Section 7: Rheumatology

CHAPTER 43: ARTHRITIS

Q.1. A 28-year-old law student comes to your office complaining of back pain. He has been reading information on the Internet and is concerned that he has ankylosing spondylitis. Which of the following symptoms would support that diagnosis?

A. Back pain worsening on forward flexion with radiation to the buttocks

B. Severe back pain in the evening

C. Morning stiffness for two hours in the back that improves with exercise

D. Diarrhea alternating with constipation

E. Acute onset of pain after lifting

Answer: C. Ankylosing spondylitis is an inflammatory process, and thus pain is worse in the morning and is associated with prolonged morning stiffness. The other symptoms can be present in mechanical back problems.

Q.2. A bad batch of potato salad at a company picnic causes 100 people to develop severe gastroenteritis. A 36-year-old man recovers from the diarrhea, but two weeks later develops arthritis in his right knee and severe Achilles tendinitis. What is the most likely diagnosis?

A. Septic arthritis likely from staphylococcal food poisoning

B. Acute gout caused by dehydration and stress

C. Lyme disease contracted at the picnic

D. Reactive arthritis

E. Osteoarthritis

Answer: D. This is likely a reactive arthritis as a sequela from his gastroenteritis. The infection to which the arthritis is "reactive" has resolved. As is often the case, he has no other features of Reiter's syndrome (e.g., pustular rash on the palms, conjunctivitis, or urethritis). The arthritis will generally resolve over several days to a few weeks but may reoccur.

Q.3. A 52-year-old man has a 10-year history of initially episodic but now chronic, seronegative polyarthritis involving his hands, knees, and first MTP joints. He has now developed nodules. NSAIDs were effective, but not hydroxychloroquine or MTX for six months. Why?

A. He has gout

B. Dose of DMARD was inadequate

C. Seronegative RA is not responsive to DMARDs

D. He has psoriatic arthritis

E. Duration of DMARD trial was inadequate

Answer: A. Tophi may be mistaken for rheumatoid nodules in patients with gouty polyarthritis. The clue to this mistaken identity is the observation that this "nodular arthritis" is "seronegative," while patients with rheumatoid nodules are expected to be rheumatoid factor positive.

Q.4. A 21-year-old man presents with chronic right knee synovitis for three weeks. He has had no uveitis or urethritis, and has otherwise been in normal health. His diagnostic evaluation should include

A. Complete skin exam for psoriatic plaques

B. Lyme serology

C. Sacroiliac joint radiographs

D. Endoscopic evaluation for occult IBD

E. All of the above

Answer: E. Psoriatic arthritis, Lyme disease, occult inflammatory bowel disease, and any of the other seronegative spondyloarthropathies can present with an isolated knee joint arthritis.

Q.5. A 21-year-old woman presents with fever, migratory arthralgias/arthritis, and scattered sites of pustular "folliculitis" for five days. She is sexually active with two partners, but has had no vaginal discharge or urethritis. She will most likely respond to treatment with

A. Antibiotics for Lyme disease

B. NSAIDs for parvovirus B19

C. High-dose steroids for vasculitis

D. Antibiotics for disseminated gonococcal infection

E. Nothing; she is likely malingering

Answer: D. The triad of migratory joint symptoms, rash, and fever is typical of disseminated gonococcal infection (DGI), as is the absence of genitourinary symptoms in affected women.

Q.6. A 52-year-old man with no significant past medical history presents complaining of the acute onset of pain and swelling of the left knee. He also notes feeling tired and having a low-grade fever. He denies any recent trauma. He denies any penile discharge or history of sexually transmitted disease. He is married and reports being monogamous. His physical examination reveals a temperature of 100.5□□F, pulse of 90, and blood pressure of 140/85. Physical examination reveals swelling, erythema, warmth, and tenderness of the left knee. The pain is

exacerbated by flexion/extension of the knee. His blood work reveals a WBC of 12,000/□L with 80% PMNs. Uric acid level is 10.4 mg/dL. An arthrocentesis is performed and reveals: 10,000 WBC/□L with 75% PMNs; crystal exam positive for negative birefringent, needle-shaped crystals inside PMNs; and Gram stain—many PMNs but no organisms identified. Which of the following treatments is most appropriate?

A. Allopurinol

B. Colchicine

C. Intra-articular steroid injection

D. B or C

E. A, B, or C

Answer: D. This patient has findings consitent with an acute gout attack. Treatment options would include nonsteroidal anti-inflammatory agents, colchicines, and/or an intra-articular steroid injection. If numerous joints are involved, systemic steroids are an option. Allopurinol has no role in the acute management of gout.

Q.7. A 45-year-old man with no significant past history presents for a physical. His only complaint is pain in both his hands that has been going on for the last five years. He denies any morning stiffness. He denies any joint swelling. He also denies alcohol use, drug use, or occupational exposures. His physical examination reveals prominence of his second and third MCP joints bilaterally. There is no active synovitis. Laboratory studies reveal the following:

WBC6800

HCT 42%

MCV 90 fl

PLT 250K

AST 100 U/L

ALT 97 IU/L

Alkaline phosphatase 60 IU/L

Na 140 mEq/L

K 4.9/L

Cr 0.9 mg/dL

Rheumatoid factor negative

Glucose 105 mg/dL

What is the most likely diagnosis?

