Neoplasia and Behçet's disease: Rare case of association of Behçet's disease and multiple myeloma

S.MOURABIT,K.ECHCHILALI, M. BENZAKOUR, M. MOUDATIR, H. EL KABLI

internal medicine department - university hospital IBN ROCHD, Casablanca, Morocco

INTRODUCTION

Behçet's disease is a multisystemic inflammatory disorder characterized by recurrent acute attacks with a clinical and evolutionary polymorphism and whose pathogenesis remains obscure. Ocular involvement in Behçet's disease is classically manifested by non-granulomatous anterior uveitis, and posteriorly by hyalitis and vasculitis. We describe the case of a young patient with an atypical association of Behçet's disease and multiple myeloma.

OBSERVATION

A 34-year-old female patient, followed for Behçet's disease revealed by a bipolar aphthosis evolving for 4 years, admitted for a decrease of visual acuity and ocular redness of sudden onset. The ophthalmological examination showed bilateral episcleritis and Roth's spots on the fundus, suggesting retinal vasculitis. The extraocular signs were characterized by an episode of oral aphthosis, arthromyalgia, and altered general condition. The biological workup revealed an inflammatory syndrome, malignant hypercalcemia, renal failure with a GFR of 24ml/min, and a profound regenerative anemia of 4g/dl. An emergency myelogram showed 85% plasma cells, confirming the diagnosis of multiple myeloma. Plasma protein electrophoresis showed a monoclonal peak in gamma globulin at 30 g/l. Serum immunofixation identified a monoclonal IgG Kappa immunoglobulin. Serum free light chain assay showed an elevation of the Kappa chain level to 569.5mg/L and a Lambda chain level to 1.98mg/L with a pathological ratio of 287.63. The bone workup showed cookie cutter lesions on the skull X-ray and at the spinal level. The MRI showed a cervical-dorsal-lumbar myeloma with no signs of compression. The patient was put on polychemotherapy (bortezomib, dexamethasone and thalidomide) with bone marrow autotransplantation scheduled.

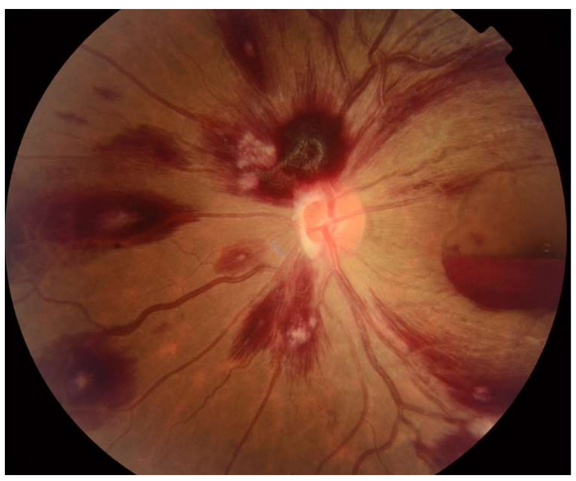
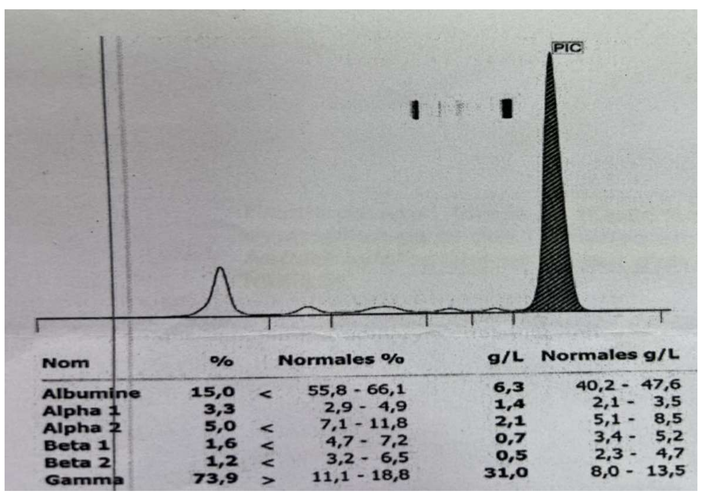


