Brainstem encephalitis as the onset of Behçet disease

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Background

Behçet disease is often regarded as the manifestation of recurrent orogenital aphtosis, together with lesions such as pseudofolliculitis, eritema nodosum, uveitis and a positive result of the patergy test. However, its clinical onset may be extremely variable, thus making the diagnostic evaluation a difficult challenge for clinicians. Central nervous system involvement in Behçet disease can affect both the meningeal structures and the cerebrospinal tissue, sometimes mimicking infectious diseases or neoplastic lesions; an accurate differential diagnosis is therefore mandatory, in order to provide adequate treatment. We describe an Italian female patient, aged 42, who received a diagnosis of Behçet disease after the onset of brainstem encephalitis.

Case report

P.A. Female aged 42. Taken to the Emergency Department due to progressive headache and vertigo, right hemiparesis and slurred speech. CT scan revealed a pontine hypodensity. Her medical hystory was relevant for recurrent, self-limiting manifestations of orogenital ulcers.

Cerebrospinal fluid analysis detected mononuclear leucocytosis (62 cells/mL), without evidence of viral or bacterial infection, nor oligoclonal bands. MRI showed a non specific brainstem lesion (figure 1) with irregular contrast enhancement and FLAIR hyperintensity.

Ophtalmological evaluations detected right retinal vasculitis (figure 3), togehter with lymphocytic infiltrate at the skin biopsy (figure 4).

Steroids were administered, with clinical recovery and progressive resolution of the lesion on MRI follow-up scans (figure 2). Without steroid medications, orogential ulcers were reported again.

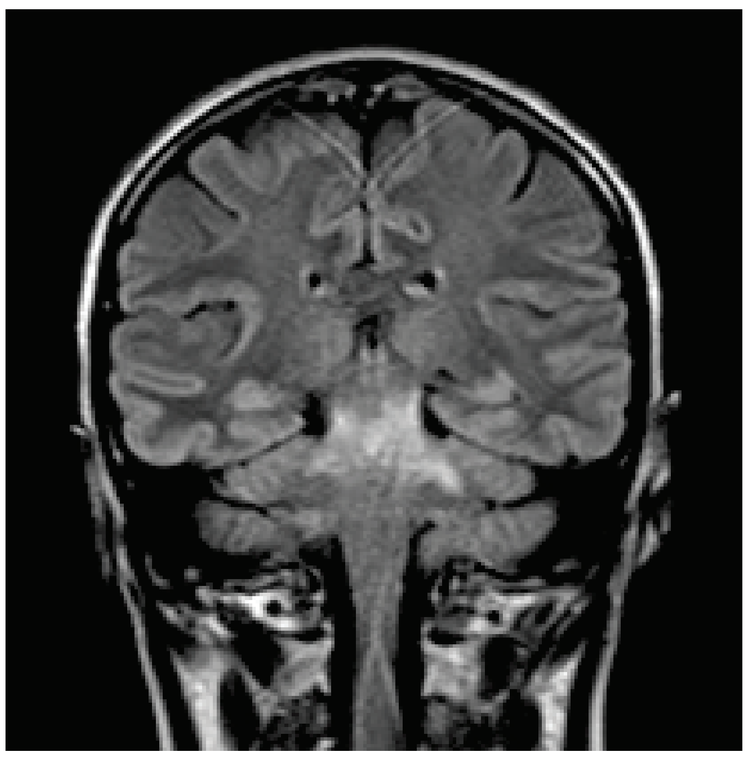

Figure 1: Admission MRI scan (FLAIR sequence, 05/2015) showing a massive pontine hyperintensity.

