Patient with Daily Headache

R. Allan Purdy, MD, FRCP, FACP
Professor of Medicine (Neurology)
Learning Issues

• Headaches in the elderly always require a look for a secondary causation
• The history is important as on examination there few clinical signs
• Systemic symptoms have to be sought out to make the diagnosis
• Failure to diagnose this condition has serious consequences
Case History

- JS is a 72-year-old man with a three-month history of diffuse headache, somewhat more left sided and frontal temporal in location. The headache is moderately severe and boring in nature and without relieving or aggravating factors.
Case History

• Over time the headache is gradually worsening and he has begun to feel ill. He develops pain in his face, jaw and tongue.

• He noticed increasing tiredness and some weight loss. He has history of vascular disease.

• In the past few weeks he has noticed recurrent episodes of transient loss of vision in his left eye, coming down like a curtain.
Physical Examination

• On examination he looks somewhat ill and in some distress from the pain.
• His blood pressure is 140/90 and PR is 68.
• He has no focal neurological signs and his general examination appears normal.
• His fundi showed some atherosclerotic and hypertensive arteriole changes.
• There are no bruits in his head or neck.
Initial Impression

- JS has a steady headache which is not specific for any primary or secondary headache disorder and one which could be a chronic daily headache since it is present more than 15 days a month and longer than 4hrs in duration, however it has yet to last six months which is important in the differential diagnosis.
Other Historical Facts

• JS did not report any triggering or relieving factors. This is very important since most primary headaches, including migraine, tension-type headache and cluster headache, have definite modifying factors.

• As this is associated with a progressive worsening over time, it is a definite ‘red flag’ with respect to looking for a secondary cause.

• He has also had his gall bladder removed.
First Stage Questions

• At this stage can you consider three differential diagnoses for this case?
• Is there any other information you would like to know about the case?
• What initial investigations, if any, would you like to consider?
More Analysis

• He feels unwell, has fatigue and weight loss, symptoms on their own which could suggest a systemic disease. On their own one might consider something like a malignant process which can present with headaches as well, however one should consider the differential diagnosis more broadly in any patient with constitutional symptoms.
More Analysis

• Also patients with primary headache can have anorexia (tension-type headache) and nausea and vomiting (migraine) but again these are episodic and his age starts to play a role in the diagnostic formulation in his case.
Symptom Analysis

• JS has volunteered symptoms of pain in the face, jaw and tongue, but the latter two are worse with eating and talking, so these need to be evaluated carefully. These symptoms suggest some need for activity to bring them on and are termed, jaw and tongue claudication respectively.
Clinical Pearl

- Claudication suggests the blood supply to the pertinent muscles is compromised to ischemic levels during eating and chewing so that the subsequent pain is most likely ischemic in nature.
Symptom Analysis

• The visual symptoms here are classic for ‘amaurosis fugax’ which implies reduced blood supply to the central retinal artery.
• The classic loss of vision described as a blind coming down is based on the anatomical blood supply to the retina, that is the lower vessels become involved first and then upper arterioles.
Any need to think Otherwise?

- There are no other important features in his history. He has had some mild hypertension over time and prior gall bladder removal, but nothing that suggests any other systemic disorder.
Lab and Investigations

- Preliminary blood work including a complete blood count, CRP and ESR were ordered along with and EKG, chest x-ray and a baseline unenhanced CT scan.
- He also had electrolytes, renal function and lipids done.
Secondary Impression

• A secondary headache in an older patient is likely. Transient ischemic attack or TIA, might be important, because of the visual symptoms, so carotid artery disease needs consideration.

• However the headache is daily which is unusual in a TIA and is progressive so another vascular diagnosis should be kept in mind.
Further impression

• A less likely concern would be cardiac emboli from non-bacterial endocarditis associated with an underlying malignancy, which might fit with some of the general and systemic symptoms.
Second Stage Questions

• At this stage can you narrow your diagnosis to one entity?
• Can you analyze the case further based on what you know about this diagnosis?
• Any other investigations you would do and how would they be justified?
• Any immediate or long term management suggestions in this case?
Diagnosis

• The visual symptoms in this case along with jaw and tongue claudication, are very characteristic of temporal arteritis, as well as pain in the shoulders which is termed *polymyalgia rheumatica*, a feature missing in his early presentation but present later.

