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Medicine

JUVENILE TEMPORAL ARTERITIS—A RARE ENTITY

Pranav J. Nadkarni	MBBS, Junior resident, Department Of Medicine, Goa Medical College, Bambolim - Goa. 403202.
Tanaya D. Dhopeshwarka	MBBS, Junior resident, Department Of Medicine, Goa Medical College, Bambolim - Goa. 403202.
Kedareshwar Narvencar*	M. B. B. S., M.D.(Int. Medicine)Assistant Professor, Department of Medicine, Goa Medical College, Bambolim -Goa. 403202. *Corresponding Author

ABSTRACT Superficial Temporal arteritis or Giant Cell Arteritis is very rare below the age of 50 years old. While the disease is common in older individuals, one has to keep this diagnosis in mind in young individuals presenting with typical symptoms. We present a case of biopsy-proven giant cell temporal arteritis in 14-year-old girl who presented with jaw claudication pain and headache for 6 months. The current case is reported in view of its rarity.

KEYWORDS : Juvenile Temporal arteritis, Giant cell arteritis, superficial temporal arteritis

INTRODUCTION:

Temporal arteritis, also known as giant cell arteritis (GCA), is a granulomatous vasculitis that commonly occurs in older adults, mainly those over 50 years old. Although rare, young patients under 40 years of age could also develop vasculitis in the temporal artery.[1]

The presentation could be varied, some may present with systemic vasculitis involving temporal artery, others may present with giant cell arteritis similar to the one seen in adults, while some others may have eosinophilic arteritis localised to temporal artery (Juvenile temporal arteritis). While this disease is rare in young individuals, it should be considered in differential diagnosis in patients presenting with temporal pain or nodular lesions in temporal area.

PATHOLOGY

The histopathologic features of juvenile temporal arteritis are eosinophilic panarteritis and thrombosis with or without micro aneurysmal disruption of the artery. Intimal proliferation, disruption of the media, and an extensive inflammatory infiltrate, consisting predominantly of lymphocytes and a moderate number of eosinophils and plasma cells, are noted. The absence of granulomatous inflammation and giant cells helps distinguish this variant from the classic type of temporal arteritis.[2]

Although JTA is a localized disease, it is often accompanied by peripheral blood eosinophilia, so some authors have suggested the concept of JTA with eosinophilia.[3] However, its association with pathogenesis, clinical features, and prognosis of JTA remains unclear.

CLINICAL FEATURES

The clinical features that characterize juvenile temporal arteritis are nodular swelling in the region of temporal artery which may be painful or painless, unilateral or bilateral and headache.

Its occurrence in children or young adults and the absence of typical features of classical temporal arteritis such as myalgia, visual disturbance, fever, anemia, and an increased erythrocyte sedimentation rate, are the characteristics of this condition. The visual symptoms noted in classic temporal arteritis reflect the involvement of the ophthalmic artery, however this has not been observed in juvenile temporal arteritis. [2]

As discussed above, juvenile temporal arteritis is a rare entity. A clear distinction between juvenile temporal arteritis and classic temporal arteritis is clinically important because high-dose corticosteroid therapy is almost standard management for the latter but may be unnecessary in juvenile temporal arteritis.

TREATMENT

Case reports of JTA have stated that surgical excision of superficial temporal artery is the treatment of choice and has shown no recurrence of symptoms at follow up. Use of corticosteroids or immuno supressants in this condition is controversial.[1]

CASE REPORT

A 14 year old female presented to our OPD with jaw claudication for 6 months.

The patient also had bilateral throbbing headache and malaise for the same duration. The patient denied having visual disturbance or muscle pain.

On examination, there was severe tenderness on both sides of scalp over the superficial temporal artery. No mass or thickening was palpable. The patient's past medical history was insignificant.

