CASE THREE

Short case number: 3_26_3
Category: Musculoskeletal System & Skin
Discipline: Medicine
Setting: General Practice
Topic: Systemic Vasculitis_Polymyalgia Rheumatica.

Case

Joan Hayes, is an active, normally well, 70 year old, she presents complaining that she thinks old age is catching up with her. *"Recently my muscles seems to be aching all the time and sometimes I can hardly get out of my chair"*
She explains that she is also having trouble sleeping because of the pain.

You notice that Joan does seem to be moving more slowly than she usually does and that she is in pain.

Questions

1. In your assessment of Joan what key features of history and examination would support a diagnosis of polymyalgia rheumatica? What other conditions would you consider in your differential diagnosis?
2. What investigations would you undertake and what findings would support a diagnosis of polymyalgia rheumatica?
3. A few days after your initial assessment of Joan and while you are awaiting the results of your investigations, she presents complaining of increasing fatigue and headaches, she states *"it even hurts to touch my head."* You are concerned that Joan temporal arteritis. What features of history and examination would support this diagnosis? What would be your immediate concern and why?
4. You explain to Joan that you are concerned she has temporal arteritis and you need to commence her on immediate treatment. Joan asks you what this is and what treatment is required. What would you explain to Joan? Outline management of temporal arteritis
5. Joan is referred for a temporal artery biopsy, what histopathological features would you expect to see in the biopsy?

Suggested reading:

- Kumar P, Clark ML, editors. Kumar & Clark's Clinical Medicine. 8th edition. Edinburgh: Saunders Elsevier; 2012.
- Colledge NR, Walker BR, Ralston SH, Penman ID, editors. Davidson's Principles and Practice of Medicine. 22nd edition. Edinburgh: Churchill Livingstone; 2014.

ANSWERS

1. In your assessment of Joan what key features of history and examination would support a diagnosis of polymyalgia rheumatica? What other conditions would you consider in your differential diagnosis?

   Polymyalgia rheumatica (PMR) is a relatively common clinical syndrome of unknown aetiology.
   It is characterized by proximal myalgia of the hip and shoulder girdles with accompanying morning stiffness that lasts for more than 1 hour. Typically, this disorder affects people older than 50 years of age who were previously in good health. People may present with both systemic and musculoskeletal problems. Systemic problems include low-grade fever and weight loss, malaise, fatigue, and depression, difficulty rising from bed in the morning, difficulty getting up from the toilet and difficulty completing daily life activities.
   Musculoskeletal problems include morning stiffness or stiffness after prolonged inactivity. Carpel tunnel syndrome may accompany this disorder in about 15% of people.
   Differential diagnosis includes:
   - Depression
   - Other types of vasculitis including Giant Cell Arteritis,
   - Other arthropathies
   - Hypothyroidism
   - Polymyositis.

2. What investigations would you undertake and what findings would support a diagnosis of polymyalgia rheumatica?

   Erythrocyte sedimentation rate is the most sensitive study for polymyalgia rheumatica, but it is not specific. The ESR is frequently elevated and greater than 40 mm/h, but the rate can exceed 100 mm/h.
   C-reactive protein is also often elevated and may parallel the ESR.
   Longitudinal studies suggest that it may be a more sensitive test than ESR for the diagnosis.
   The full blood count may be normal but it may also show a mild normocytic, normochromic anaemia with an elevated white cell and platelet count.
   The creatine kinase level is normal and this helps differentiate PMR from other disorders such as polymyositis.
   Serum IL-6 levels are elevated and often closely parallel inflammatory activity of the disease.

3. A few days after your initial assessment of Joan and while you are awaiting the results of your investigations, she presents complaining of increasing fatigue and headaches, she states *"it even hurts to touch my head."* You are concerned that Joan temporal arteritis. What features of history and examination would support this diagnosis? What would be your immediate concern and why?

   People with temporal arteritis present with headache and a tender, thickened, or nodular artery. This may be pulsatile early in the disease but become occluded later.
   If left untreated, this may result in ischemic optic neuropathy, which may lead to serious visual symptoms and blindness.

4. You explain to Joan that you are concerned she has temporal arteritis and you need to commence her on immediate treatment. Joan asks you what this is and what treatment is required. What would you explain to Joan and outline management of temporal arteritis?

   The goals of treatment in giant cell arteritis are to reduce symptoms and, most importantly, to prevent visual loss. Giant cell arteritis and its associated symptoms are very sensitive to glucocorticoid therapy. Treatment should begin immediately with prednisone 40-60 mg/ day for at least one month, though the optimal duration of treatment is not known. Giant cell arteritis also responds to tocilizumab (Actemra), a

monoclonal antibody that blocks the interleukin-6 receptor (IL-6R). The availability of this therapy is an important development in the treatment of GCA because it can treat patients with a poor response to corticosteroids and can be used to significantly reduce the dose of corticosteroids, thus reducing the significant adverse consequences associated with long term and/or high dose steroids.

5. Joan is referred for a temporal artery biopsy, what histopathological features would you expect to see in the biopsy?

   Temporal artery biopsy is the gold standard to establish the diagnosis of temporal arteritis. Features of the biopsy include inflammation of the arterial wall with fragmentation and disruption of the internal elastic lamina. Often multinucleated giant cells with a dense perivascular inflammatory infiltrate are visible.