# MCQs

# Case 1

- A 54-year-old female with rheumatoid arthritis is treated with infliximab for refractory disease. ***All the following are potential side effects of this treatment except:***

A. demyelinating disorders

B. disseminated tuberculosis

C. exacerbation of congestive heart failure

D. pancytopenia

E. pulmonary fibrosis

# The answer is E.

- Serious infections are most frequently seen, with a marked increase in disseminated tuberculosis. Other side effects include:

✓ pancytopenia

✓ demyelinating disorders

✓ exacerbations of congestive heart failure hypersensitivity to the infusion or injection

✓ drug-induced systemic lupus erythematosus.

✓ An increased incidence of malignancy is of theoretical concern, but this has not been borne out in the limited follow-up of patients treated with these drugs.

# Case 2

- A 48-year-old male has a long-standing history of ankylosing spondylitis. His most recent spinal film shows straightening of the lumbar spine, loss of lordosis, and “squaring” of the vertebral bodies. He currently is limited by pain with ambulation that is not improved with non-steroidal anti-inflammatory medications. Which of the following treatments has been shown to improve symptoms the best at this stage of the illness?

A. Celecoxib

B.Etanercept

C. Prednisone

D. Sulfasalazine

E.Thalidomide

# The answer is B

- Before the introduction of anti (TNF) α therapy, the mainstay of treatment for ankylosing spondylitis was (NSAIDs) and exercise therapy. In 2000, infliximab and etanercept were introduced and since that time have been shown to confer a rapid, profound, and sustained reduction in all clinical and laboratory measures of disease activity. Even patients with long-standing disease and ankylosis show significant improvement in spinal mobility and pain relief.
- Other treatments for AS can be used, including NSAIDs and COX-2 inhibitors, to decrease pain, especially in mild cases.
- An ongoing exercise program is encouraged to maintain posture and range of motion. In patients with more severe pain, sulfasalazine or methotrexate may be added with modest benefit, especially in those with peripheral arthritis.
- ***Glucocorticoids have no role in the treatment of this disease.***

# Case 3

- A 72 y/o woman presents to the ER for an episode of vision loss in her Rt eye. The vision loss came on abruptly and is described as a curtain falling across her visual field. She immediately called her daughter and upon arrival to the ER 40 min later, her vision had returned to normal.
- Recently she also has been experiencing dull throbbing headaches for which she is taking acetaminophen, with limited relief. She has a past hx of hypercholesterolemia & CAD, undergoing PCI of the RCA 8 years previously.
- She does not smoke currently but has a 40-pack-year hx of tobacco, quitting only after her diagnosis of CAD. On review of systems, the pt recalls pain in her scalp with combing her hair, particularly on the Rt side, occasional pain with chewing food.
- She has also recently noticed stiffness and pain in her hips, making it difficult to stand from seated position. O/E, she has 20/30 visual acuity in the Lt eye, and 20/100 Rt eye.

- Funduscopic examination suggests anterior ischemic optic neuropathy. There are no carotid bruits present, but palpation of the temporal artery is painful. The neurologic examination is otherwise normal. (ESR) is 102 mm/h. The Hb is 7.9 g/dL, and Hct is 25.5%. A head CT shows no acute ischemic event. ***Which of the following is the next most important step in the management of this patient?***

