Giant Cell Arteritis in Mumbai

S Singh*, C Balakrishnan*, G Mangat†, R Samant†, M Bambani**, S Kalke**, VR Joshi*  

Abstract  
Objective: To study the clinical profile of patients with giant cell arteritis in Mumbai.  
Methods: From our database, patients with a diagnosis of giant cell arteritis (GCA) over a fifteen year period (January 1990 to December 2005) were included. Clinical manifestations, temporal artery biopsy, treatment, and follow-up data of these patients were analyzed.  
Results: Twenty one patients with GCA were identified. However, data were available only for sixteen patients. The median age at onset was 66.5 years (58 – 78 yrs) with male to female ratio of 1:1. The mean time from symptom onset to diagnosis was 5.18 months (0.5 – 24 months). Clinical manifestations included new onset headache (15), fever (9), weight loss (9), jaw claudication (9), polymyalgia rheumatica (5), visual disturbances (3), scalp nodule (1), temporal artery tenderness (11), tortuosity (9), and scalp tenderness (6). ESR was elevated in 15 patients with a median of 106.5 mm at 1 hr (25 – 135 mm/hr). Temporal artery biopsy was done in 11 patients and confirmed the diagnosis in 10 patients. Color doppler study of the temporal arteries (9 patients) revealed halo sign (indicating arterial wall edema) in 6 patients. Biopsy as per site by color doppler study was performed in 6 of these patients and was positive in 5. All patients had a good initial response to steroids, however, on follow up, 3 patients required addition of methotrexate. At a median follow up (n = 14) of 6 months (range 6 – 156), steroids were successfully stopped in 7 patients at 1 to 3 years interval. The disease relapsed in 1 patient. Of the remaining 7 patients, 2 were steroid dependent and 5 patients were doing well on low dose prednisolone.  
Conclusion: GCA, though uncommon in India, should be suspected in all elderly patients with a new onset headache, fever, jaw claudication, or high ESR. Color doppler sonography is a useful noninvasive method for the diagnosis of GCA and also helps to identify the site to biopsy. Most respond to steroid therapy while some need addition of steroid sparing agents.

Table 1: Clinical manifestations in patients with GCA (n = 16)  

<table>
<thead>
<tr>
<th>Manifestation</th>
<th>Count</th>
</tr>
</thead>
<tbody>
<tr>
<td>New headache</td>
<td>15</td>
</tr>
<tr>
<td>Fever</td>
<td>9</td>
</tr>
<tr>
<td>Weight loss</td>
<td>9</td>
</tr>
<tr>
<td>Jaw claudication</td>
<td>9</td>
</tr>
<tr>
<td>Polymyalgia rheumatica</td>
<td>5</td>
</tr>
<tr>
<td>Visual disturbances</td>
<td>3</td>
</tr>
<tr>
<td>Neurological features</td>
<td>3</td>
</tr>
<tr>
<td>Temporal artery tenderness</td>
<td>11</td>
</tr>
<tr>
<td>Tortuosity</td>
<td>9</td>
</tr>
<tr>
<td>Scalp tenderness</td>
<td>6</td>
</tr>
<tr>
<td>Scalp nodule</td>
<td>1</td>
</tr>
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</table>

Introduction

Giant cell arteritis (GCA) affects mainly the elderly. Large sized arteries are mainly involved. Though frequently seen in European countries, it is uncommon in Asians. There are no large series reported from India. We present our experience from Mumbai.

Material and Methods

Three rheumatology centers in Mumbai participated in this retrospective study. All patients who satisfied the ACR criteria for GCA over a 15 year period were included. The clinical and investigatory parameters recorded were: demographic features, duration of symptoms before diagnosis, clinical features, ESR, color doppler study, temporal artery biopsy, treatment and follow up.

Results

Twenty one patients with GCA were identified. However, data were available only for sixteen patients. The median age at onset was 66.5 years (58 – 78 yrs) with male to female ratio of 1:1. The mean time from symptom onset to diagnosis was 5.18 months (0.5 – 24 months).

The clinical manifestations were as shown in the Table 1. New onset headache was the commonest presenting manifestation. Visual disturbances were present in 3 patients, of which 2 had temporary blurring while 1 had diplopia. Neurological manifestations were present in 3 patients (2 had transient ischemic attacks and 1 had peripheral neuropathy). Permanent visual loss or extra-cranial manifestation was not found in any patient.

The ESR was elevated in 15 patients with a median of 106.5 mm at 1 hr (range 25 – 135 mm/hr). Eight patients had an ESR more than 100 mm while 7 had values between 50 – 100 mm at 1 hr. Color doppler study of the temporal arteries was done in 9 patients. The findings included halo sign, indicating arterial wall edema (Figure 1) in 6 patients and circumferential thickening of the vessel wall in 7 patients. Biopsy as per site by color doppler study was performed in 6 of these patients and was positive in 5. All patients had a good initial response to steroids (1 mg/kg...
However, on follow up 3 patients required addition of methotrexate. Follow up data were available for 14 patients with a median duration of 16 months (range 6 – 156). Steroids were successfully stopped in 7 patients at 1 to 3 years interval. However, 1 patient relapsed after 3 years but responded to steroids again. Two patients were steroid dependent and 5 patients were doing well on low dose prednisolone. The only serious complication of therapy was military tuberculosis in 1 patient, which responded to antituberculous treatment.