Figure 1 : roth spots



Nom	%		Normales %	g/L	Normales g/L
Albumine	15,0	<	55,8 - 66,1	6,3	40,2 - 47,6
Alpha 1	3,3		2,9 - 4,9	1,4	2,1 - 3,5
Alpha 2	5,0	<	7,1 - 11,8	2,1	5,1 - 8,5
Beta 1	1,6	<	4,7 - 7,2	0,7	3,4 - 5,2
Beta 2	1,2	<	3,2 - 6,5	0,5	2,3 - 4,7
Gamma	73,9	>	11,1 - 18,8	31,0	8,0 - 13,5

Figure 2 : important peak in gamma globuline

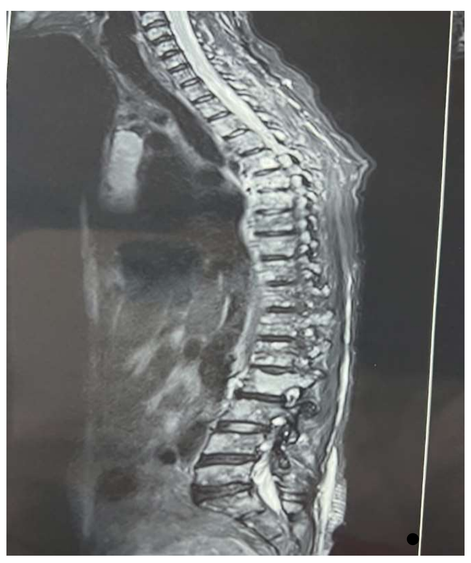


Figure 3 :cervical-dorsal-lumbar myeloma

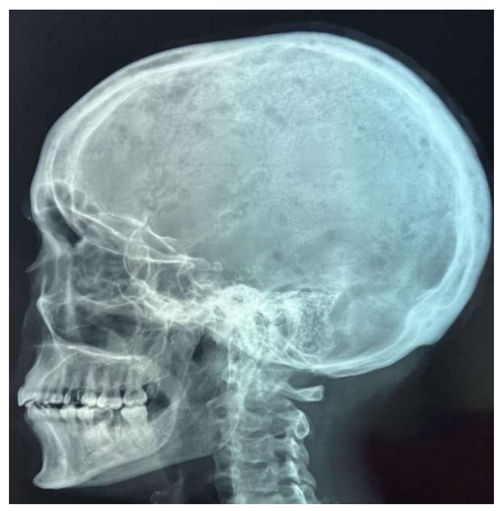


Figure 4: Cookie cutter lesion

DISCUSSION / CONCLUSION

Ophthalmologic involvement in Behçet's disease is very frequent, including retinal involvement, whereas ophthalmologic involvement in multiple myeloma is very rare but can affect all structures of the eye: orbital involvement (affecting the orbital muscles and the adnexa of the eye), intraocular involvement (retinal vasculitis, uveitis, chorioretinitis) and neuro-ophthalmologic involvement. The mechanisms that explain ophthalmologic involvement in myeloma are: either extra-medullary infiltration of tissues by monoclonal plasma cells; or a hyperviscosity syndrome secondary to monoclonal gammopathy. When retinal vasculitis revealed by Roth's spots is diagnosed, the etiological work-up should include a systematic search for monoclonal gammopathy that may reveal the existence of multiple myeloma.

And in conclusion, it is true that ocular involvement in the course of Behçet's disease is frequent and serious, dominated by uveitis and retinal vasculitis. Nevertheless, one must be wary when faced with a particular symptom, notably the presence of ROTH spots, or when one has other systemic manifestations, or when one is faced with a therapeutic failure in conventional therapies.