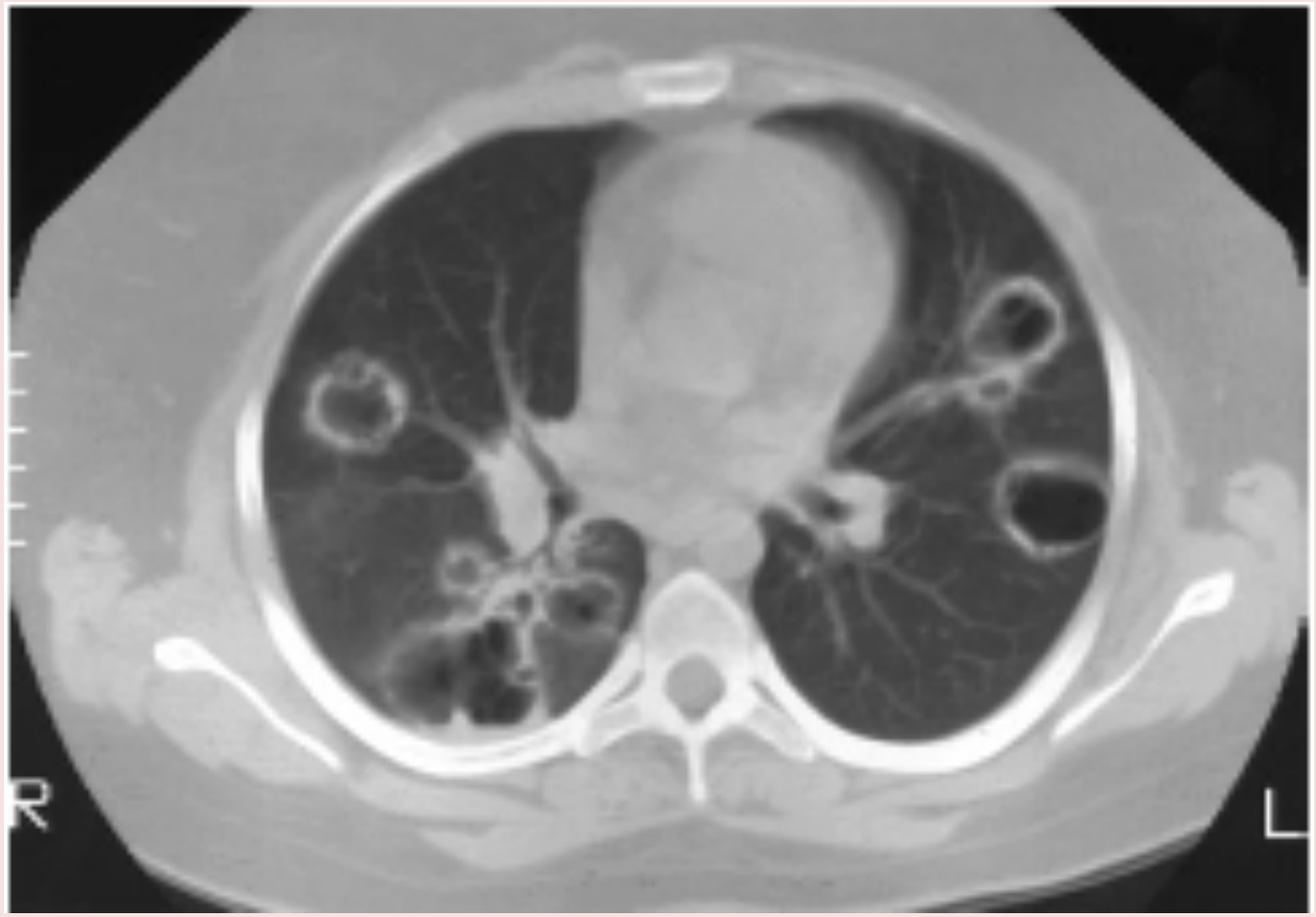
A. Initiate treatment with indomethacin, 75 mg twice daily.
B. Initiate treatment with prednisone, 60 mg daily.
C. Initiate treatment with unfractionated heparin adjusted based on activated partial thromboplastintime to obtain full anticoagulation.
D. Perform magnetic resonance angiography of the brain.
E. Perform a temporal artery biopsy.

# The answer is B

- This pt is presenting with amaurosis fugax with evidence of decreased visual acuity and anterior ischemic optic neuropathy in the setting of a compatible clinical hx of GCA (temporal arteritis).
- In an individual >50 y, this clinical hx should ***prompt immediate initiation of glucocorticoids*** in order to prevent the development of monocular blindness.
- GCA is ***exquisitely sensitive to steroid therapy***, and initiation of prednisone, 40–60 mg daily, is usually effective at managing the sx. If ocular symptoms recur, prednisone may be increased further. Once symptoms are controlled, gradual tapering of the steroid dose should occur. Most patients do require prolonged courses of steroid, usually for >2 years. ***The elevation in ESR can be a useful marker of disease activity during a steroid taper.***
- ***Aspirin is often used in combination with glucocorticoids as it has been shown to decrease ischemic complications.***
- Indomethacin is not frequently used and should not be used alone in a pt presenting with symptoms of ischemic optic neuropathy.
- ***There is no role for anticoagulation.***
- Definitive dx is confirmed by temporal artery bx.
- ***Treatment should not be withheld for performance of the biopsy as sudden and irreversible blindness may occur.***

