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Case Report

Atypical presentation of giant cell arteritis: a case report

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ABSTRACT

A 58-year-old female, known hypertensive and hypothyroid, presented with complaints of diffuse headache, decreased appetite, and easy fatiguability over 2 months. Laboratory tests revealed elevated acute phase reactants (ESR-140 mm/1st hour, CRP-70 mg/l), positive ANA profile with anti-Pm-SCL 100 and AMA-M2 positivity. During subsequent visits, she had a recurrence of headaches along with dyspnea on exertion (DOE) being her predominant symptom limiting her daily activities. A superficial temporal artery doppler was done which showed a positive 'halo sign' and right temporal artery biopsy was compatible with the diagnosis of giant cell arteritis (GCA).

Keywords: Giant cell arteritis, Dyspnea on exertion, Biopsy

INTRODUCTION

Giant cell arteritis (GCA) is a chronic autoimmune vasculitis. It is characterized by monocyte-derived giant cell infiltration of large and medium arteries. GCA results in local and systemic inflammation.[1] It is defined as a pan-arteritis that predominantly involves the extracranial branches of the carotid artery. Temporal headache, jaw claudication, fever and visual disturbances are the classical symptoms.[2] We are herewith presenting an interesting and atypical presentation of GCA.

CASE REPORT

A 58-year-old female, known hypertensive and hypothyroid, presented to the OPD with complaints of diffuse headache, decreased appetite, and easy fatiguability over 2 months. She also complained of dyspnea on exertion without cough, dependent edema, and chest pain during this period. She denied history of fever, weight loss, visual disturbances, limb/jaw claudication, joint pains, and shoulder/pelvic girdle pain. There is no history of tightening of skin, darkening of the skin, Raynaud's phenomenon, muscle weakness, and orthopnea. On examination, patient was afebrile with normal general physical findings. All peripheral pulses were felt well and had no significant difference in blood pressure amongst the limbs. Temporal arteries were non-tender and had no nodularity on examination. Musculoskeletal and systemic examination also did not reveal any significant findings.

Preliminary evaluation revealed a normal complete blood picture, renal and liver function tests, and raised acute phase reactants (ESR-140 mm/1st hour, CRP-70 mg/l). ANA profile (immunoblot) was done which showed anti Pm-SCL 100 and AMA-M2 mild positivity. Imaging revealed normal study on MRI brain and USG abdomen.

The patient complained of persisting exertional breathlessness and reported significant ocular and oral SICCA symptoms during the next OPD visit 2 weeks later. The headache had resolved by this time. Ophthalmology evaluation revealed evaporative dryness of eyes and hypertensive retinal changes only. Chest CT showed minimal pericardial effusion with no features of interstitial lung disease. Echocardiography was normal without evidence of pulmonary hypertension. Further investigations showed normal serum protein electrophoresis, and serum angiotensin-converting

enzyme levels, with mildly elevated urine creatine ratio. The possibility of undifferentiated connective tissue disease was considered and the patient was put on hydroxychloroquine and other symptomatic treatment.

On review after 3 weeks, patient complained of recurrence of headache and DOE which had progressed to the extent of limiting her daily activities. Her oxygen saturation and respiratory rate were normal. Acute phase reactants (ESR-96 mm/1st hour and CRP-65 mg/l) persisted to be elevated. Possibility of GCA was considered. Superficial temporal artery Doppler done thereafter showed wall thickening with hypoechoic halo and luminal narrowing in bilateral superficial temporal arteries suggestive of temporal arteritis. A right temporal artery biopsy done under local anesthesia was compatible with the diagnosis of GCA.

Treatment

The patient was treated with leflunomide (10 mg OD) and oral steroids (Deflazacort 1 mg/kg and tapered).

Follow up

On follow-up after 1 month, the patient had resolution of headache and exertional breathlessness with a reduction in acute phase reactants (ESR-79 mm/1st hour, CRP-9.8 mg/l). After 3 months ESR reduced to 57 mm/1st hour and CRP-8.9 mg/l. She was on leflunomide 10 mg OD and a tapering dose of steroids (deflazacort 6 mg OD) during this visit. On the last follow-up after 12 months, patient was asymptomatic with near normal values of acute phase reactants (ESR-56 mm/1st hour and CRP-4.9 mg/l).

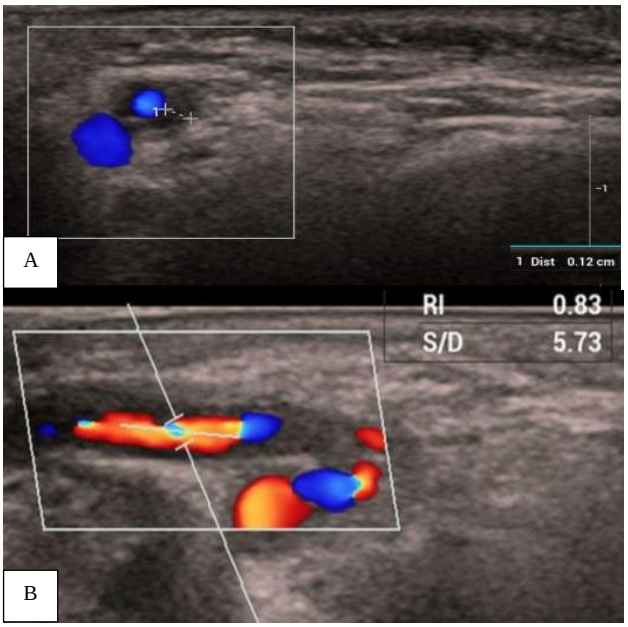

Figure 1 (A and B): Superficial temporal artery doppler-wall thickening with hypoechoic halo (halo sign) and luminal narrowing in bilateral superficial temporal arteries.