A. Hemochromatosis

B. Wilson’s disease

C. □-1-antitrypsin deficiency

D. Seronegative rheumatoid arthritis

E. Primary biliary cirrhosis

Answer: A. This patient presents with arthralgias, elevated hepatic enzymes, and prominence of his MCP joints. Hemochromatosis can present with arthritis or arthralgias in up to 50% of patients. A high iron level, saturation, and ferritin level would be expected. This is not a typical presentation for either Wilson's disease or alpha-1-antitrypsin deficiency. Although he does have symmetrical prominence of his MCP joints, he does not meet the criteria for RA. Primary biliary cirrhosis usually affects women and causes an elevation in the alkaline phosphatase (which is not seen in this patient).

CHAPTER 44: OFFICE ORTHOPEDICS

Q.1. A 52-year-old man presents to you with the complaint of lateral elbow pain after a long weekend of fly-fishing. He has point tenderness over the lateral aspect of the elbow, with pain exacerbated on resisted wrist extension. This patient has

A. Lateral epicondylitis

B. Medial epicondylitis

C. Ulnar neuropathy

D. Olecranon bursitis

E. Gouty arthritis of the elbow

Answer: A. These symptoms and findings are typical of lateral epicondylitis. Medial epicondylitis presents with medial elbow pain and tenderness, provoked by resisted wrist flexion. Ulnar neuropathy would present with pain, paresthesias, or numbness in the ulnar nerve distribution. Olecranon bursitis and gouty arthritis would both be expected to have inflammatory findings (heat, erythema, and swelling), the former over the olecranon process, and the latter within either the joint or olecranon bursa.

Q.2. A 42-year-old woman presents with the complaint of anterior shoulder pain that is worse when lifting overhead or carrying something in front of her body. The pain is brought on with resisted supination of the forearm (Yergason's test). This patient has

A. Rotator cuff tendonitis

B. Complete rotator cuff tear

C. Adhesive capsulitis of the shoulder

D. Bicipital tendonitis

E. Cervical radiculopathy

Answer: D. Bicipital tendonitis presents as described in this case, with Speed''s and Yergason''s tests providing clinical confirmation. Rotator cuff tendonitis would be more likely to cause lateral pain, and with abduction rather than the anterior motion described. A complete rotor cuff tear might prevent her from abducting her arm. Adhesive capsulitis would virtually immobilize the joint. No radiculopathic symptoms are described.

Q.3. A 43-year-old man presents with a four-day history of back and left buttock pain, with radiation down the leg. Examination shows decreased knee reflex with loss of sensation over the medial aspect of the ankle. The patient has a positive straight-leg- and crossed straight-leg-raising test. Which of the following is false?

A. This is most likely an L4 nerve root compression.

B. The majority of patients with these symptoms will improve with time and rest.

C. Operative intervention will prevent the recurrence of future events.

D. The presence of a crossed straight-leg-raising test would support the clinical diagnosis of lumbar radiculopathy

Answer: C. Operative intervention will prevent the recurrence of future events. The majority of low back pain is due to self-limited conditions, so moving directly to surgery without evidence of a worrisome etiology (see Table 44-5) is premature. There is no reason at this point to believe that the current symptoms might not resolve spontaneously, as even ruptured disc material may be resorbed.

Q.4. A 46-year-old man presents with a three-day history of left knee pain after playing tennis. Examination of the knee demonstrates an effusion. There is medial knee pain with medial torsion applied to the knee as it is moved from flexion to extension (positive McMurray’s sign). This patient most likely suffers from

A. Medial collateral ligament injury

B. Medical meniscus injury

C. Rupture of the anterior cruciate ligament

D. Prepatellar bursitis

E. Rupture of the posterior cruciate ligament

Answer: B. The history of “trauma” followed by the development of pain, an effusion, and a positive McMurray sign are characteristic of a meniscal injury. These patients are also likely to demonstrate tenderness along the joint line. Valgus stress on the knee would be expected to exacerbate the pain from a medial collateral ligament injury. Prepatellar bursitis presents with fullness anterior to the patella, but no knee joint effusion. Anterior or posterior cruciate ligament injuries would be manifested as instability of the knee (anterior and posterior drawer signs).

Q.5. A 62-year-old man with osteoarthritis presents with a two-day history of knee pain and calf swelling that were acute and apparently spontaneous in onset. Examination is remarkable for a small knee effusion, swollen calf, and ecchymoses at the ankle. This patient has

A. Anserine bursitis

B. Deep venous thrombosis

C. Ruptured Baker''s cyst

D. Achilles'' tendonitis

E. Plantar fasciitis

Answer: C. Ruptured (popliteal) Baker's cyst (or "pseudo-thrombophlebitis") may track into the thigh or more commonly the calf. Ultrasound is useful in distinguishing true deep venous thrombosis from a ruptured Baker's cyst. Anserine bursitis presents with anteromedial pain, just distal to the knee joint margin. Achilles' tendonitis and plantar fasciitis present with symptoms involving the posterior aspect of the ankle and the plantar surface of the foot, respectively.