Laboratory investigations at admission revealed erythrocyte sedimentation rate of 60 mm/h, and a positive C-reactive protein. ANA and P ANCA were both negative. Peripheral blood examination showed iron deficiency anemia with Hb of 7.1g%, a white cell count of 6300/microL, and a normal differential count. No peripheral eosinophilia was noted. Serum electrolytes, renal and liver function test were all within normal limits.

An ophthalmologic evaluation was unremarkable.

Superficial temporal artery biopsy revealed congested fibrocoll agenous tissue around the artery with lymphocytic infiltration of arterial wall and surrounding tissue with absence of granulomatous inflammation and giant cells suggestive of JTA.

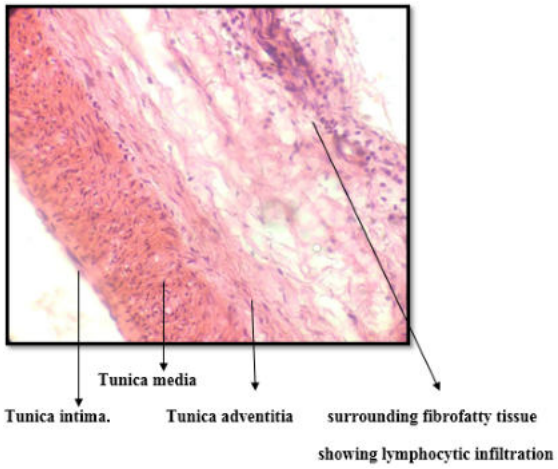

Fig 1—histopathology image showing inflammation in temporal artery (magnification 40 X)

The patient was treated with oral prednisone (1mg/kg daily). However patient had no response to treatment after one month of therapy.

In view of deterioration in symptoms, patient was started on oral methotrexate 10mg once a week in addition to steroid. The patient showed dramatic improvement in 1 month of this therapy.

Prednisone was tapered over 3 months currently patient is on regular follow up with methotrexate and tapering doses of steroids and is in complete remission.

DISCUSSION

Vasculitis involving temporal artery is typically rare in individuals below 40 years of age. Juvenile temporal arteritis (JTA), polyarteritis nodosa, Eosinophilic Granulomatosis with polyangiitis and Elderly type vasculitis like GCA should be considered in the differential diagnosis in a young patient presenting with features of inflammation of temporal artery.

Clinical manifestations of JTA are as follows: age younger than 40 to 50 years, a palpable nodule or vessel engorgement in the area of temple (which can either be painful or painless), and a localized disease. Systemic involvement is rare. ESR is usually normal.4 Peripheral blood eosinophilia could be present.

In 1975, Lie et al. first reported four cases of JTA in the young.[5] These patients complained of painless nodules on their temple, and the pathologic findings revealed nongiant cell inflammation of temporal arteries

From then on until February 2019, 19 more cases have been reported; their mean age was 27 years (range, 7 to 44 years) with a male predominance (18 men and 5 women).[1,6,7,8] Only seven cases out of 23 cases involved both sides of temples. Two out of twenty three presented with headache without presence of a nodule similar to the presentation in our patient.

JTA is a localized disease, hence excision of the artery is curative, steroid treatment is usually not required after excision, recurrence is rare.[6]

Our case, though fitting in the diagnosis of Juvenile temporal arteritis, had some atypical features. The biopsy of Superficial temporal artery showed panarteritis in absence of granulomatous inflammation or giant cells, hence the diagnosis of classical Giant Cell Arteritis was excluded. Eosinophilic granulomatosis with polyangiitis was also excluded as the histopathology was inconsistent with the same and PANCA was negative.

Though our patient did not present with the features of juvenile temporal arteritis such as nodular lesion in the temporal arteries, the typical histological picture and absence of systemic involvement in this young patient makes the diagnosis of Juvenile Temporal arteritis more likely. However there was no eosinophilia on peripheral smear and ESR was raised.

Our patient was unresponsive to steroids however symptoms subsided with use of Methotrexate. Hence the excision of the artery was not considered.

In conclusion, when any young patient presents with nodular lesions at temple, JTA should be considered in the differential diagnosis.

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