• One physical finding helped in the diagnosis, seen on next slide!
Examination of Temple
Diagnostic Points

• Systemic symptoms of being and “looking” ill are classical but not specific.
• Also remember that “temporal arteritis” is a systemic disorder and a misnomer since other major arteries can be involved including the carotid, basilar and coronary arteries, which could cause a major stroke or myocardial infarction.
The Headache of TA

- The headache in temporal arteritis is not specific but does flag raise the diagnosis early in the case of any elderly patient.
- The pain is usually located over the temporal artery region and is the commonest presenting symptom of the disorder.
Another Clinical Pearl

• Anterior ischemic optic neuropathy or AION can occur in patients with temporal arteritis and can lead to permanent blindness if not recognized and treated and even if treated.

• The diagnosis in his case was supported by the clinical observation of a swollen, tortuous and painful left temporal artery (Figure 1).
Management Issues

• Obtaining results of blood work.
• Treatment with corticosteroids.
• CT baseline scan to look for other diagnoses in this case.
• Carotid duplex dopplers in this case to rule out carotid disease with TIA.
• Temporal Artery Biopsy
Lab Results

• The results of the blood work can be very informative and strongly support the diagnosis in this case.
  – The CBC showed a microcytic anemia, the ESR was 125, with a CRP of 42.
  – His serum chemistry including was normal.
  – His EKG was essentially normal.
Comments on Steroids

- Treatment should be started as early as possible to prevent major complications of the arteritis, especially loss of vision, usually due to an AION.

- The treatment is oral steroids with the dosage range of prednisone being 40 mg/day to 80 mg/day, although 60 mg/day or above has been more commonly suggested than lower doses.
Justification for CT

- A CT scan is reasonable in his case as he could have a systemic malignancy with secondary brain metastases or small infarctions from prior carotid TIAs or cardiac microemboli.
- In his case this test was normal for age and that is the usual finding in this disorder.
Reason for Carotid Dopplers

• Again this test is reasonable since he presented with amaurosis which usually is due to artery to artery embolic from the internal carotid artery to the ophthalmic artery and retinal vessels.
• In his case the test was normal.
The Temporal Artery Biopsy

• The temporal artery biopsy is an important test to do as soon as reasonable. A good section of artery is required to avoid skip lesions.

• It should be done by a surgeon who has a lot of experience obtaining such specimens and read by a pathologist familiar with the disorder.

• Delay of therapy however is not warranted while waiting for the biopsy.
Options on the biopsy result

- If biopsy positive treatment continues and if negative or equivocal, the treatment should be guided by clinical impressions and/or other laboratory tests, especially the ESR.
- See Figure 2 for a biopsy which shows typical arteritis as seen in this classic temporal arteritis.
Temporal artery biopsy
Long-term Followup

• Patients with temporal arteritis can be managed by most practitioners, but referral to a neurologist, internist or rheumatologist maybe helpful for diagnosis, and ongoing management.

• Surgery and pathology are involved early as well as an ophthalmologic opinion can be helpful for assessment and follow up of the eye symptoms.
Treatment Follow-up

• The duration of therapy depends on his clinical course and follow-up assessments.

• Most patients require high dose steroids for several weeks and this may last up to a year or longer.

  – Steroid sparing or other immunosuppressive agents sometimes are employed in some cases. Long term problems with steroids are to be avoided and must be managed.
Back to the Case!

- Shortly after receiving prednisone orally, his symptoms started to subside within hours. The next day he was headache free on 60 mgs per day.
- He continued on this dosage for a month and then it was gradually tapered and discontinued several months later as he became asymptomatic in all respects and his blood work normalized.
Final Diagnosis

- This is a case of *temporal arteritis* presenting with a visual symptom of amaurosis fugax and not AION.
- The case demonstrates the usual features of the disorder as well as concepts about immediate and long term management.
- The headache of this disorder is not specific and on its own non-diagnostic!
References


• Goodwin, J. Temporal Arteritis. Medlink http://www.medlink.com