Q.6. A 26-year-old woman presents with a three-week history of burning pain between her third and fourth toes. She recalls no injury and the area is normal to visual inspection. Her symptoms are reproduced by deep palpation of the web space between toes 3 and 4. This process is most consistent with the diagnosis of

A. Tarsal tunnel syndrome

B. Rheumatoid arthritis

C. Gout

D. Morton's neuroma

E. Hammer toe

Answer: D. Paresthetic pain between the third and fourth toes, exacerbated by deep palpation or walking, is characteristic of Morton's neuroma, caused by compression of the digital nerves. Tarsal tunnel syndrome causes pain over the plantar surface of the foot. There is no evidence of an inflammatory process consistent with the diagnosis of rheumatoid arthritis. The tip of a hammer toe may be tender due to local trauma with weight-bearing.

Q.7. An 83-year-old woman complains that she is no longer able to lie in a left lateral decubitus position to sleep, due to pain. She recalls no injury. She was seen in an urgent care facility yesterday where hip radiographs revealed only mild superior acetabular sclerosis and joint space narrowing. Which maneuver(s) would you expect to provoke her symptoms?

A. Internal and external rotation of the hip

B. Palpation of the greater trochanter

C. Resisted abduction of the hip

D. B and C

E. None of the above

Answer: D. This patient has trochanteric bursitis, and both B and C would be expected to provoke her symptoms. Pain on internal and external rotation of the hip is characteristic of hip joint arthritis.

Q.8. A 35-year-old man complains of pain affecting the plantar surface of his right foot in the morning when he first bears weight. The symptoms then resolve within a few minutes after he begins ambulating. He weighs 287 pounds and has a family history of ankylosing spondylitis, though he has been free of any low-back symptoms. What is the cause of his symptoms?

A. Tarsal tunnel syndrome

B. Plantar fasciitis

C. Ankylosing spondylitis

D. Morton''s neuroma

E. Tarsal coalition

Answer: B. The presence of plantar pain with first weight-bearing, which then resolves with continued use, is characteristic of plantar fasciitis. While plantar fasciitis may present as an enthesopathic manifestation of ankylosing spondylitis, there is no other reason to suspect this diagnosis on clinical grounds. Both tarsal tunnel syndrome and Morton's neuroma, would be expected to present with paresthetic pain. Tarsal collation would present with mechanical foot pain to this developmental abnormality of the tarsal bones.

CHAPTER 45: VASCULITIS

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Q.1. A 74-year-old white woman presents with a four-week history of headache, scalp tenderness when combing her hair, and pain in her jaw when chewing. Her examination, however, reveals no temporal artery tenderness and her erythrocyte sedimentation rate is 45 mm/hour. You suspect that she may have giant cell arteritis, but you are reluctant to start steroids without more objective information. A temporal artery biopsy is performed and found to be floridly positive. What percentage of cases of giant cell arteritis is associated with an erythrocyte sedimentation rate of less than 50 mm/hour?

A. 75%

B. 50%

C. 10%

D. 0%

E. None of the above

Answer: C. The erythrocyte sedimentation rate is a useful test in giant cell arteritis, but it is not perfect. Most patients have very elevated erythrocyte sedimentation rates, but in approximately 10% of cases the ESR is less than 50 mm/hour (a level not strikingly abnormal in the elderly population that giant cell arteritis usually afflicts).

Q.2. Which disorder occurs predominantly in children?

A. Wegener's granulomatosis

B. Takayasu's arteritis

C. Behçet's disease

D. Henoch-Schönlein purpura

E. Both C and D

Answer: D. Henoch-Schönlein purpura tends to occur in children but may be underdiagnosed in adults with cutaneous vasculitis because of failure to perform direct immunofluorescence on skin biopsies.

Q.3. Which form of vasculitis does not usually involve large arteries?

A. Takayasu's arteritis

B. Giant cell arteritis

C. Cogan's syndrome

D. Cryoglobulinemia

E. All of the above are large vessel vasculitides

Answer: D. Cases of cryoglobulinemic vasculitis, 90% of which are associated with hepatitis C, are associated with involvement of small- and medium-sized vessels. Consequently, cutaneous involvement in cryoglobulinemia is very common, and the lesions include palpable purpura (often with small central foci of necrosis), urticaria, vesiculo-bullous lesions, nodules, and ulcers. Other manifestations of cryoglobulinemia (e.g., vasculitic neuropathy and glomerulonephritis) also result from vasculitis of small and medium-sized vessels. Takayasu's arteritis, giant cell arteritis, and Cogan's syndrome, in contrast, are large vessel vasculitides and therefore more likely to involve the aorta and its large branches.

Q.4. A 50-year-old woman comes to your office with complaints of severe joint pain and stiffness in multiple joints including her shoulders, wrists, hands, and feet, bilaterally. She has morning stiffness lasting several hours and severe fatigue for the past two years. She has been told that she has rheumatoid arthritis (RA). She

has no other prior history except surgery following a severe automobile accident in the 1970s. On examination, she has some joint tenderness but no synovitis and hyperpigmentation on her lower extremities. Her laboratory examination shows a positive rheumatoid factor at 89 IU/mL (normal <21 IU/mL), 2+ proteinuria without active urinary sediment, normal serum creatinine, ESR of 80 mm/hr, and an ALT of 86 U/L. What is your plan?