Discussion

Giant-cell arteritis affects large and medium-sized vessels usually involving the cranial branches of the arteries originating from the aortic arch. Prevalence varies considerably in different ethnic and racial groups. It occurs commonly in the Caucasians (annual incidence in Scandinavia of 15-35 cases per 100,000 persons over the age of 50) and is uncommon in blacks, Hispanics or Asians (1.47 per 100,000 populations in Japan).1-7 In India, GCA is considered to be a rare entity. The data available has been in the form of case reports or small case series.8-10 To the best of our knowledge, this is the largest series being reported from India.

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The median age at onset in our study was 66.5 years with no sex predilection. GCA is about three times more common in females among the Caucasians.3 Hu Z et al from China reported an early age of onset with 81.5% of patients under the age of 50, predominantly affecting the males.5 However, Hashimoto H et al (Japan) and Vas CJ et al (India) did not find a significant male preponderance.6,7

The clinical presentation of our patients had some important differences (Table 2). Jaw claudication was present in 56.25% of our patients. This is much higher than that reported from other Asian countries. In comparison to the Caucasian population, polymyalgia rheumatica and permanent visual loss were less common.

Visual symptoms occur in about 30% of patients, with loss of vision reported in 15 %.12 Fortunately none of our patient had permanent visual loss. The less common manifestations included scalp nodule, peripheral arthritis, erythema nodosum and peripheral neuropathy. The extracranial manifestations in GCA include aortic involvement manifesting as thoracic aortic aneurysm and dissection of the aorta or large-artery involvement which may manifest as claudication. Though none of our patients presented with extra-cranial manifestations, they were not routinely investigated for these features.

Temporal artery biopsy remains the definitive test for diagnosis of GCA with a sensitivity of 60 to 80 percent.13 Color doppler sonography has been found useful in the diagnosis of GCA.14-17 Evidence of a dark halo on ultrasonography, representing arterial wall edema is considered to be a specific sign of active disease. In a recent meta-analysis, the halo sign achieved an overall sensitivity of 69% and an overall specificity of 82 % compared with temporal artery biopsy.17 We performed color Doppler study of the temporal artery in 9 patients. The halo sign was seen in 6 patients while circumferential thickening of the vessel wall in 7 patients. Temporal artery biopsy was done in 6 of these patients and was positive in five.

Glucocorticoids have been the treatment of choice for GCA. All patients in our study received 1 mg/kg of prednisolone with initial good response. However, on follow up, methotrexate (10 -15 mg/week) was added in 3 patients. Of these, 1 had developed side effect of steroids in the form of excess weight gain and diabetes mellitus while other 2 patients were requiring high maintenance doses of steroids. There have been conflicting reports in the literature regarding the use of methotrexate in GCA.18 The only serious complication of therapy included military tuberculosis in 1 patient, which responded to anti tuberculous treatment. There was no mortality related to GCA.

GCA is usually a self limiting disease with most patients requiring treatment for 1 to 2 years.19 However, some may require continued drug therapy for longer periods of time. In our study, steroids were successfully stopped in 7 patients (50%) at 1 to 3 years interval. However, 1 patient relapsed after 3 years of discontinuation but responded to steroids again.

Table 2 : Comparative data of patients with GCA

<table>
<thead>
<tr>
<th></th>
<th>Present study</th>
<th>India (8)</th>
<th>Japan (7)</th>
<th>Caucasian (3)</th>
<th>China (6)</th>
<th>N = 7</th>
<th>N = 16</th>
<th>N = 55</th>
<th>N = 66</th>
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<tbody>
<tr>
<td>Mean age (years)</td>
<td>66.5</td>
<td>66.85</td>
<td>74</td>
<td>71.5</td>
<td>43.3</td>
<td>1.3:1</td>
<td>1.1:1</td>
<td>1:1</td>
<td>1:1</td>
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<tr>
<td>Jaw claudication (%)</td>
<td>56.25</td>
<td>31.25</td>
<td>31.25</td>
<td>32.7</td>
<td>30.3</td>
<td>3.1</td>
<td>28.5</td>
<td>14</td>
<td>14</td>
</tr>
<tr>
<td>Visual symptoms (%)</td>
<td>18.75</td>
<td>28.5</td>
<td>28.5</td>
<td>30.2</td>
<td>18.75</td>
<td>1.7:1</td>
<td>1:1</td>
<td>1:1</td>
<td>1:1</td>
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</table>

Fig. 1 : Color Doppler study of the temporal artery showing halo sign

Fig. 2 : Biopsy of superficial temporal artery showing multinucleated giant cell
Conclusion

GCA, though uncommon in India, should be suspected in all elderly patients with a new onset headache and high ESR. The epidemiology is similar to the Caucasian population. Although jaw claudication is more frequently present, polymyalgic symptoms and permanent visual loss are less common. Color doppler sonography is useful in documenting disease activity and identifying the site to biopsy. The present study has limitations due to small number of patients and the retrospective study design. A nationwide prospective study may give a clearer picture regarding this disease.

References