# Case 4

- A 34 y/o man is admitted for evaluation & Rx of RF & an abnormal CT of the chest. For the past 2 months, he has had fatigue, malaise, and intermittent fevers to as high as 38.2°C. About 3 weeks ago, he sought treatment from his primary provider for sinus pain and congestion with a purulent and bloody nasal discharge.
- He was treated for 2 weeks with ampicillin- sulbactam, but his symptoms have only minimally improved. When he returned to his physician, a basic metabolic panel was performed which showed a creatinine of 2.8 mg/dL. A urinalysis showed 1+ protein with 25 red blood cells per high-power field. RBC casts were present. His chest CT is shown below.
- ***Which of the following tests would be most likely to be positive in this individual?***


R
L

A. Antiglomerular basement membrane antibodies
B. Antiproteinase-3 antibodies
C. High titers of antibodies to antistreptolysin O
D. Perinuclear antineutrophil cytoplasmic antibodies
E. Positive blood cultures for Staphylococcus aureus

# The answer is B

- The presenting Sx of this pt include rapidly progressive renal failure, sinusitis, and cavitary lung disease. Consistent with (WG). WG is characterized by granulomatous vasculitis of small vessels that primarily manifests in the airways and kidneys.
- The male-to-female ratio is equal. The upper airway is involved in 95% of patients and, in this setting, the disease often presents as chronic sinusitis unresponsive to antibiotic therapy. Facial pain and bloody nasal discharge are commonly present.
- Untreated disease can progress to complete cartilaginous destruction with nasal septal perforation and saddle- nose deformity.
- ***The lungs are the second most commonly affected organ in about 85% of individuals with WG.*** The spectrum of lung disease may vary widely from asymptomatic pulmonary infiltrates to massive hemoptysis

- In this pt, there are characteristic cavitary lung lesions that help to differentiate WG from microscopic polyangiitis, as no cavitary disease is seen in microscopic polyangiitis.
- ***Rapidly progressive glomerulonephritis is present in 77% of patients and is responsible for the majority of deaths in WG***.
- Non specific sx are also present when the disease is active including fatigue, weight loss, and fevers.
- Dx of WG is made by demonstration of necrotizing vasculitis on tissue biopsy of an affected organ.

- Serologic testing can offer supporting evidence for the Dx of WG.
- ***90 % of individuals with WG will demonstrate (c-ANCA).***
- ***The specific c-ANCA target in WG is proteinase-3***.
- Rapid initiation of therapy is important. Prior to the use of cyclophosphamide, WG was almost universally fatal within 5 years, even with the use of glucocorticoids.
- With the combined use of glucocorticoids and cyclophosphamide, survival is now 75–80% at 5 years.
- ***(p-ANCA) are usually directed against myeloperoxidase***.
- The p-ANCA are seen in a minority of pts with WG but are more commonly present in microscopic polyangiitis.

# Case 5

- A 29 y/o male with episodic abdominal pain and stress-induced edema of the lips, the tongue, and occasionally the larynx is likely to have ***low functional or absolute levels of which of the following proteins***?

A. C5A (complement cascade)

B. IgE

C. T cell receptor, α chain

D. Cyclooxygenase

E. C1 esterase inhibitor

# The answer is E

- Complement activity, which results from the sequential interaction of a large number of plasma and cell-membrane proteins, plays an important role in the inflammatory response. The classic pathway of complement activation is initiated by an antibody-antigen interaction.

- Patients with a ***deficiency of C1 esterase inhibitor*** may develop **angioedema**, sometimes leading to death by asphyxia.
- Attacks may be precipitated by ***stress or trauma***. In addition to low antigenic or functional levels of C1 esterase inhibitor, patients with this autosomal dominant condition may have normal levels of C1 and C3 but low levels of C4 and C2.
- ***Danazol therapy*** produces a striking increase in the level of this important inhibitor and alleviates the symptoms in many patients.
- An ***acquired*** form of angioedema caused by a deficiency of C1 esterase inhibitor has been described in patients with ***autoimmune or malignant disease***.

# Case 6

- A patient with primary Sjögren's syndrome that was diagnosed 6 years ago and treated with tear replacement for symptomatic relief notes continued parotid swelling for the last 3 months.
- She has also noted enlarging posterior cervical lymph nodes. Evaluation shows leukopenia and low C4 complement levels. ***What is the most likely diagnosis?***

A. Chronic pancreatitis

B. Secondary Sjögren's syndrome

C. HIV infection

D. Lymphoma

E. Amyloidosis

# Answer is D

- Lymphoma is well known to develop specifically in the late stage of Sjögren's syndrome.
- Common manifestations of this malignant condition include persistent parotid gland enlargement, purpura, leukopenia, cryoglobulinemia, and low C4 complement levels.
- ***Most of the lymphomas are extranodal***, marginal zone B cell, and low-grade. Low-grade lymphomas may be detected incidentally during a labial biopsy.
- ***Mortality is higher in pts with concurrent B symptoms*** (fevers, night sweats, and weight loss), a lymph node mass ***>7 cm***, and a ***high or intermediate histologic grade***.