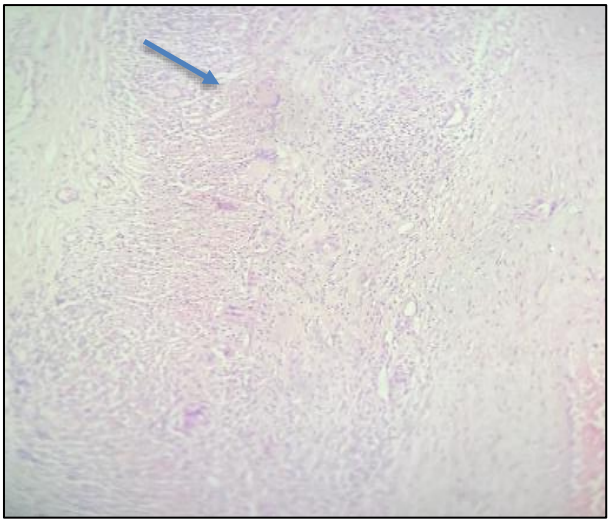

Figure 2: Temporal artery biopsy transmural inflammation with multinucleate giant cells.

DISCUSSION

GCA is the vasculitis of large and medium-sized vessels which present typically with classical symptoms like temporal headache, jaw claudication, fever, and visual disturbances. Respiratory symptoms in GCA are rare. Studies show that large vessel vasculitis can involve the aorta and its branches which may lead to subclinical or overt pulmonary complications. Hunder et al detail pulmonary involvement in GCA which presents as dyspnea due to inflammatory changes in the large vessel.[1] The pathogenesis of GCA includes an aberrant immune response characterized by macrophage infiltration and T-cells into the arterial wall. Here, pro-inflammatory cytokines such as tumor necrosis factor (TNF-α) and interleukin (IL)-6 play a major role in the inflammatory cascade. This type of immune dysregulation can extend to large vessels which may present as systemic symptoms like dyspnea and fatigue.[3]

One of the key diagnostic markers of GCA is the 'halo sign' on doppler study. It is a hypoechoic ring around the artery wall. Its specificity and sensitivity are above 90%.[4] But the gold standard remains temporal artery biopsy. It confirms the findings of granulomatous inflammation and multinucleated giant cells.[5] The main treatment of GCA is high-dose corticosteroids which can rapidly alleviate symptoms and reduce complications such as aneurysm formation and vision loss.[6] As per EULAR 2018 guidelines, an initial dose of 40-60 mg/day of prednisolone should be given. It should be tapered gradually over months depending upon normalization of inflammatory markers as well as clinical response.[7]

Various studies have documented respiratory involvement in GCA, although it is in a limited number of cases. Gonzalez-Gay et al reported dyspnea in a group of patients with GCA, leading to subclinical inflammation of the thoracic aorta.[8] Also Sonnenblick et al highlighted

cases with interstitial lung changes in biopsy-proven GCA, reinforcing the possibility of GCA's pulmonary involvement.[9] In addition to these studies, Weyand and Goronzy also emphasized that systemic inflammatory pathways implicated in GCA could extend beyond cranial arteries, affecting major vessels and leading to pulmonary symptoms.[10] Although rare, identifying respiratory complaints in GCA patients is crucial, as it may cause a wide range of involvement of vascular inflammation necessitating prompt immunosuppressive therapy.

Table 1: Reported respiratory manifestations in patients with giant cell arteritis: literature overview.

Name of author	No. of GCA patients studied	Percent of patients with respiratory involvement	Presenting symptoms
Larson et al[11]	16	4-6	Cough, sore throat, hoarseness, diffuse tenderness of the anterior neck
Zenone et al[12]	88	13.60	Dry cough
Sonneblick et al[9]	1	-	Dry cough, direct lung involvement: diffuse reticular pattern, visceral disseminated form Involvement of pulmonary artery
Manganelli at al[13]	1	-	Non-productive cough, sore throat, hoarseness, choking sensation, thoracic pain, pleural effusion, interstitial lung disease, basal interstitial fibrosis, intra-alveolar hemorrhage

CONCLUSION

Very limited literature exists on exertional breathlessness as a clinical manifestation of giant cell arteritis. An evolving pulmonary vasculitis could be the cause for the exertional breathlessness in our patient and the symptom responded promptly to immunomodulation. This highlights the importance of considering atypical presentations in GCA. Early diagnosis and treatment are very crucial to prevent complications associated with GCA, such as vision loss and large vessel involvement.

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