ORIGINAL RESEARCH PAPER

Rheumatology

KEY WORDS: Giant cell arteritis (GCA), superficial temporal artery (STA), headache, polymyalgia rheumatica (PMR)

AN OVERLOOKED HEADACHE

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ABSTRACT

Giant cell arteritis, otherwise known as temporal arteritis or cranial arteritis is a common cause of vasculitis among older adults. We report a case of Giant Cell Arteritis in an elderly female patient who presented with a recent onset unilateral headache and symptoms of polymyalgia rheumatica. Prompt diagnosis and comprehensive workup can help an alert physician to clinch the diagnosis and be instrumental in early treatment and thereby prevent permanent blindness.

INTRODUCTION

Giant cell arteritis (GCA) is a chronic granulomatous inflammatory vasculitis involving medium & large sized vessels [1]. This disease predominantly affects elderly, with peak incidence occur in 70-80 age group. Most common in females than males with ratio 3:1 in some countries. Activated T-cells produces IFN-gamma, which seems to play a role in the pathogenesis and clinical expression.

The American College of Rheumatology, has laid down the diagnostic criteria for GCA [2], demands the fulfillment of at least two of the following five criteria(i)Age>50 years (ii)New headache (iii)superficial temporal artery tenderness or decreased pulsation (iv)elevated ESR>50 mm in the first hour (v)abnormal findings on temporal artery biopsy. Below we present a case of an overlooked headache in an elderly female, the prompt diagnosis & treatment prevented the catastrophe of blindness [7].

Figure 1: Thickened Nontender Superficial Temporal Artery

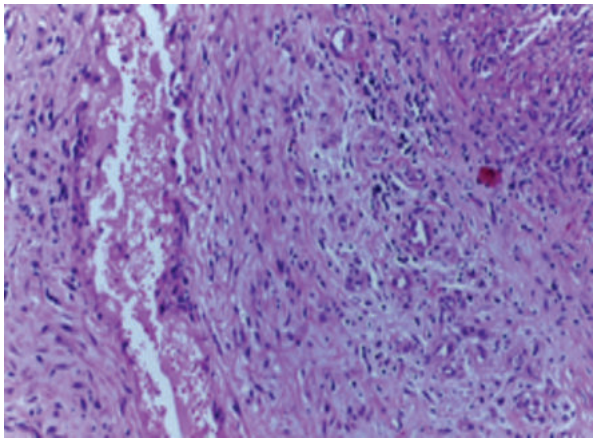

Figure 2: Section showing blood vessels with extensive transmural & perivascular lymphoplasmocytic infiltrate.

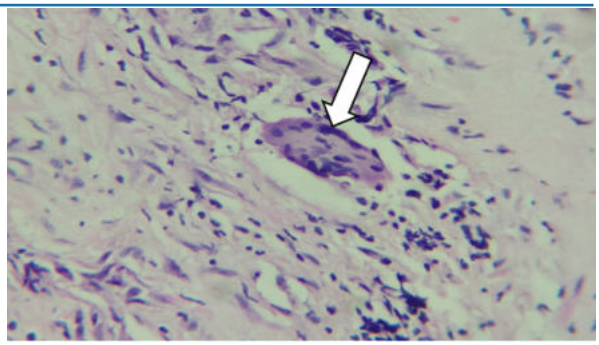

Figure 3: Arrow Indicates Giant Cells.

CASE SUMMARY

A 70-year-old lady with no known comorbidities presented with history of headache, malaise, weight loss & arthralgia of 2 months duration. Headache was unilateral, throbbing pain predominantly over the left hemicranium, with maximum intensity over the left temple. She has associated malaise, weight loss, loss of appetite and shoulder pain aggrevated by movements & had morning stiffness.

With these symptoms she had multiple contacts with physicians and neurologists, she was evaluated for chronic meningitis, intracranial space occupying lesion with CSF study, CTbrain, MRI brain. Also worked up for tuberculosis, malignancy, sarcoidosis etc all turned out to be negative. She was on various migraine prophylaxis & analgesics, without much relief. She denied any history of visual disturbance, other cranial nerve symptoms, scalp tenderness, jaw claudication, fever.