# Question 7

- All the following organisms have been implicated in reactive arthritis except:

A. Chlamydia trachomatis

B. Neisseria gonorrhoeae

C. Salmonella enteritidis

D. Shigella dysenteriae

E. Yersinia enterocolitica.

# The answer is B

- The most common organisms that are implicated are bacteria that cause acute infectious diarrhea.
- ***All four Shigella species*** have been reported to cause reactive arthritis, although S. flexneri is only rarely implicated.
- Other bacteria that have been identified as triggers include several ***Salmonella species, Yersinia enterocolitica, and Campylobacter jejuni***.
- Some organisms that cause urethritis are also causative; these include ***Chlamydia trachomatis and Ureaplasma urealyticum***.
- Arthritis associated with disseminated gonococcal infection is directly related to an infectious cause and responds to antibiotics, unlike reactive arthritis.

# Question 8

- What is the most common extraarticular manifestation of ankylosing spondylitis?

A. Anterior uveitis

B. Aortic regurgitation

C. Cataracts

D. Inflammatory bowel disease

E. Third-degree heart block

# The answer is A

- Anterior uveitis occurs in up to 30% of AS pts and may antedate the onset of the spondylitis.
- Attacks usually occur ***unilaterally*** with pain, photophobia, and blurred vision. ***Recurrent attacks are common***, and ultimately cataracts may result.
- Other commonly seen problems include inflammation in the colon and ileum in up to 60% of AS pts, but only ***rarely do these patients develop IBD.***
- Cardiac disease is present in only a few percent of these pts and ***most commonly presents as AR***. Other cardiac manifestations include CHB and CHF.
- Rare complications are ***upper lobe pulmonary fibrosis and retroperitoneal fibrosis.***

# Case 9

- A 64 y/o man with CAD and AF is referred for evaluation of fevers, arthralgias, pleuritis, and malar rash. The sx have developed over the past 6 months. The pleuritis has responded to steroid Rx, but prednisone has been unable to be tapered off due to recurrence of symptoms at daily steroid doses <15 mg of prednisone. His medications include aspirin, procainamide, lovastatin, predni- sone, and carvedilol. ***Antibodies directed against which of the following proteins is most likely to be positive?***
- A. Cardiolipin
- B.Double-strand DNA
- C. Histone
- D.Ribonucleoprotein (RNP)
- E.Ribosomal P

# The answer is C

- Drug-induced lupus can occur with a variety of medications and should be considered when individuals present atypically.
- Individuals with drug-induced lupus are more likely to be ***male*** and of Caucasian race.
- Drug-induced lupus usually presents with fever, malaise, intense arthralgias/ myalgias, serositis, and rash.
- The ***brain and kidneys are rarely*** involved.
- ***Discontinuation of the medication usually leads to resolution of the symptoms*** over a period of weeks, although anti-inflammatory medications may need to be utilized to control symptoms until the inflammation subsides.
- Common drugs that cause lupus include ***procainamide, propafenone, hydralazine, propylthiouracil, lithium, phenytoin, carbamazepine, sulfasalazine, and minocycline. Beta blockers, angiotensin-converting enzyme inhibitors, lovastatin, and simvastatin*** have also been reported to cause drug-induced lupus.
- Antibody testing usually reveals a positive ANA and antihistone antibodies.
- ***Anti-RNP antibodies are seen with mixed connective tissue disease*** that usually presents with features of lupus, rheumatoid arthritis, and/or scleroderma.
- ***Anti-ribosomal P antibodies*** are associated with depression and psychosis with central nervous system involvement of SLE.

# Case 10

- A 65 y/o woman with scleroderma presents with progressive SOB. CXR shows interstitial changes. Dry bibasilar rales are heard O/E. Diffusing capacity is reduced. High-resolution CT shows diffuse bibasilar ground-glass appearance. In addition to prednisone, ***which of the following is the best treatment for her lung involvement?***

a. Azathioprine

b. Cyclophosphamide

c. Methotrexate

d. Mycophenolate mofetil

e. Plasmapharesis

# Answer b

- Ground-glass appearance on chest CT correlates with active alveolitis. This finding can be responsive to cyclophosphamide treatment.
- A honeycomb appearance on CT is more indicative of fibrosis, which is not responsive to medication.