Temporal arteritis: A case series from south India and an update of the Indian scenario

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Abstract

Objective: To study the clinical, pathological and prognostic profile of patients with temporal arteritis in India. **Materials and Methods:** The study was conducted in a tertiary care center from south India from 2005 to 2010 in the departments of neurology and medicine. The details of all patients that satisfied the ACR 1990 criteria for diagnosis of temporal arteritis were reviewed. The clinical presentation, laboratory parameters and biopsy findings of the patients were analyzed and compared with other studies from India done over the last decade. **Results:** A total of 15 patients were diagnosed with temporal arteritis. The male:female ratio was 1.5:1. The mean age of onset was 67.58 years. Mean time for detection after onset of symptoms was 2.56 months. Typical manifestations included headache (100%), temporal artery tenderness (100%), jaw claudication (20%), polymyalgia rheumatica (53%) and visual manifestations (20%). The erythrocyte sedimentation rate was elevated in all patients. Biopsy was done in 13 patients, with 11 of them being positive. All patients responded to steroids well, with most patients being symptom-free within the first 48 h of treatment. **Conclusions:** Temporal arteritis seems to be underdiagnosed in India, with all patients previously misdiagnosed, and with a mean time from symptom onset to diagnosis of 2.5 months. The clinical presentation of temporal arteritis in India appears to be similar to that of the West, with no gender preference and a slightly younger age group.

Key Words

Blindness, giant cell arteritis, headache

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Ann Indian Acad Neurol 2012;15:27-30

Introduction

Temporal arteritis or giant cell arteritis is the most common vasculitis in the elderly population, with an incidence of 15–30 per 100,000 persons aged greater than 50 years in the North American and European countries.[1-4] However, the incidence is believed to be much lower in Asian countries, with the incidence in Japan reported to be as low as 1.47 per 100,000.[5] In the past, only a few case studies have been reported from India.[6-9] The present study describes the clinical, pathologic and prognostic profile of 15 cases of temporal arteritis diagnosed and treated in our institution, a tertiary care hospital in south India from 2005 to 2010. We have compared our data with those from other case series conducted from India in the last decade.

Access this article online

Quick Response Code:

Website: www.annalsofian.org

DOI: 10.4103/0972-2327.93272

Materials and Methods

The inpatient and outpatient records of all elderly individuals above 50 years of age who had presented with features suggestive of temporal arteritis to the neurology and medicine services of our institution between 2005 and 2010 were reviewed. A clinical diagnosis was made to determine whether the symptoms satisfied any three criteria of the American College of Rheumatology 1990 Criteria for the diagnosis of giant cell arteritis.[10] The demographic profile, duration of symptoms before diagnosis and associated symptoms such as polymyalgia rheumatica, visual loss and neurological manifestations were noted. Laboratory parameters such erythrocyte sedimentation rate (ESR) and C-reactive protein were also documented. The histopathological features of temporal artery biopsies were reviewed in detail. Patient outcomes were also analyzed.

Results

A total of 15 patients fulfilled the criteria for diagnosis of temporal arteritis. Nine of the 15 patients were male and six were female, with a male to female ratio of 1.5:1. The mean age of onset was 67.53 years (SD 9.13, range 52–81). The mean duration

of symptoms was 77 days (2.56 months; range 20–120 days). All the patients had recent onset of bilateral or unifrontal headache. Tender temporal arteries with absent or diminished pulsations were common to all the patients. Nodularity on palpation was noted in seven patients (47%). Jaw claudication was present in only three of the patients (20%). Three of the patients had ipsilateral blindness due to central retinal artery occlusion (20%). Eight patients had features of polymyalgia rheumatica (53%). All patients had a raised ESR (mean 90.8 mm/1st h) and raised C-reactive protein (mean 5.38). Superficial temporal artery biopsy was planned in all, but two patients did not give consent. Summary of the findings are included in Table 1.

Biopsy length varied from 3 to 5 cm and was conducted within 48 h of initiation of treatment. Eleven of the 13 cases who underwent biopsy were positive for temporal arteritis (84.6%). Nine of them had characteristic histological features of temporal arteritis on biopsy, which included multinucleated giant cells [Figures 1a–d]. While in the other two cases, changes reflected chronicity with medial hypertrophy and fibrosis causing luminal narrowing, lymphohistiocytic infiltration but no granulomatous response. The two patients that were biopsy-negative refused repeat biopsy on the opposite side.