A. Start methotrexate for severe rheumatoid arthritis

B. Start hydroxychloroquine for mild rheumatoid arthritis

C. Draw hepatitis serologies

D. Obtain x-rays of her hands and wrists to look for erosions

E. Order gonococcal cervical cultures

Answer: C. Although she is the right host for RA, and her joint symptoms are compatible with the diagnosis of RA, renal disease is not a feature of RA, nor is her lower extremity rash. Her diagnosis is likely chronic hepatitis C with cryoglobulinemia. Her lower extremity hyperpigmentation is the result of recurrent purpura. Many cryoglobulins have rheumatoid factor activity. Arthralgias without synovitis are a common complaint in chronic hepatitis C. Her 2-year course makes gonococcal disease unlikely.

Q.5. A 35-year-old man presents with a three-year history of recurrent painful aphthous oral ulcers, a two-month history of scarring scrotal ulcers, and a one-week history of visual blurring in the right eye. He is seen urgently by his ophthalmologist, who notes a hypopyon and posterior uveitis marked by retinal vasculitis and perivascular changes, both on the right. Which form of vasculitis is most likely to cause this patient's posterior uveitis (retinal vasculitis)?

A. Behçet's disease

B. Giant cell arteritis

C. Churg-Strauss syndrome

D. Microscopic polyangiitis

E. Polyarteritis nodosa

Answer: A. One of the most serious complications of Behçet's disease is eye involvement. Behçet's can cause both anterior and posterior uveitis. Anterior uveitis is usually highly symptomatic. In contrast, posterior uveitis or retinal vasculitis may be clinically silent until substantial retinal damage has occurred. The ocular complications of Behçet's disease, if untreated, may lead to blindness. Blindness may also result from giant cell arteritis, of course, but the usual mechanism of visual loss in that disease is anterior ischemic optic neuropathy caused by occlusion of the ophthalmic or posterior ciliary arteries. Central retinal artery occlusion may also occur in giant cell arteritis. Uveitis, however, is not

associated with giant cell arteritis and is an exceptionally unusual complication of the other disorders listed as well.

Q.6. A 48-year-old male account executive complains of the sudden onset of pain and weakness in his left foot and right hand. On further history-taking, he reports that the hand had become weak first but he hadn't paid too much attention to it because he is left-handed. He had also noted increased numbness and tingling in all of his extremities, but he had attributed this to his 16-year history of type 2, insulin-reliant diabetes. Of further concern, the patient reports that over the past couple of years he has developed significant problems with adult-onset asthma, requiring several emergency room visits and frequent glucocorticoid tapers. Physical examination shows significant weakness of the foot dorsiflexors on both sides, left greater than right, a right wrist drop, and weakness of the right hand grip. There is a small cluster of three to four nodules on the left elbow, and one nodule on the right. You are now suspicious that a form of systemic vasculitis ties together many of the patient's complaints. Which test in the evaluation test would help confirm your suspicion?

A. Rheumatoid factor

B. Anti-CCP antibodies

C. A complete blood count with differential

D. An antinuclear cytoplasmic antibody leading to C-ANCA immunofluorescence

E. An erythrocyte sedimentation rate

Answer: C. The patient's adult-onset asthma, nodules over the extensor surfaces of the elbows (probably Churg-Strauss granulomas), and mononeuritis multiplex are all consistent with a diagnosis of the Churg-Strauss syndrome. On the patient's complete blood count, one would expect to see peripheral eosinophilia, with eosinophils comprising between 10% and 60% of the peripheral white blood cells. The diagnosis of Churg-Strauss syndrome could be clinched by biopsy of one of the skin nodules.

Q.7. A 27-year-old woman recently delivered a healthy baby girl. During her pregnancy, it was noted on many occasions that her blood pressure was extremely difficult to detect in both arms, but her pregnancy was uncomplicated. The patient has been quite healthy for the past several years, although while she was in college she had experienced an inflammatory illness lasting for several months, characterized by fevers, weight loss, and profound malaise. On examination, the both the brachial and radial pulses are absent bilaterally, but the pedal pulses are intact. Blood pressure taken in the left leg is 190/80 mm Hg. Auscultation of the large vessels reveals bilateral subclavian bruits and a faint diastolic murmur at the right upper sternal border. What diagnostic test is an essential next step for this patient?

A. Measurement of acute phase reactants, to determine whether or not she would benefit from immunosuppressive agents

B. A purified protein derivative (PPD) test

C. A chest x-ray, to exclude dilatation of the ascending aorta

D. A lipid profile and further querying about her family history of early atherosclerotic disease

E. An aortogram, with measurement of her central aortic pressures

Answer: E. This patient's history and physical examination are strongly consistent with Takayasu's arteritis, also known as "pulseless disease." The stenotic phase of the illness is often preceded by several years by an inflammatory phase characterized by the kinds of symptoms the patient experienced in college. The absence of pulses in the upper extremities, caused by narrowing of the subclavian arteries, is far more common than pulselessness in the feet. (In this sense, Takayasu's arteritis is the opposite of coarctation of the aorta). The descending aorta, iliac, and femoral arteries may also be involved in Takayasu's arteritis, rendering even blood pressure measurements obtained in the legs unreliable. Because involvement of the renal arteries can lead to a rennin-mediated hypertension, patients with Takayasu's arteritis are at substantial risk for developing blood pressure elevations that are undetected by conventional measurements. At diagnosis, therefore, and possibly at occasional intervals over the course of management, patients should undergo full aortography with runoff, evaluating the ascending and descending aorta, the aortic arch, and the primary branches off of the aorta (particularly the renal arteries). In this case, the patient's physical findings are concerning for both aortic regurgitation (often caused by dilatation of the ascending aorta) and renal artery involvement, both complications that may require thoughtful intervention.

