

Destructive Sinusopathy and Middle Ear Involvement in Behçet's Syndrome

A case report

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1. INTRODUCTION

Behçet's syndrome is a multisystemic disease with internationally agreed diagnostic criteria¹. Vascular, central nervous system (CNS) and gastrointestinal (GI) involvement have been documented². We report a first case with destructive sinusopathy and middle ear involvement on a 47-year-old Caucasian male, studied for recurrent oral ulcerations. Previously healthy, he suffered from recurrent oral ulcerations and dysphagia from 1995 to 1998. In 1998, nasal discharge and facial "congestion" started together with right side ear pain without hearing loss. During 1999 right-side repletion and hearing loss sensation began. In 2000-2001 the patient suffered a crisis with ear, nose and throat (ENT) symptoms, dysphonia, strength-loss and worsening of muco-cutaneous lesions; recurrent genital ulcerations and subcutaneous nodules. There was never eye, CNS or GI involvement.

2. DIAGNOSIS, INVESTIGATION AND DISCUSSION

Behçet's syndrome was diagnosed on the basis of the negative biochemical and immunological results, positive pathergy test, recurrent oral ulcerations, and the presence of both recurrent genital ulcerations and skin lesions. Additional periodontal disease and dysphonic laryngitis were

diagnosed (laryngoscopy showed glottis oedema and a 1-2 mm white lesion on the right ventricular band). On the basis of synchronous symptoms we investigated two associations: ear involvement with hearing loss and rhinitis with sinusopathy.

Rhinitis with sinusopathy was diagnosed based on rhinoscopy showing hyperaemia with no focal lesion and perinasal sinuses CT-scan images compatible with chronic pansinusopathy of ethmoido-maxilar predomination, with destructive characteristics (amputation of the conchae and sept perforation). ENT findings were: normal otoscopy; positive hearing tests for mixed conductive and sensory hearing loss of light degree on the left side and of moderate degree on the right side. CT scan of the temporal bones showed bilateral middle ear involvement; right side occlusion of the oval and round windows by liquid collection; no signs of cholesteatoma or of ossicle destruction, compatible with bilateral medial chronic otitis, particularly on the right ear. It is known that sensory hearing loss is common in Behçet's syndrome as part of neural involvement^{1,3} and that in several diseases, external, middle or inner ear structures are subject to immunological injury⁴. What seems strikingly new in this case is the existence of a chronic bilateral middle ear otitis, especially on the right ear, which could explain the conductive component of the patient hearing loss. CT-scan images, hearing tests and clinic show a good correlation, especially on the right side. We cannot yet prove that Behçet's syndrome is the cause but it seems acceptable that part of these complaints can be attributed to this non-sensory hearing loss component. To our knowledge there is no published material about destructive sinusopathy in Behçet's syndrome, and one of the few diseases that show these features is Wegener's granulomatosis which is excluded in this patient. Without a reliable histological marker for the nasal sinus biopsy histological evaluation, a clear documentation of the association between Behçet's syndrome and this entity will have to be postponed.

3. CONCLUSIONS

Dysphonic laryngitis caused by a documented lesion and correlated with a typical clinical picture was present in this case. Rhinitis with destructive sinusopathy caused by Behçet's syndrome is most probably present in this patient; given a reliable histological marker a nasal sinus biopsy may come to support this conclusion. Though middle ear involvement with chronic bilateral middle ear otitis is undoubtedly proved in this patient, no histological confirmation can be presented.

ACKNOWLEDGEMENTS

We would like to thank the patient and patient's family; A.C. Roque, Dr. A.G. Fonseca, Prof. P.F. Costa, Dr. J.P. Boléo-Tomé, Dr. T. Palma and Dr. L. Cuña for their contributions; Dr. H. Yazici for his expert opinions. Hospital Fernando Fonseca support is gratefully acknowledged. Contact: H.M.G. Martins (henriquemartins@hotmail.com).

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