All our patients responded to steroids, with more than 80% of the patients having full symptomatic recovery within 48 h. Only one patient developed complication of lung abscess on steroid therapy and was treated with antibiotics. All patients were on regular follow-up for at least 6 months after diagnosis and were kept on treatment with low-dose steroids with no relapse of symptoms during the treatment period. As ours is a tertiary care hospital with many of the patients coming from long distances, it was possible for us to retain only three patients for long-term follow-up of more than 2 years. Steroids was tapered and stopped after 2 years of diagnosis in these patients and no relapse was reported after stopping the steroids. The clinical presentation was compared with other studies from India [Table 2].

Discussion

Temporal arteritis is a granulomatous inflammatory disease affecting large- and medium-sized arteries. The pathogenesis of temporal arteritis is still unclear. Activated T cells produce interferon-γ, which seems to play a pivotal role in the pathogenesis and clinical expression.[11] Both environmental[12,13] and genetic factors[14,15] seem to play a role in the etiology of temporal arteritis.

Temporal arteritis mostly affects the elderly population, appearing almost exclusively in patients aged >50 years, with the mean age of onset around 75 years of age.[1] It is also more

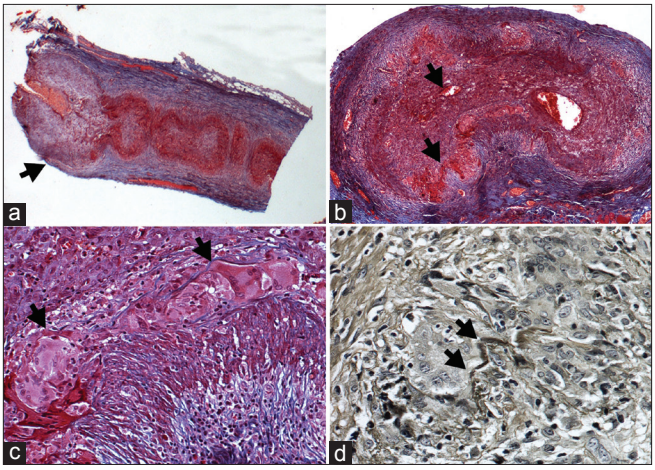

Figure 1: Temporal artery biopsy viewed in the longitudinal axis demonstrates striking beading of the wall with nodular distensions causing luminal narrowing (a). Note the bulbous end (arrow, a) that shows dense inflammation expanding the medial coat. Close-up view of the vessel in the transverse plane (b) reveals expansion of the medial coat by multiple aggregates of epitheloid cells forming granulomas (b, arrows) with large multinucleate giant cells (c, arrows). Several of the multinucleate giant cells have ingested fragments of elastica from the internal elastic lamina (d, arrow)

Table 1: Demographic, clinical and laboratory characteristics of the patients

Age/sex	Location	Symptom duration in days	Jaw claudication	PMR	Visual symptoms	ESR (mm in 1st hr)	CRP (mg/dL) (normal <0.33)	Biopsy
57 F	Left	30	No	No	No	80	3.49	Positive
52 F	B/L	60	No	No	No	75	1.6	Positive
70 F	Left	60	No	No	No	83	0.7	Negative
72 F	B/L	90	No	No	No	160	14.24	Not done
55 M	B/L	120	No	No	No	71	1	Positive
57 M	B/L	30	No	Yes	No	75	0.80	Positive
69 M	LEFT	60	Yes	Yes	No	68	1.3	Positive
82 M	LEFT	180	Yes	Yes	No	88	5.57	Positive
76 M	B/L	60	No	Yes	No	80	9.9	Negative
81 F	RIGHT	30	No	No	Yes	150	7.7	Positive
65 M	LEFT	30	Yes	Yes	Yes	100	12.3	Positive
70 M	B/L	90	No	Yes	No	130	Positive*	Positive
64 F	B/L	180	No	Yes	No	60	Positive*	Not done
72 M	Right	120	No	Yes	Yes	92	Positive*	Positive
71 M	Right	20`	No	No	No	50	6.75	Positive

*Quantitative CRP was not performed in three cases, *ESR = Erythrocyte sedimentation rate; CRP = C-reactive protein; PMR = Polymyalgia rheumatica; TA = Temporal artery

Table 2: Comparative data of patients with temporal arteritis in India: Demographic and clinical profiles