Q.8. A 37-year-old man has suffered from nonhealing ulcers over the distal lower extremities for six months. When they began on one leg just above the medial malleolus, they were attributed to infection but failed to respond to antibiotics. More recently the question of Buerger's disease has been raised because the patient smokes a half pack of cigarettes per day. The patient reports that the lesions usually begin as tender red nodules, but then ulcerate and expand. The patient is of Western European heritage, and denies oral and genital ulcers. On examination, the patient has a half-dozen or more ulcerated lesionsranging between 0.5 and 1 cm over the dorsum of both feet, as well as larger lesions over the left lateral malleolus and right medial malleolus. There are several areas on the lower extremities that appear to be healed ulcers, as well. Which form of vasculitis is a better explanation for the patient's chronic ulcers than Buerger's disease?

A. Behçet's disease

B. Cholesterol emboli

C. An ANCA-associated vasculitis

D. Polyarteritis nodosa

E. Hepatitis C-related cryoglobulinemia

Answer: D. Polyarteritis nodosa involves medium-sized muscular arteries, precisely the types that are involved in the ulcerated lesions described in this case. Buerger's disease tends to involve most intensely the arteries at the levels of the wrists and ankles, leading to profound digital ischemia but not to ulcerations in this patient's distribution. Behçet's disease can be associated with erythema nodosum–like lesions that lead to breakdown and ulceration, but the absence of oral and genital ulcers (particularly oral ulcers) make this diagnosis unlikely.

Q.9. Which form of vasculitis is most likely to be associated with a saddle nose deformity?

A. Polyarteritis nodosa

B. Wegener's granulomatosis

C. Churg-Strauss syndrome

D. Urticarial vasculitis

E. None of the above

Answer: B. Ninety percent of patients with Wegener's granulomatosis have involvement of the nose. Nasal congestion, bloody nasal discharge, nasal crusting, and epistaxis are all common symptoms of this disease. Wegener's granulomatosis may lead to destruction of the nasal septum, causing nasal septal perforation, and in advanced cases results in collapse of the nasal bridge resulting in a "saddle nose deformity."

Q.10. Coronary artery aneurysms are a frequent complication of which type of vasculitis?

A. Kawasaki's disease

B. Takayasu's arteritis

C. Henoch-Schönlein purpura

D. Wegener's granulomatosis

E. Buerger's disease

Answer: A. Coronary artery aneurysms may be a lethal complication of Kawasaki's disease. This complication may be prevented by the timely administration of intravenous immunoglobulin in children with this illness.

Q.11. Which type of vasculitis often requires cyclophosphamide in combination with steroids as treatment?

A. Giant cell arteritis

B. Buerger's disease

C. Hypersensitivity vasculitis

D. Wegener's granulomatosis

E. Henoch-Schönlein purpura

Answer: D. Cyclophosphamide is required for the effective treatment of many patients with Wegener's granulomatosis, particularly those with glomerulonephritis, pulmonary hemorrhage, vasculitic neuropathy, and potentially destructive ocular complications such as scleritis. In such patients, steroids alone are insufficient to induce disease remission. Giant cell arteritis responds promptly to steroid treatment in nearly all cases, and seldom requires a steroid-sparing agent. The only treatment for Buerger's disease is smoking cessation. Hypersensitivity vasculitis usually resolves after removal of the offending agent (e.g., a medication), but short courses of steroids are sometimes required in severe cases. Henoch-Schönlein purpura is usually a self-limited illness that resolves within several weeks, and in most cases requires no treatment.

Q.12. A 27-year-old man presents with a three-month history of fevers, a 12-pound weight loss, numbness over his left calf, and left testicular pain. His physical examination is noteworthy for his chronically ill appearance, blood pressure of 163/103 mm Hg in the absence of a prior history of hypertension, diminished sensation to pinprick over his left calf, and left testicular tenderness without palpable masses. His labs are notable for a hematocrit of 32.3% and mean corpuscular volume of 89 fl. Which of the following diagnoses best explains his testicular pain?

A. Henoch-Schönlein purpura

B. Kawasaki's disease

C. Polyarteritis nodosa

D. Takayasu's arteritis

E. Giant cell arteritis

Answer: C. Testicular pain due to vasculitic involvement is a classic finding in polyarteritis nodosa. None of the other vasculitides listed typically involve this organ.

Q.13. A 72-year-old white woman presents with a three-week history of headaches, scalp tenderness, jaw claudication, and transient visual loss the evening before her appointment with you. Which of the following laboratory studies is least likely to be abnormal?

A. Hematocrit

B. Alanine aminotransferase

C. Erythrocyte sedimentation rate

D. Aspartate aminotransferase

E. Mean corpuscular volume

Answer: E. While low-level anemia is commonly seen in giant cell arteritis, this is generally normochromic and normocytic. The ESR is typically elevated in this disease process, as are the hepatic transaminases.

