

Is temporal artery biopsy the gold standard for the diagnosis of Giant Cell Arteritis?

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Background: The only test that confirms diagnosis of Giant Cell Arteritis (GCA) is a temporal artery biopsy showing vasculitis with mononuclear cell inflammatory infiltrates, often with giant cells. Due to the focal and segmental nature of the infiltrates, areas of inflammation may be missed by the biopsy. Some imaging modalities may aid in the diagnosis such as color duplex ultrasonography of the temporal arteries. When the clinical suspicion is strong and temporal biopsy is negative or can't be performed, patients are treated as GCA. Our objective was to analyze all patients with GCA seen at our hospital in order to address value of temporal biopsy result in relation to the clinical course.

Methods: we retrospectively reviewed electronic medical records of patients registered in our hospital between 2000-2013 with the problem: vasculitis, Giant Cell Arteritis or Temporal Arteritis. Patients fulfilling ACR 1990 criteria for GCA were included. Clinical and treatment information was collected. Temporal biopsies were reviewed. Ultrasound of temporal arteries was performed by an experienced vascular sonographer if requested by the treating rheumatologist and the finding of the halo sign was considered compatible with GCA diagnosis.

Results: 101 patients were included with GCA diagnosis, 79 females (78.2%), with a mean age at diagnosis of 74.9 years (SD 8.1). Temporal biopsy was positive in 52 patients, negative in 21 and was not performed in 28. Clinical characteristics are shown in table 1 grouped by biopsy result. Multivariate analysis showed that abnormal temporal pulse on examination had an OR of 19.7 (CI 2.9-131.9) for predicting a positive biopsy. Having symptoms of polymyalgia rheumatica (PMR) and age were also associated with a positive biopsy (OR 4.5, CI 1.03-19.4, and OR 1.11, CI 1.01-1.24 respectively). No differences were found in clinical presentation, treatment, relapses or recurrences between groups. Ultrasound was performed in 42 patients (41.6%). Results according to temporal biopsy are shown in table 2. Ultrasound had an overall sensitivity of 29%, and a sensitivity of 25% and a specificity of 84% versus temporal biopsy; it helped in diagnosing 2 patients with negative biopsy and 6 patients without biopsy.

Conclusion: Abnormal temporal pulse, PMR symptoms and age were associated with a positive biopsy. Clinical presentation, course, treatment and relapses/recurrences didn't differ between patients with positive or negative or unperformed biopsy. In our experience, clinical judgement continues to be relevant in the diagnosis of GCA, aided partially by biopsy, less so by ultrasound.

Table 1. Patients' characteristics according to temporal biopsy results

	Total of GCA patients (n=101)	Positive temporal biopsy (n=52)	Negative temporal biopsy (n=21)	Temporal biopsy not performed (n=28)
Females, n (%)	79 (78.2)	39 (75)	24 (86)	22 (79)
Age at diagnosis, average, years (SD)	74.9 (8)	76 (6)	71 (9)	76 (10)
Follow-up, median, years (IQR)	2.6 (2.3)	3.2 (2.7)	2.2 (3.6)	2 (2.1)
Headaches, % (CI 95%)	80.2 (72-88)	78.8 (67-90)	71.4 (51-91)	88.8 (77-101)
Ocular symptoms, % (CI 95%)	45.5 (36-55)	48 (34-62)	48 (25-70)	39 (21-58)

- Blurred visión, %	20.8 (13.3-30)	28.8 (17.1-43.1)	9.5 (1.2-30.4)	14.3 (4-32.7)
- Diplopia , %	8.9 (4.1-16.2)	5.8 (1.2-15.9)	9.5 (1.2-30.4)	14.3 (4-32.7)
- Partial or total blindness, %	15.8 (9.3-24.4)	13.5 (5.6-25.8)	28.6 (11.3-52.2)	10.7 (2.3-28.2)
Jaw claudication, % (CI 95%)	39.6 (30-49)	48 (34.2-62)	28.5 (8.5-48.6)	33.3 (15-52)
Polymyalgia Rheumatica, % (CI 95%)	44.6 (35-54)	53.8 (40-68)	33.3 (12-54)	33.3 (15-52)
Temporal tenderness, % (CI 95%)	63.4 (53.2-72.7)	65.4 (50.9-78)	57.1 (34-78.2)	64.3 (44.1-81.3)
Abnormal temporal pulse, % (CI 95%)	43.6 (33.7-53.8)	55.8 (41.3-69.5)	9.5 (1.2-30.4)	46.4 (27.5-66.1)
Temporal tenderness or abnormal temporal pulse, % (CI 95%)	63.4 (54-73)	65 (52-79)	57 (35-79)	64 (46-82)
Erythrocyte sedimentation rate at diagnosis, mm/1 h, average (SD)	67 (31)	71(27)	68 (30)	60 (36)
Methylprednisolone IV pulse, n (%)	15 (14.9)	10 (19,2)	4 (19,1)	1 (3,6)
Initial oral meprednisone dose, mg/day, average (SD)	42.8 (16.9)	44 (20)	42 (14)	39.9 (12)
Duration of treatment, months, median (IQR)	19 (25)	22 (19)	18 (31)	14 (18)
Relapses while tapering steroids, n (%)	31 (30.7)	17 (32.7)	7 (33.3)	7 (25)
Recurrence after finishing initial treatment, n (%)	7 (6.9)	4 (7.7)	1 (4.8)	1 (3.7)
Use of other immunosuppressive agents, n (%)	11 (10.9)	7 (13.5)	3 (14.3)	1 (3.6)

Table 2. Ultrasound findings according to temporal biopsy result

	Total of GCA patients with temporal Doppler ultrasound performed (n=42)	Positive temporal biopsy (n=16)	Negative Temporal Biopsy (n=13)	Temporal biopsy not performed (n=13)
Ultrasound with halo sign	12 (28.6%)	4 (25%)	2 (15.4%)	6 (46.2%)
Ultrasound without halo sign	30 (71.4%)	12 (75%)	11 (84.6%)	7 (53.8%)