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Research Article

The Demographic, Clinical and Laboratory Profile of Patients with Giant Cell Arteritis: Case Series

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Abstract: The aim of the study was to describe the demographic, clinical and laboratory profile of patients with giant cell arteritis in northeast of Iran. In a retrospective study we reviewed the records of all patients that satisfied the American College of Rheumatology 1990 criteria for diagnosis of giant cell arteritis. The clinical features, laboratory parameters and biopsy findings of the patients were analyzed. A total of 30 patients were diagnosed with giant cell arteritis. The male: female ratio was 1.1:1. The mean age of onset was 63.9 ± 10.4 years. The most common manifestations included headache (96.7%), temporal artery tenderness (62.1%), jaw claudication (23.3%), polymyalgia rheumatica (40%), visual manifestations (36.7%) and fever (20%). The erythrocyte sedimentation rate was elevated in all patients (mean: 88.43 mm/hour). Anemia (Hb<12g/dl) was found in 26% of patients. Biopsy was done in 21 patients, with 15 of them being positive. Of 14 patients with eye involvement, 11 had unilateral and 3 bilateral eye involvement. Visual loss was permanent in all of the affected patients. All patients responded to steroids well (Except for visual loss). There was not any mortality due to giant cell arteritis. In northeast of Iran, it seems that giant cell arteritis has minimal gender preference and it occurs in younger population. Headache and scalp tenderness were more common but, fever, jaw claudication, vertigo and diplopia were less common in our patients. Other features were similar to that of the West. The response to steroids was excellent**.** Irreversible visual loss was an important complication.
Keywords: Giant cell arteritis, temporal arteritis, polymyalgia rheumatica, Iran.

INTRODUCTION

Giant cell arteritis (GCA) –or temporal arteritis- was first described in 1890 by Hutchinson [1], and then was more clearly defined in 1932 by Horton et al[2]. GCA is a systemic vasculitis that predominantly affects the extracranial branches of the carotid artery. It occurs among individuals over 50 years of age and is two times more common in women[3]. The most commonly affected populations include Scandinavian and northern Europeans[4]. The etiology and pathogenesis of GCA are unknown. However, increasing age, genetic factors, and infection may have causative roles. Both the humoral and cellular immune systems have been implicated in the pathogenesis[5]. The classic symptoms of GCA include headache, jaw claudication, symptoms of polymyalgia rheumatic, visual symptoms, fever and fatigue. The onset may be gradual or sudden. The most important complication of GCA is blindness, which usually can be prevented by early diagnosis and treatment[6].

Temporal artery biopsy (TAB) is the gold standard for diagnosis of GCA, revealing inflammatory cell infiltration of arterial walls with or without giant cells and disruption of the internal elastic lamina leading to luminal occlusion and tissue ischemia[7].

There is no single clinical feature or laboratory test specific for GCA. In 1990, the American College of Rheumatology (ACR) formulated classification criteria that were effective in distinguishing GCA from other forms of vasculitis[8].

The clinical and laboratory characteristics of this disease have never been studied in Iran. We present the first study evaluating clinical and laboratory characteristics of GCA in the population of Iran; comparing the characteristics of the GCA to internationally reported figures.

MATERIALS AND METHODS

We retrospectively reviewed the case records of all patients diagnosed with giant cell arteritis at the clinic and Department of rheumatology of the Imam Reza Hospital, Mashhad, Iran between January 1, 2002, and December 31, 2012. Patients included in the current study were diagnosed as having GCA when they fulfilled at least three of the 1980 American College of rheumatology criteria for classification of giant cell arteritis [8]. Cases lacking sufficient medical

information were excluded. We use the presence of all of the following three criteria for the clinical diagnosis of PMR: 1) Age ≥50 years at disease onset. 2) Bilateral pain and morning stiffness (lasting 30 minutes or more) persisting for at least one month. The stiffness should involve at least two of the following three areas: neck or torso, shoulders or proximal regions of the arms, and hips or proximal aspects of the thighs. 3) Erythrocyte sedimentation rate (Westergren) ≥40 mm/h[9].

The erythrocyte sedimentation rate (ESR) was measured using Westergren method. An abnormal ESR defined as the patient's age divided by 2 for men, and the patient's age plus 10, divided by 2 for women.

We analyzed demographic, clinical and laboratory data (ESR, CBC) for all patients.

Statistical analysis

We described continuous data as mean and standard deviation (mean ± SD), and categorical variables as percentage.

RESULTS

There were 30 patients (16 male, 14 female). The mean age at diagnosis of giant cell arteritis was 63.9 ± 10.4 years (minimum 44 years, maximum 83 years). The disease was presented as fever of unknown origin (FUO) in one patient. Erythrocyte sedimentation rate (ESR) was elevated in all patients (mean 88.43 mm/1th hour, maximum140 mm/1th hour, minimum 43 mm/1th hour). Temporal artery biopsy was done in 21 patients, which 15 of them being positive. The clinical and laboratory features of studied patients are showed in table 1. Initial dose of prednisolone was 1mg/kg/day (maximum 60 mg). All patients responded to steroids well (Except for visual loss). Visual loss was permanent in all of the affected patients. There was not any mortality in our patients due to temporal arteritis.