CHAPTER 46: SELECTED TOPICS IN RHEUMATOLOGY

Q.1. A previously healthy 46-year-old right-handed woman awakens with right-sided weakness and presents to the emergency room. Her past history is of note only for childhood asthma. She is on no medicines. She is afebrile on physical examination; blood pressure is 128/76 mm Hg, pulse is 76, and respiratory rate is 14. She has an expressive aphasia and right hemiparesis. Laboratory studies show WBC 6000, Hct 36, platelets 90,000, PT 12, aPTT 48. ANA and FTA are negative, RPR is 1:8, and head CT scan shows infarct left hemisphere. What is the most likely diagnosis?

A. Systemic lupus erythematosus

B. Primary CNS angiitis

C. Primary antiphospholipid antibody syndrome

D. Churg-Strauss syndrome

E. Thrombotic thrombocytopenic purpura

Answer: C. This woman has primary antiphospholipid antibody syndrome, a cause of stroke. The negative ANA makes SLE very unlikely. Thrombocytopenia, biologic false positive test for syphilis (BFP-STS), and prolonged aPTT indicate the presence of antiphospholipid antibodies. Primary CNS angiitis is not usually associated with antiphospholipid antibodies. Churg-Strauss syndrome rarely causes stroke. Thrombotic thrombocytopenic purpura does not affect coagulation assays.

Q.2. A 38-year-old woman presents with a chief complaint of hand "tightness." She was well until about 10 months ago, when she had an episode of painful blue discoloration (cyanosis) of her right third digit. Eight months ago, she began to note cold sensitivity and cyanosis in all digits, with occasional digital tip ulcerations. Six months ago she noted the onset of proximal muscle weakness. On physical examination she has no heliotrope rash or Gottron's papules, and

sclerodactyly, periungual erythema, and telangiectasias present, along with proximal weakness. The following statements are true except

A. Raynaud's phenomenon is characteristic

B. Lab abnormalities include increased CPK

C. Typical lab abnormalities include anti-Ro

D. Calcium channel-blockers may be helpful

E. Prednisone useful for managing myositis

Answer: C. The objective here is to recognize the clinical and laboratory features of scleroderma/systemic sclerosis. Raynaud's phenomenon is usually present in patients with scleroderma (Answer A), and calcium channel-blockers (Answer D) may be helpful (although these agents are often more beneficial in patients with primary Raynaud's phenomenon). Proximal muscle weakness suggests the emergence of myositis, which would be associated with elevated muscle enzymes (including CPK), and for which prednisone is appropriate treatment (Answer B). Anti-Ro antibodies associate with photosensitivity, sicca symptoms, subacute cutaneous lupus and neonatal lupus and would not be expected in scleroderma, thereby making Answer C the correct choice.

Q.3. A 63-year-old man presents with joint pain and weakness. His past history is of note only for having been a heavy smoker for years, but he has no cough, dyspnea, or chest pain. A few weeks before evaluation, he became aware of malaise and some generalized musculoskeletal aching. Ten days ago, he had arthralgias in his fingers, shoulders, and knees, but he was not aware of any joint swelling or warmth. Seven days ago, he noted trouble climbing stairs, rising from a chair, and reaching above his head. Physical examination reveals a temperature of 100° F, and pain on motion of fingers and shoulders, but no warmth or synovitis. He has slight tenderness in his right deltoid and weakness in his proximal muscles, and upper and lower extremities. His distal strength is excellent. He has trouble clearing his throat. Which of the following is not likely?

A. Labs indicate CPK 4200, aldolase 26

B. CXR reveals single large nodule

C. Surgical resection/biopsy is necessary

D. Labs indicate positive centromere

E. High-dose prednisone is indicated

Answer: D. This patient's labs indicate positive centromere, which suggest features of myositis and possible association with malignancy. Proximal weakness, muscle tenderness, and dysphagia suggest myopathy; no features suggest another specific process. Elevated CPK and aldolase are consistent with myositis, and the absence of cutaneous features makes dermatomyositis less likely. In patients over 50 years old with new myositis, the possibility of an associated

malignancy should be considered, and age and/or risk-factor-appropriate screening should be undertaken. Biopsy of a suspicious lesion is important to confirm diagnosis of malignancy. High-dose steroids are the initial treatment of choice for myositis. There are no features to suggest the CREST syndrome or limited scleroderma, which would be associated with anticentromere antibodies.

Q.4. A 53-year-old woman presents with profound fatigue and diffuse muscle pains. She reports that her symptoms began about six months ago, when she began to note malaise, fatigue, and generalized aching. She reports that she awoke feeling stiff and like she had been "run over by a truck." She has tried ibuprofen and naproxen without any benefit. She has not had headache, fever, weight loss, or joint swelling. Physical examination reveals no synovitis, xxcellent muscle strength, marked tenderness over muscle groups, bilaterally, and upper and lower extremities. Her labs show Hct 39, ESR 35, CPK 90, and ANA positive 1:80. What should you do next?

