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A case of vision loss in a patient with Giant Cell Arteritis

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A missed case of Giant Cell Arteritis – Awareness for the PCP

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Abstract

Introduction: Giant Cell Arteritis (GCA) is the most common large vessel vasculitis and carries the potential for life-changing morbidity manifested as permanent vision loss or other neurological deficit. Despite these well understood complications, literature suggests that a significant delay to diagnosis in GCA exists. Owing to the gradual and non-specific symptoms associated with GCA, patients are likely to present in the ambulatory setting. This case report aims to provide an example of a delay to diagnosis in GCA and improve awareness of the diagnostic challenges facing primary healthcare providers.

Case Description: An 80 year old Caucasian female presented to her PCP with a 1 week history of aching left sided neck pain with radiation to posterior scalp. She denied any associated symptoms. Medical history included Coronary Artery Disease and Right Supraspinatus Tear. Neck pain was initially diagnosed as cervical spondylosis and the patient was discharged with physical therapy referral. Over the following weeks the patient presented twice to her PCP and once to the Emergency Department (ED) with symptoms including scalp pain, fatigue, persistent headache, pain in her mouth and weight loss. Work-up during this time included routine labs and MRI Brain and C-spine which did not reveal a diagnosis. 8 weeks following the onset of symptoms the patient presented to the ED with partial vision loss in her right eye. ESR was found to be elevated at 47mm/hr. She was started on Prednisone 60mg PO daily and underwent Temporal Artery Biopsy (TAB) which confirmed the diagnosis of Giant Cell Arteritis. Despite oral steroids she developed vision loss in the contralateral eye and was admitted to hospital to receive IV corticosteroids. She was subsequently started on Tocilizumab (Actemra) with tapering of corticosteroids. 4 months following her initial vision loss she reported no improvement in vision despite adherence to therapy. 6 months after the diagnosis of GCA she was admitted for severe sepsis and lower GI bleeding secondary to terminal ileitis. She had poor functional recovery following discharge from hospital and died under hospice care 7 months later.

Discussion: A recent meta-analysis estimates the mean time from presentation to diagnosis of GCA at 9 weeks. This case demonstrates a similar pattern and highlights the challenge and importance of achieving a prompt diagnosis. Existing literature suggests that few of the classic symptoms and physical signs are predictive of biopsy-confirmed GCA. Furthermore, none of these history and physical points are reliable to rule out GCA. However, Erythrocyte Sedimentation Rate (ESR) has demonstrated utility in ruling out GCA with reported sensitivity ranging from 85-96% for any ESR elevation and Negative Likelihood Ratio of 0.2 for normal ESR. The American College of Rheumatology published Classification Criteria for GCA in 1990, however comprehensive management guidelines are still under development. Existing European and British guidelines endorse maintaining a high-degree of clinical suspicion to prompt urgent referral for TAB and specialist evaluation. To improve clinical outcomes through rapid diagnosis, studies of fast-track referral pathways have demonstrated reduced rates of permanent-vision loss compared to conventional care. Nonetheless, increased primary care awareness is necessary for consideration of this less common but potentially catastrophic diagnosis.

Background

WHY THIS CASE?

- Giant Cell Arteritis (GCA) is the most common large vessel vasculitis (1) and carries the risk of permanent vision loss which occurs in 15-20% of patients. (2)
- GCA often presents subtly and with non-specific symptoms leading to frequent delays to confirming this challenging diagnosis. (3)
- This delay is of particular importance as prompt initiation of Corticosteroid therapy has been demonstrated to significantly decrease the incidence of vision loss (4).
- Fortunately, existing literature has evaluated the diagnostic significance of history, physical examination and laboratory findings to offer an evidence basis to guide clinical suspicion.

GCA BASICS

- GCA is a systemic vasculitis with a predilection to supra-aortic arteries, particularly the branches of the external carotid artery. (5)
- It is seen overwhelmingly in individuals over 50 years of age and incidence increases with age. (5) The highest incidence of GCA is seen in persons of Northern European ancestry. (6)
- The clinical presentation of GCA may include cranial symptoms including headache, temporal or scalp tenderness and jaw or tongue claudication. Extra-cranial manifestations may include unexplained fever, malaise, weight loss and fatigue. (7)
- Vision loss in GCA is caused by anterior ischemic optic neuropathy (5) with images from our case patient seen to the right (Figure 1).