Table 1: Clinical and laboratory characteristics of studied patients

Feature	Frequency (%)
Headache	96.7
Fever	20
Any visual symptom	46.6
Unilateral visual loss	36.6
Bilateral visual loss	10
Jaw claudication	23.3
Polymyalgia rheumatica	40
Arteritis	25
Sore throat	6.9
Tinnitus	3.3
Vertigo	3.3
Scalp tenderness	62
Abnormal ESR	100
Anemia	26
Positive biopsy	71

DISCUSSION

To our knowledge, the present report is the first case series of Giant cell arteritis from Iran. The prevalence of GCA in Iran is unknown. There are only a few case reports of GCA from Iran[10-11]. GCA is common in the whites and is uncommon in blacks and Asians [12-15]. Although it has been reported in individual patients younger than 50 years, GCA usually is encountered in the elderly. The mean age of onset is 79 years, with a range from approximately 50 to older than 90 years [16]. In our study, the mean age of onset was 16 years lower (63.97 years). Only one patient was younger than 50 years of age. This incidence pattern of younger age group when compared with the Caucasian population was also observed in other case studies from Asia [12-15]. Many series in the literature report that more women are affected than men. We didn't find this gender preference in our patients. In our study the male: female ratio was 1.1:1 like most reports from Asian population[12-15]. Hu Z et al from China reported a male to female ratio of 15:1.[14]. However, other case studies from Asia did not find a significant male preponderance[12-15].

Headache is the most common systemic manifestation. The frequency of headache in our study was 96.7% which was more than most other studies (Table 1).

The incidence of ocular involvement in GCA ranges between 14 and 70%. Bilateral visual loss has been noted to occur in up to one third of patients with GCA[17]. Although visual manifestations may be intermittent initially, visual deficit is usually irreversible once established[18]. In our patients the frequency of visual symptoms was 43.3%. In 10% of our patients (21.4% of patients with eye involvement) visual symptoms were bilateral. Visual loss was permanent in all of our patients.

The association between temporal arteritis and polymyalgia rheumatica (PMR) is well-known. PMR has been reported in 34%–60% of patients with GCA [19]. Forty percent of our patients had clinical features of polymyalgia rheumatica, which is in comparison with that of the West.

The erythrocyte sedimentation rate is characteristically elevated in patients with temporal arteritis. No definite normal value can be given to the sedimentation rate in the elderly. However, most individuals (more than 95%) with temporal arteritis have a markedly elevated sedimentation rate[20]. All of our patients had elevated ESR.

Temporal artery biopsy (TAB) is the gold standard in the diagnosis of GCA; however, a negative biopsy does not rule out the diagnosis. A biopsy should be performed on all patients suspected of having temporal arteritis. Bilateral TAB has a range of sensitivities

between 69%, and 98.7%, with an overall mean TAB sensitivity of 87.1% [21]. In our study the sensitivity of TAB was 71%. Contralateral biopsy was not done in any of our patients with negative TAB.

The clinical and laboratory features of GCA in our study are compared with some other studies from Asia and Europe in table 2.

Glucocorticoid treatment should be started once the diagnosis of GCA is suspected strongly, often even before it is confirmed. For patients in whom the diagnostic suspicion of GCA is high, especially those with recent or threatened vascular complications such as visual loss, therapy should be started immediately [22]. The initial dose of glucocorticoid is equivalent to 40 to 60 mg prednisolone per day [23]. All of our patients had a good initial response to steroids (1 mg/kg of prednisolone).

The life expectancy of this population is not significantly different than the general population[24]. There was no mortality related to GCA in our patients.

CONCLUSION

In northeast of Iran it seems to have minimal gender preference. In our study, the mean age of onset was lower than that of the West. Headache and Scalp tenderness were more common but, fever, jaw claudication, vertigo and diplopia were less common in our patients. Other features of our patients were similar to that of the West. The response to steroids was excellent in our study. Irreversible visual loss was an important complication in our patients.

Table 2: Comparative data of patients with GCA

	Iran N=30	**Japan [15] N=66**	**China [14] N=16**	**India[12] N=16**	**Caucasian[17]**
Mean Age (years)	63.9	71.5	43.3	66.5	79
F:M	1:1.1	1.7:1	1:15	1:1	3:1
Headache (%)	96.7	80.3	-	93.7	76
Fever (%)	20	55.4	-	56.2	42
Visual symptoms (%)	46.6	6.5	18.75	18.75	37
Jaw claudication (%)	23.3	15.2	Uncommon	56.25	34
PMR (%)	40	30.3	-	31.25	34
Scalp tenderness (%)	62	63.3	-	67.8	31
Arteritis (%)	25	-	-	-	30
Abnormal ESR	100	84.8	-	93.7	96
Anemia (%)	26	-			44
Positive biopsy (%)	71	71.4	-	90.9	86

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