



Biopsy Proven Juvenile Giant Cell Arteritis

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Clinical Image

A 35-year-old woman was examined for slight tenderness and dysaesthesia in the right temporal region. The woman reported temporary blurred vision which resolved spontaneously. On inspection the temporal artery was slightly prominent and tender, soft to the touch.

A brain-CT scan, thoracic vessels MRI, acute phase reactants, ultrasound on the temporal artery, and PET/CT scan, resulted negative. ANCA antibodies were negative as well. After symptoms persisted a 3 mm biopsy was carried out and resulted normal. The first sample was considered possibly insufficient and a second 12 mm sample was taken. This showed acute thrombosis, rich eosinophil infiltrate, and predominantly lymphoplasmacytic inflammation with adventitial lymphoid follicles, allowing a diagnosis of juvenile giant cell arteritis (JGCA). During the follow-up the patient was well without any treatment, confirming the diagnosis of JGCA.

In contrast with classical Giant Cell Arteritis (GCA) the age of onset in JGCA is less than 40-50 years old, cranial and systemic signs of vasculitis are lacking or only mild and intense eosinophil infiltration is a histological feature.

JGCA has been classified as isolated or associated with Churg-Strauss, Panarteritis nodosa or Thrombangiitis obliterans. It could also be a form of Kimura's disease or angio-lymphoid hyperplasia with eosinophilia.

JGCA is a localized vasculitis which does not require corticosteroids. As with the patient in this case, among sixteen patients with pure JGCA, surgical excision resolved symptoms in twelve of them. Two patients required prednisone, and one patient required tocopherol nicotinate (Figure 1) [1].

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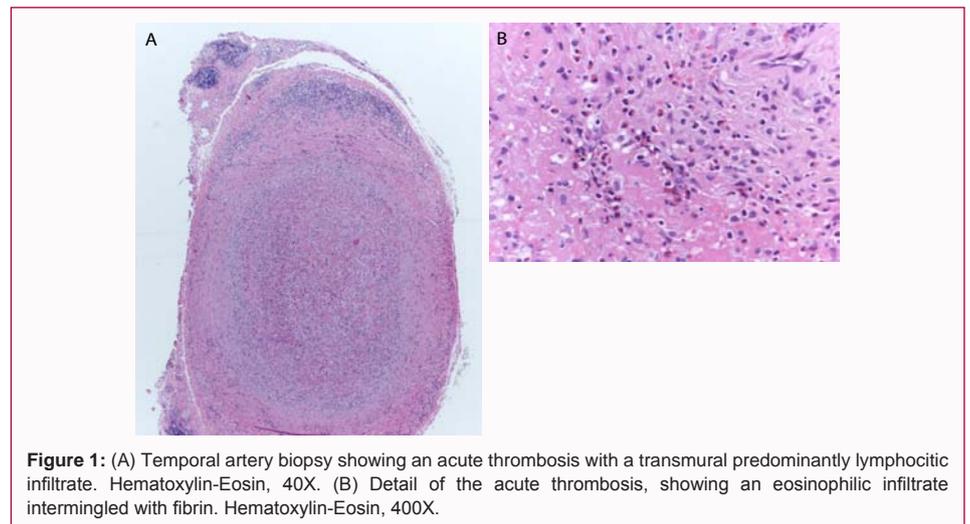


Figure 1: (A) Temporal artery biopsy showing an acute thrombosis with a transmural predominantly lymphocytic infiltrate. Hematoxylin-Eosin, 40X. (B) Detail of the acute thrombosis, showing an eosinophilic infiltrate intermingled with fibrin. Hematoxylin-Eosin, 400X.

References

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