Title

Behçet's syndrome masquerading as tumor.

Journal

Neuroradiology, 29(1)

ISSN

0028-3940

Authors

Litvan, I

Roig, C

Rovira, A

et al.

Publication Date

1987

Peer reviewed

Case note

Neuroradiology
© Springer-Verlag
(1987) 29: 103

Behcet's syndrome masquerading as tumor

I. Litvan[1], C. Roig[1], A. Rovira[2], and J. Ruscalleda[2]

Departments of Neurology[1] and Neuroradiology[2], Hospital Santa Creu i Sant Pau, Barcelona, Spain

History and clinical findings

Behcet's syndrome, with neurological symptoms mimicking an expanding cerebral mass lesion is rare although a few cases reported recently have had identifiable thalamic lesions [1, 3].

A 51-year-old male with a 30-year history of recurrent oral and genital ulcers, arthritis and conjunctivitis was admitted to another hospital in November of 1983, with progressive symptoms of headache, vomiting, right homonymous hemianopia, and hemihypoesthesia. Blood analysis showed a slight elevation of the sedimentation rate and leukocytosis. CT scan showed (Fig. 1) a left parieto-occipital lesion of diminished density with marked contrast enhancement and mass effect. Clinical symptoms promptly disappeared after treatment with steroids, and a repeated CT scan 1 month later showed a small necrotic residual lesion. Lymphoma, sarcoidosis, and bacterial infection were discarded as probable diagnoses after extensive negative laboratory, radiological and bone marrow studies.

Seven months after steroid withdrawal, the patient was admitted to our institution with subacute symptoms, including partial complex seizures of occipital origin, right homonymous hemianopia, visual agnosia, alexia without agraphia, and prosopagnosia. In addition, he presented with low-grade fever and genito-oral ulcers. Blood analysis, virus serology, bacteriological and fungus cultures were negative. There was a moderate elevation of proteins and a slight elevation of the cell count in the cerebro-spinal fluid, but the cultures and immunologic studies yielded negative results. Arterial digital substraction angiography showed a discrete "blush" in the left temporo-occipital arteries, but did not show any venous thrombosis or mass effect. A control CT scan and MRI (Fig. 2) showed a left parieto-occipital necrotic area.

Discussion

This patient met the four clinical criteria (oro-genital ulcers and oculo-dermatological lesions) and two minor clinical criteria (neurologic and arthritic) for the diagnosis of complete Behcet's syndrome. The clinical evolution of this patient included remission with steroid treatment.

In our review of the literature on neuro-Behcet's syndrome we found three types of vascular involvement: venous thrombosis [4]; avascular mass [1]; and vascular distortion [5]. We identified several case reports with documented CT scans showing unilateral hypodense thalamic lesions with or without contrast enhancement [1, 3]. Hypodense lesions of the pons [3] and cerebral hemisphere [5] with contrast enhancement have also been reported. Our case discloses radiological signs of an expanding mass lesion that was unlike any of the other cases previously reported in the literature in terms of location and size. Pseudotumoral presentation in Neuro Behcet's syndrome is extremely rare but considering that 5% of cases have neurological symptoms [1], we urge that it should be included in the differential diagnosis of expanding masses.

References

1. Rozin F, Haughton V, Bernhard G (1977) Neurology 27: 1148–1152
2. Williams A, Haughton V, Saxena V, Albers J (1979) Radiology 131: 403–404
3. Dobkin B (1980) Arch Neurol 37: 58–59
4. Harper MC, O'Neill BP, O'Duffy DJ (1985) Mayo Clin Proc 60: 419–422
5. O'Duffy D, Goldstein N (1976) Am J Med 61: 170–178

Received: 29 November 1985

J. Ruscalleda MD
Department of Neuroradiology
Hospital Sant Pau
San Antoni M Claret 167
08025 Barcelona
Spain

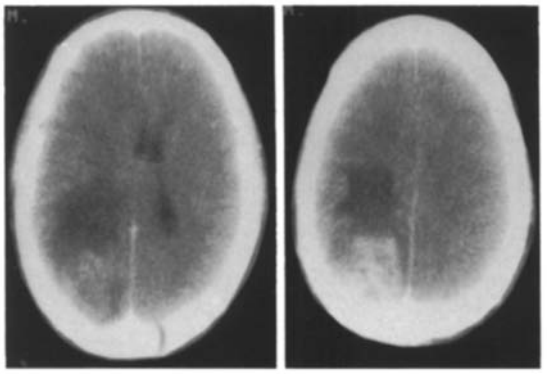

Fig. 1. Initial CT scan after contrast enhancement disclosed a cortical nodular mass simulating a tumor

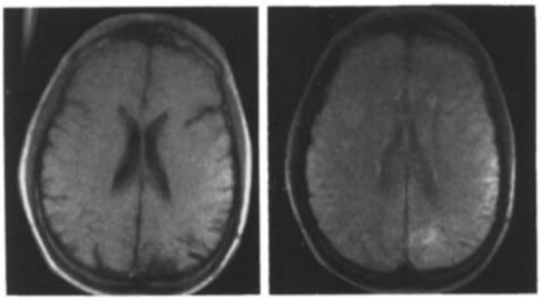

Fig. 2. MR made two years later, disclosed the morphology and location of the lesion with protons density; and increased brilliance with T2 confirmed a residual ischemic lesion