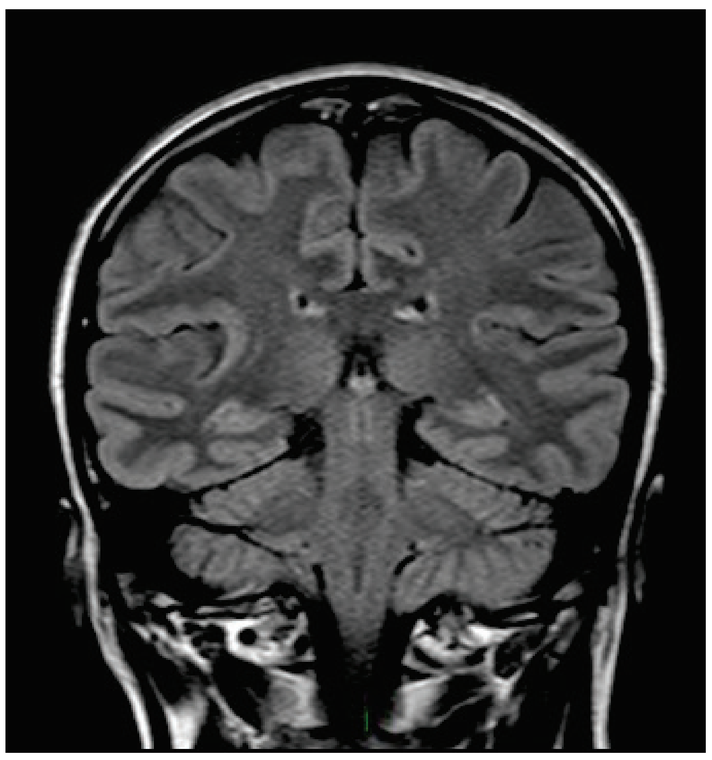

Figure 2: Follow-up MRI scan (FLAIR sequence, 09/2015), without evidence of the previous lesion.

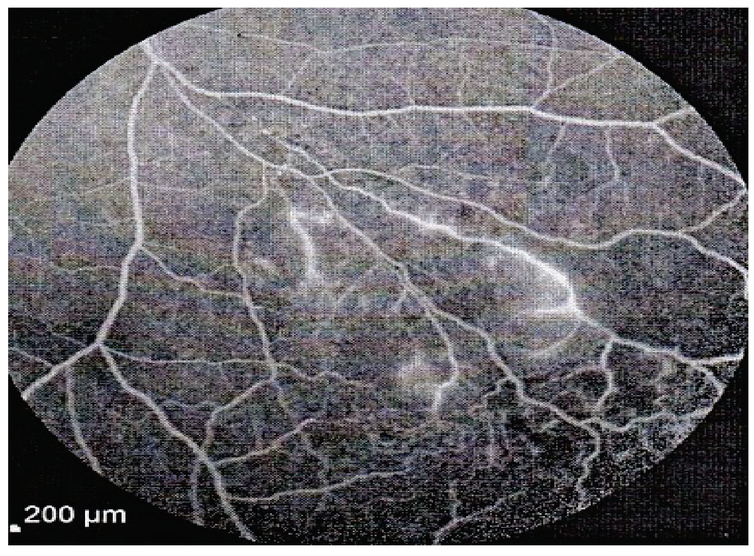

Figure 3: right-eye fluoroangiography showing contrast leakage in the infero-nasal rentinic field.

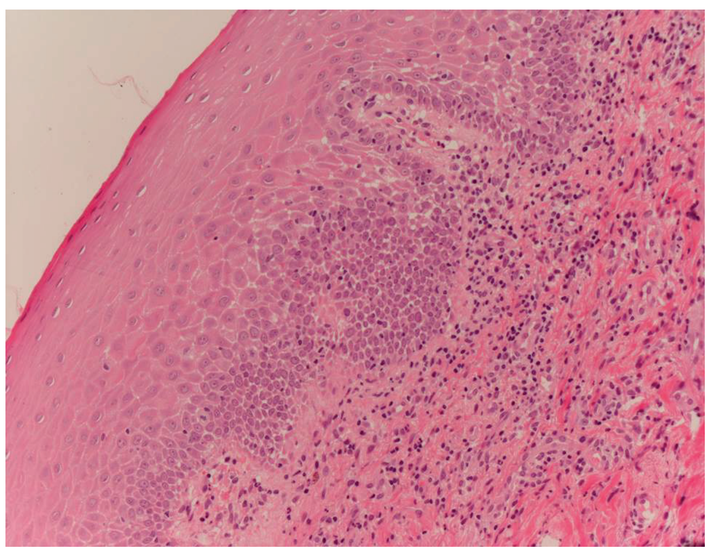

Figure 4: Oral mucosa with lymphocytes accumulation in the connective layer and lymphocytes infiltration in the epithelial layer.

Results

On the basis of clinical, radiological, histological findings and the absence of liquoral oligoclonal bands (2), according to the available diagnostic criteria (1), the diagnosis of Behçet disease was made. Azathioprine 100 mg a day was started.

Conclusions

Neuro Behçet is a rare but potentially devastating manifestation of Behçet disease. Its recognition and treatment can be difficult, due to the lack of controlled trials (3). Clinical descriptions and trials are still needed, in order to improve the disease understanding and management.

References

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2. Saruhan-Direskeneli G, Yentur SP, Mutlu M et al. Intra-thecal oligoclonal IgG bands are infrequently found in neuro-Behcet's disease. Clinical and Experimental Rheumatology 2013; 31(3 Suppl 77):25–27.
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