A. Schedule EMG and NCV

B. Schedule temporal artery biopsy

C. Start celecoxib, 100 mg twice daily

D. Start amitriptyline, 25 mg at bedtime

E. Start prednisone, 20 mg daily

Answer: D. This patient should be started on amitriptyline, 25 mg at bedtime for the management of fibromyalgia with some abnormal (although nonspecific) laboratory results. There is no weakness suggestive of an inflammatory myopathy (although resistive strength testing can sometimes be limited by pain), and the CPK is normal. There are no cephalgic symptoms suggestive of giant cell arteritis. The slightly elevated ESR is essentially normal for her age. The positive ANA is low titer, and by itself in this setting is not specific for diagnosis. There is no proximal distribution to the muscle symptoms, which would be expected in polymyalgia rheumatica (and which would warrant low-dose steroids). NSAIDs are often minimally helpful in fibromyalgia. This woman's history of diffuse pain and nonrestful or nonrestorative sleep, coupled with a physical examination that does not localize the pain, is characteristic of fibromyalgia. Thus tricyclic agents, as well as aerobic exercise, can be beneficial.

Q.5. A 28-year-old woman is referred for evaluation of possible SLE. She was well until three months ago, at which time she had a self-limited illness characterized by fever, rash, malaise, myalgias, and pleurisy. Laboratory studies at the time revealed leukopenia and positive ANA. Her family history is notable for a cousin with SLE. Physical examination reveals nontender cervical adenopathy, but is otherwise unremarkable. Laboratory studies reveal WBC 3100/mm^3, PCV 43%,

platelets 185,000/mm^3, ESR 23 mm/hr, an unremarkable urinalysis, ANA positive 1:640 homogeneous, and negative anti-dsDNA.What do you conclude?

A. She has a nonspecific viral illness, with incidentally abnormal labs, but her symptoms will resolve spontaneously and with time

B. She needs additional evaluation

C. Some features of her presentation are suggestive of SLE (or another rheumatic disease), but are insufficient for diagnosis at this time

D. She has definite SLE

E. She has Lyme disease

Answer: C. The objective here is to recognize a lupus-like illness. Physicians are often asked to evaluate for "probable," "borderline," or "incomplete" lupus. Patients and physicians may refer to such patients as having lupus, which has serious implications at many levels. Most do not develop classic SLE and do well—a clinically important distinction. This case demonstrates features consistent with but not diagnostic of SLE (making Answer C the correct choice). The situation is not a viral illness, Lyme disease, or some other rheumatic disease. Careful follow-up, rather than extensive testing now, is appropriate.

Q.6. A 19-year-old woman with a history of SLE presents with new chest pain. Her lupus began 12 years ago with polyarthritis, cutaneous vasculitis, positive ANA, positive anti-dsDNA, and a renal biopsy showing diffuse proliferative glomeulonephritis. She had deep vein thrombophlebitis nine years and seven years ago. One week ago, she developed fever and right-sided pleuritic pain. On physical examination, she has a temperature of 38.5° C and respiratory rate of 30/min with poor inspiratory effort. Laboratory studies reveal WBC 6000/mm^3, PCV 25%, platelets 139,000/mm^3, aPTT 42 seconds, and 24-hour urinary protein excretion is 2.7 g. Which of the following statements is not true?

A. SLE flare is likely

B. Cyclophosphamide is indicated for treatment of the lupus nephritis

C. Antiphospholipid antibodies are probably present

D. A deep venous thrombosis (DVT) and/or pulmonary emboli are likely

E. There is no role for anticoagulation

Answer: E. This patient presents with problems associated with antiphospholipid antibodies in the setting of background/active SLE. Active lupus is a consideration in view of the proteinuria, fever, and pleuritic pain. Lupus nephritis (especially diffuse proliferative glomerulonephritis) is an indication for cyclophosphamide therapy. The prolonged aPTT and thrombocytopenia are consistent with the presence of antiphospholipid antibodies. Thrombotic events, including DVT and pulmonary emboli, are frequent in antiphospholipid antibody syndrome. The clinical situation is consistent with antiphospholipid antibody

syndrome and vascular events, therefore anticoagulation is warranted, with a target INR of greater than 3.

Q.7. A 36-year-old woman is referred for evaluation because of a five-year history of Raynaud's phenomenon and thickening of the skin over her fingers. Two years ago, she developed several 1- to 2-mm patches of calcinosis on the distal fingers and dorsum of her right hand, and these have persisted. She reports mild dyspnea when climbing stairs. She takes a proton pump inhibitor for reflux esophagitis. On physical examination, there is sclerodactyly, with one digital tip ulcer and a few digital pit scars, minimal calcinosis, and two telangiectasias on her face. Her lungs and heart are unremarkable. Her chest radiograph is without infiltrates or interstitial fibrosis, and the heart size is normal. Routine laboratory tests are within normal limits. An echocardiogram suggests mild right ventricular diastolic dysfunction. The diffusing capacity for carbon monoxide (DLco) is 44% of normal. Within the next five years, she is at high risk for developing which of the following?

A. Fine, dry crackles indicative of pulmonary fibrosis

B. Renovascular hypertension with hyperreninemia

C. Sclerodermatous changes on the forearm and face

D. Pulmonary hypertension

E. Esophageal varices

Answer: D. This woman clearly presents with predictors of pulmonary hypertension in limited forms of scleroderma (systemic sclerosis). Pulmonary hypertension (PHT) is the leading cause of scleroderma-related mortality in patients with the limited cutaneous form of scleroderma. The DLco may be significantly reduced for many years before the diagnosis of PHT is made, even in the absence of interstitial fibrosis. The importance of suspecting, and then diagnosing, PHT in this form of the disease is that potent pulmonary vasodilators and lung transplantation have improved patient well-being and, perhaps, survival. In addition, an orally administered dual endothelin-receptor antagonist (Bosentan) has been shown to improve exercise capacity and cardiopulmonary hemodynamics in patients with pulmonary hypertension. Patients with limited cutaneous scleroderma rarely develop interstitial fibrosis, and are not at risk for scleroderma renal crisis with malignant hypertension. Those with this form of scleroderma do not develop skin changes other than sclerodactyly. Telangiectasias are part of the limited scleroderma or CREST syndrome, and are not the result of liver disease, portal hypertension, or esophageal varices.