On examination, the patient was conscious, oriented with stable vitals. Xanthelasma present, left superficial temporal artery STA (Figure 1) was thickened, non- tender and non-pulsatile. System examination was unremarkable. Laboratory investigations revealed mild anemia and elevated ESR of 120 mm/1hr. With high degree of suspicion of GCA we proceeded with STA biopsy (Figure 2) which showed blood vessels with extensive transmural & perivascular lympho-plasmocytic infiltrate & giant cells (figure 3) consistent with giant cell arteritis.

Our patient fulfilled all 5 of the ACR criteria & was diagnosed as GCA. Patient was initiated on oral corticosteroid with initial dose of 60 mg per day for the first 2 months & gradually tapered the dose. Patient had resolution of her symptoms of headache & PMR within 2 weeks of treatment initiation. She was advised to continue on low dose steroid for at least 2 years before stopping to avoid the rate of relapse.

DISCUSSION

Temporal arteritis or GCA is the most common vasculitis in elderly population. Most typical manifestations being headache, polymyalgia rheumatica (PMR), jaw claudication, tender superficial temporal artery & visual disturbance. There is an interesting yet incompletely understood relationship between PMR &GCA. Studies shows that 16-21% of the patients with PMR develop GCA [3]. Conversely, PMR is noted in 58% of cases diagnosed with GCA [4]. GCA presenting solely as PMR, without or only minimal cranial nerve symptoms carries a 27.4% increased risk of ischemic episodes, as per available data [5].

The most dreaded complication of GCA is visual loss. It is due to ischemia in the optic nerve & optic tracts, developing because of vasculitis of the ophthalmic & posterior ciliary artery. Gold standard in the diagnosis is biopsy of STA with segment length of 4-6cm. Classical histopathological picture of GCA is giant cells located at the junction between intima & media (seen in 50% of the cases), pan arteritis with mixed inflammatory cell infiltrate. The involvement is typically patchy (skip lesions). Other non-invasive modalities emerging as alternatives in aiding diagnosis are Duplex Sonography of the temporal artery, High resolution MRI & FDG-PET scan. Duplex sonograph demonstrates a characteristic dark halo around the artery with a reported specificity of 100% and sensitivity of 73% [5]. MRI & FDG-PET has a promising role in demonstrating vasculitis, especially in active disease [6].

CONCLUSIONS

This case highlights the importance of considering the possibility of GCA in elderly patients presenting with new onset headache and should prompt the physician to work up for GCA, amongst other possibilities. And the red flag signs being PMR symptoms, elevated ESR & STA abnormalities. GCA responds excellently to steroids. Timely diagnosis and treatment can prevent the most ominous complication – irreversible visual loss.

REFERENCES:

[1] Horton BT, Magath TB, Brown GE. (1932), An undescribed form of arteritis of the temporal vessels. Mayo Clin Proc 7:700-1.
[2] Hunder GG, Bloch DA, Michel BA, Steves MB, Arend WP, et al. (1990), The American College of Rheumatology1990 criteria for the classification of giant cell arteritis. Arthritis Rheum; 33:1122-8.
[3] Salvarani C, Gabriel SE, O''Fallon WM, HunderGG. [1995] Epidemiology of polymyalgia rheumatic in Olmsted Country, Minnesota,1970-1991. Arthritis Rheum; 38:369-73.
[4] Salvarani C, Gabriel SE, O''Fallon WM, HunderGG [1995] The incidence of giant cell arteritis in Olmsted Country, Minnesota. Apparent fluctuations in a cyclic pattern. Ann Intern MED; 123:192-4.
[5] Hernandez-Rodriguez J, Font C, Garcia-Martinez A, et al. [2007]. Devolopment of ischemic complications in patients with giant cell arteritis presenting with apparently isolated Polymyalgia rheumatic: A study of a series of 100 patients. Medicine (Baltimore); 86:233-41.
[6] Schmidt WA, Kraft HE, Vorpahl K, Gromnica-lhle EJ. [2007] Color duplex ultrasonography in the diagnosis of temporal arteritis. N England J Med; 337:1336-42.
[7] Walter WA [2007] Flurodeoxyglucose PET in large vessel vasculitis. Radiol Clon North Am; 45: 735-44.
[8] Khosla P,Gogia A, AggarwalPK,Jain KP. [2004] Unilateral central retinal artery occlusion followed by contralateral anterior ischemic optic neuropathy. J Postgrad Med ; 50: 219-21.