	Present study (n = 15)	Singh *et al.*[6] (n = 16)	Vankalakunti *et al.*[7] (n = 9)	Laldinpuii *et al.*[8] (n = 4)
Male:female ratio	1.5:1	1:1	1.25:1	1:1
Mean age (years)	67.5	66.5	70	74.25
Mean duration of illness (in months)	2.56	5.18	1.85	6.25
Clinical features				
Headache (%)	100.0	93.7	66.7	100.0
Jaw claudication (%)	20.0	56.2	22.2	50.0
TA tenderness (%)	100.0	68.7	22.2	100.0
PMR (%)	53.3	31.25	-	100.0
Visual symptoms (%)	20.0	18.75	22.2	25.0
ESR >50 (%)	100.0	100.0	88.8	75.0
Biopsy positive (%)	84.6 (11/13)	90.9 (5/6)	87.5 (7/8)	75.0 (3/4)

TA = Temporal artery; ESR = Erythrocyte sedimentation rate in mm after 1 hour; PMR = Polymyalgia rheumatica

common in women than in men, with a ratio as high as 3:1 in some countries. In our study, the mean age of onset was slightly lower at 67.53 years (52–81 years), with all patients older than 50 years of age. The male to female ratio was 1.5:1. This incidence pattern of slightly younger age group and male predominance when compared with the Caucasian population was also observed in other case studies from India.[6-8]

The mean time taken to detect the disease after onset of symptoms was 77 days or 2.56 months in our study. In other case series from Mumbai, India, it was 5.14 months, and it was 6.25 months from a case series in northeast India.[6,8] In another study from China, the mean time taken for diagnosis after symptom onset was 5.5 months, with a misdiagnosis rate of 87.5%.[16] All patients had multiple visits to many physicians before presenting to our hospital. The mean time taken for diagnosis after presenting in our hospital was 5.6 days.

The most typical manifestations of temporal arteritis include headache, tenderness on palpation, jaw claudication, polymyalgia rheumatica and visual disturbances.[17] Headache is seen in about 87% of the patients[17] with temporal arteritis. All our patients presented with headache, and about half the patients (seven of the patients) gave a history of bifrontal headache. Jaw claudication is present in nearly 40% of the patients with temporal arteritis. Only three of our patients presented with jaw claudication (20%), which was similar to another case study from south India.[7] Eight patients had clinical features of polymyalgia rheumatica (53.3%), which is in comparison with that of the West.[17] The percentage of cases with polymyalgia rheumatica varied widely from zero to 100% in the other case series from India. Visual disturbances are seen in about 30% of the cases, with permanent loss of vision in about 15% of the cases.[18] Three of our patients who had visual symptoms (20%) had ipsilateral blindness that did not improve upon treatment. Permanent visual loss was less common in the other case studies from India.

Less common manifestations from India included pyrexia of unknown origin,[7,19] peripheral neuropathy[6] and scalp tenderness.[6] None of our patients had any of these symptoms.

Superficial temporal artery biopsy is the definite test for diagnosis of temporal arteritis and, hence, is advised for all our patients. Eleven of the 13 patients who underwent biopsy were positive for the same (84.6%). Steroids have been the drug of choice for the treatment of temporal arteritis. The initial dose of corticosteroids recommended is 40–60 mg for the first 2 months, with gradual tapering of the dose. It is advised that the patient should be on low-dose steroids for at least 1–2 years before stopping steroids to reduce the rate relapse.[20] A relapse rate of up to 77% has been reported in patients that have abruptly stopped steroids.[21] Steroids were tapered and stopped after 2 years in all our patients who were on long-term follow-up, and no relapse has occurred in any of these patients.

Conclusion

Although ethnicity may have a role in the incidence of temporal arteritis, it is still underdiagnosed in India. All the patients that were diagnosed as temporal arteritis at our institution had prior visits to many physicians with a misdiagnosis rate of 100% and mean time to diagnosis of 2.5 months.Temporal arteritis in India seems to have minimal gender preference, with a slight male predominance in our study. It also seems to present at a slightly younger age group than in the Caucasian population. Permanent blindness is an important complication. The response to steroids was excellent in our study. In conclusion, the clinical presentation of temporal arteritis in India appears to be similar to that of the West.

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How to cite this article: Mathew T, Aroor S, Devasia AJ, Mahadevan A, Shobha V, Nadig R, Varghese R, Shankar SK, Sarma G. Temporal arteritis: A case series from south India and an update of the Indian scenario. Ann Indian Acad Neurol 2012;15:27-30.

Received: 20-11-11, **Revised:** 10-12-11, **Accepted:** 17-12-11

Source of Support: Nil, **Conflict of Interest:** Nil