Unusual Manifestations Revealing Behçet's Disease: A Study of 43 Cases.

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Introduction:

Behçet's disease (BD) is a vasculitis with venous tropism affecting young individuals in countries bordering the eastern Mediterranean. Cutaneous and mucosal manifestations in this pathology occur in 70 to 90% of cases and are often the initial presentation of the disease, alongside ocular, neurological, or vascular involvement. Other unusual manifestations may also reveal this disease. In this study, we will investigate these manifestations in patients with BD.

Patients and Methods:

This is a retrospective monocentric study of BD cases selected according to the international diagnostic criteria of the International Study Group conducted over an 18-year period from 2006 to 2023.

Results:

Among a series of 563 patients followed for BD, 44 cases, accounting for 7.8% of cases, had an unusual mode of presentation. Acute fever was indicative in 21 patients, accounting for 47.7% of cases. In this group of patients, infectious workup was negative with marked inflammatory syndrome. BD presented in its benign cutaneous-mucosal form in 2 cases, in an articular form in one patient, and in the severe form involving the eyes, neurological system, vessels, and digestive system in the other 18 cases. It included panuveitis in 1 patient, thrombotic event in 7 cases such as lower limb venous thrombosis (3 cases) and pulmonary embolism (2 cases), Budd-Chiari syndrome (3 cases), cerebral venous thrombosis in one case, Behçet's enteritis complicated by severe digestive hemorrhage in one case, and central parenchymal neurological involvement in 7 patients. Prolonged fever was indicative in 15 patients, accounting for 34% of cases, and was correlated with severe vascular involvement in 8 cases: pulmonary artery aneurysm associated with pulmonary embolism (5 cases), intracardiac thrombus (2 cases), and thrombosed coronary artery aneurysm (1 case). Central parenchymal neurological involvement was found in 6 patients, and benign articular involvement in 1 case.

Orchiepididymitis was indicative in 6 patients, accounting for 13.6% of cases.

Parotidomegaly associated with dry mouth syndrome revealed BD in only 1 patient, accounting for 2.3%. It was correlated with severe vascular involvement, and lip biopsy concluded to AA-type amyloidosis.

Stage III papilledema associated with signs of intracranial hypertension revealed BD in only one patient, accounting for 2.3%, and was correlated with retrobulbar optic neuritis.

Discussion/Conclusion:

Although Behçet's disease is primarily revealed by classical symptoms, it is crucial to recognize the existence of unusual manifestations that may reveal this pathology, such as fever evolving acutely or chronically, which may be correlated with severe systemic involvement. Other even rarer revealing manifestations were found in our series, such as papilledema, testicular involvement, and AA-type amyloidosis as a mode of revelation. These symptoms should be particularly sought after in regions with a high prevalence of the disease to ensure optimal management.