Q.8. Which of the following factors is predictive of an unfavorable outcome in adults with polymyositis or dermatomyositis?

A. Presence of a rash

B. Level of increase in serum creatine kinase

C. Presence of dysphagia

D. Age at onset

E. Absence of myositis-specific antibodies

Answer: C. The objective here is to recognize predictors of outcome in adults with inflammatory myopathy. The presence of dysphagia predicts a complicated and difficult course in myositis; none of the other variables do so. Myositis-specific antibodies are associated with interstitial lung disease, and their absence is a good prognostic sign.

Q.9. A 75-year-old woman is referred for evaluation because of persistent pain, fatigue, and laboratory abnormalities. Three months ago, she was diagnosed with polymyalgia rheumatica, based on diffuse pain in the neck and shoulder girdle, and an elevated erythrocyte sedimentation rate. She has been treated with prednisone in the past, as much as 20 mg daily, but the diffuse pain has persisted and progressed to involve her lower back and buttocks. She is able to perform her activities of daily living, but movements are painful. She is constantly fatigued and reports that her sleep is not restful. She notes low-grade fevers in the mornings. She currently takes no medications; her symptoms did not increase when the prednisone was tapered off over a two-week period. On physical examination, there is no scalp tenderness. Her temporal arteries are palpable, firm, and nontender. There is no muscle weakness or synovitis. Laboratory studies reveal a WBC of 12,400/mm^3, PCV 24.2%, ESR 98 mm/hr, fasting glucose 143 mg%, and serum creatinine 0.8 mg%. Bilateral temporal artery biopsy specimens reveal atherosclerotic changes but no vasculitis. Which of the following is the most appropriate next step?

A. Restart prednisone

B. Start amitriptyline at bedtime

C. Start prednisone and methotrexate

D. Prescribe a short-acting hypnotic agent at bedtime and an aerobic exercise program

E. Evaluate the elevated ESR

Answer: E. This complicated case illustrates several important points, the most important of which is that an ESR as high as 98 mm/hr must be considered a sign of serious organic disease, and its cause must be sought, particularly when a significant anemia is also present. Polymyalgia rheumatica and fibromyalgia are the most common causes of pain syndromes in the elderly. It would be unusual for polymyalgia rheumatica not to improve with prednisone 20 mg daily. Fibromyalgia can exist in a patient who has other organic pathology, and the ESR in fibromyalgia only is typically not elevated. Diagnoses to be considered in this

situation include lymphoma, granulomatous disease, and carcinoma. Tricyclic agents and aerobic exercise programs can be useful for some patients with fibromyalgia. Selective serotonin reuptake inhibitors may also be effective. Methotrexate fails to substitute effectively for corticosteroid therapy in giant cell arteritis.

Q.10. Which pair does not represent a correct association?

A. Neonatal lupus : Ro

B. Lupus nephritis : antibodies to double-stranded DNA

C. Drug-induced SLE : antihistone antibodies

D. Diffuse scleroderma : anticentromere antibodies

E. Interstitial lung disease : antibodies to Jo-1

Answer: D. The objective is to distinguish among the antibodies associated with rheumatic diseases. Anti-Scl 70 (topoisomerase I) is seen in up to 50% of patients with diffuse scleroderma, whereas anticentromere antibodies are seen in patients with limited scleroderma or the CREST syndrome.

Q.11. A 36-year-old woman presents with facial telangiectasias, Raynaud's phenomenon, nodules on her digital pulp that may occasionally drain a white chalky substance, dysphagia, and waxy bound-down skin over her fingers. Which additional disorder is she likely to experience?

A. Vascular blindness

B. Pulmonary hypertension

C. Carpal tunnel syndrome

D. Meralgia paresthetica

E. Aplastic anemia

Answer: B. This patient exhibits all of the manifestations of the CREST syndrome. Although the CREST syndrome is generally less likely to have major organ system involvement than classic scleroderma, pulmonary hypertension may be seen.

Q.13. A 23-year-old woman is pregnant with her first child. She undergoes screening ultrasonography at five months gestation and the fetus is determined to have an abnormally low heart rate, although it appears to be otherwise normal. Serologic screening of the mother is most likely to reveal which of the following autoantibodies?

A. Anti-Ro

B. Anticentromere

C. Antidouble-stranded DNA

D. Anti-Jo-1

E. Anti-Mi2

Answer: A. Anti-Ro antibodies are associated with congenital heart block and cutaneous eruptions in affected neonates, as well as photosensitive eruptions in adults and Sjögren's syndrome. Anticentromere antibodies are associated with the CREST syndrome, antidouble-stranded DNA with SLE, and anti-Jo-1 and anti-MI2 antibodies with inflammatory muscle disease.