Case Description

- An 80 year old Caucasian female with a history of Coronary Artery Disease, Hypertension and Right Supraspinatus Tear presented to her PCP office with a 1 week history of aching left sided neck pain with radiation to posterior scalp. She denied any associated symptoms. She was ambulatory and functioning independently.
- Neck pain was initially diagnosed as cervical spondylosis, the patient received osteopathic manipulation and was discharged with a physical therapy referral.
- Two weeks later she was seen by a different provider in the same practice and endorsed persistence of neck pain with development of bilateral headache and fatigue. MRI Brain and Cervical Spine were ordered and patient received prescriptions for Norco and Valium. Imaging results revealed mild cervical spondylosis with no intracranial pathology.
- Three weeks later she was seen again by a third provider for self-limited cough and shortness of breath. She had persistence of fatigue for which labs including routine chemistry, complete blood count, HbA1c, TSH, Cortisol and Iron Studies were ordered. Patient reported resolution of her previous headache for which a diagnosis of Migraine Headache was documented.
- 8 weeks following the onset of symptoms the patient presented to the ED with partial vision loss in her right eye. ESR was found to be elevated at 47mm/hr. She was started on Prednisone 60mg PO daily and underwent Temporal Artery Biopsy (TAB) which confirmed the diagnosis of Giant Cell Arteritis.
- Despite oral steroids she developed vision loss in the contralateral eye and was admitted to the hospital to receive IV corticosteroids. She was subsequently started on Tocilizumab (Actemra) with tapering of corticosteroids.
- 4 months following her initial vision loss she reported no improvement in vision despite adherence to therapy. 6 months after the diagnosis of GCA she was admitted for severe sepsis and lower GI bleeding secondary to terminal ileitis. She had poor functional recovery following discharge and died under hospice care 7 months later.

Photos

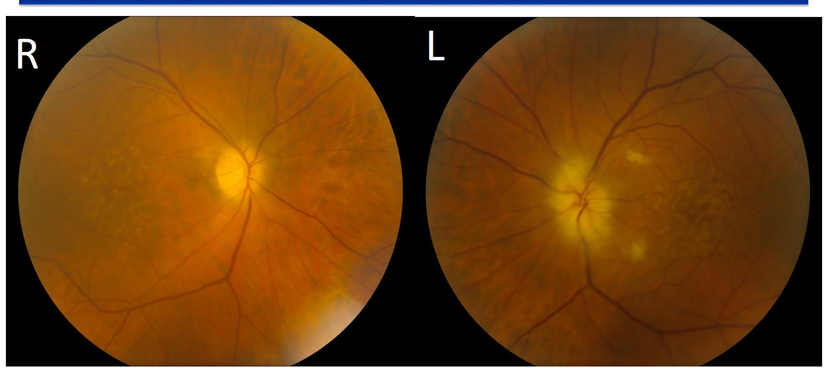

Figure 1. Fundoscopy images demonstrating marked optic disc edema of left eye secondary to Ischemic Optic Neuropathy.

Discussion

- A recent meta-analysis estimates the mean time from presentation to diagnosis of GCA at 9 weeks.(3) This case demonstrates a similar pattern and highlights the challenge and importance of achieving a prompt diagnosis.
- Existing literature suggests that few of the classic symptoms and physical signs are predictive of biopsy-confirmed GCA.(7)
  - **Jaw Claudication: Positive Likelihood Ratio (LR) 4.2**
  - **Diplopia: Positive LR 3.4**
  - **Beaded Temporal Artery: Positive LR 4.6**
  - **Prominent or Enlarged Temporal Artery: Positive LR 4.3**
- Furthermore, none of the classic history and physical findings are reliable to rule out GCA.
- **Erythrocyte Sedimentation Rate (ESR**) has demonstrated some utility in ruling out GCA with reported sensitivity ranging from 85-96% for any ESR elevation and Negative Likelihood Ratio of 0.2 for normal ESR (7,8). With a positive result threshold of >50mm/h the negative LR based on these studies is 0.35. Despite our patient's presentation with vision loss, her ESR was less than this threshold.
- In our patient's clinical course there were epidemiologic and symptomatic clues to raise suspicion for GCA. Identification may have been hampered by lack of continuity with a single provider.
- The American College of Rheumatology published Classification Criteria for GCA in 1990, however comprehensive management guidelines are still under development. Existing European and British guidelines endorse maintaining a high-degree of clinical suspicion to prompt urgent referral for TAB and specialist evaluation. (9,10).
- To improve clinical outcomes through rapid diagnosis, studies of fast-track referral pathways have demonstrated reduced rates of permanent-vision loss compared to conventional care (11). Nonetheless, increased primary care awareness is necessary for consideration of this less common but potentially catastrophic diagnosis.

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