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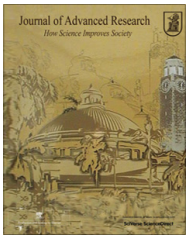

CASE REPORT

Development of a low grade lymphoma in the mastoid bone in a patient with atypical Cogan's syndrome: A case report

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ABSTRACT

Cogan's syndrome is a rare disorder characterized by ocular and audiovestibular manifestations in its typical form and caries a wide variety of atypical manifestations. It is considered as an autoimmune disease. We present the first case in the literature of a 67 year old woman with the development of low grade non-Hodgkin lymphoma (NHL) in the mastoid bone in a pre-existing history of atypical Cogan's syndrome. The anatomical development of NHL was to a "target" organ of Cogan's syndrome, which is the inner ear.

Introduction

We present a rare case of the development of low grade non-Hodgkin lymphoma (NHL) in the mastoid bone in a patient with an atypical Cogan's syndrome without progression of NHL and with symptomatic deterioration of Cogan's syndrome, responding only to TNF-a modulation.

Case presentation

A 67 year old female Caucasian patient from Greece presented in April 2003 with intermittent fevers up to 38 °C. Two months later she complained for additional persistent headaches, bilateral hearing loss, vertigo, tinnitus, and episodes of ataxia. Audiovestibular manifestations were classified as sensorineural

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deafness. In September 2003 she was admitted to the hospital where an extensive workup was negative except of a brain MRI which showed a presence of a mass lesion over the right mastoid outgrowth (Fig. 1A and B). She underwent surgical biopsy and histologic evaluation revealed a low grade B-cell non-Hodgkin's lymphoma (NHL) (ICD10:C85) (Fig. 2A and B). Staging evaluation proved to be of IB. She was managed with six cycles of chlorambucil till June 2004, with complete remission of the malignant lesion.

In April 2005 the patient referred to the ophthalmology department with main complains a severe impairment of visual acuity and ocular pain in both eyes. It is worth mentioning that medical history revealed that the patient had experienced episodes of mild visual disturbances during the last semester of 2003 and throughout 2004 overlooked by her. Quite long intervals between visits for follow up and management occurred because of patient poor compliance. In ophthalmologic examination Snellen visual acuity was found to be 0.2 on the right and 0.3 on the left eye; bilateral panuveitis (anterior chamber reaction and vitritis) along with papilledema and increased intraocular pressure in both eyes was diagnosed. Laboratory workup including intraocular fluid studies with PCR, cultures and flow cytometry was not diagnostic; elevated serum IgG titers against CMV were only found. Investigation for tuberculosis, syphilis and sarcoidosis was also negative. The patient was initially considered as a case of CMV associated uveitis treated with intravitreal injection of ganciclovir, cycloplegics, topical steroids and periocular steroid injections. Patient's ocular manifestations were markedly improved (Snellen visual acuity: 0.7 in each eye and remission of uveitis signs).

However, audiovestibular and institutional manifestations were gradually deteriorated and in June 2006 she was presented with deafness, arthritis, fever, anemia and skin rash whereas, neither oral aphthous along with genital ulceration were observed nor had been ever reported. Ocular manifestations were still under control. The clinical presentation mainly the audiovestibular and ocular manifestations was indicative of Cogan's syndrome in its atypical form. Full serum autoimmune profile (including antinuclear antibodies, anti-dsDNA antibodies and c-antineutrophil cytoplasmic antibodies) and infectious profile were negative, except for the presence of an IgG monoclonal protein band as well as for elevated erythrocyte sedimentation rate and C-reactive protein levels.

Due mainly to the continuous clinical deterioration of fever, fatigue, headache, skin rash and arthralgias led in November 2007 to the re-administration of chlorambucil and methylprednisolone for another six cycles. During the administration of methylprednisolone skin rash, fever and fatigue got better, only for a short period of time. The patient was practically deaf, with mild visual disturbances, fever, fatigue, malaise, symmetric polyarthritis and cutaneous manifestations. A cutaneous lesion biopsy revealed granuloma annulare. Systematic follow up was negative for NHL progression. The patient was managed from December 2008 till January 2009 with two cycles of cyclophosphamide, vincristine and methylprednisolone and from January till February 2009 with two cycles of rituximab without response.

In February 2009 patient's ocular disturbances recurred with ocular pain and markedly decreased visual acuity (Snellen visual acuity: 0.025 on the right and 0.1 on the left eye). Cytology of aqueous humor demonstrated inflammatory cells with the predominance of lymphocytes, findings suggestive of chronic active inflammation (uveitis). In the absence of progression of NHL disease and given the fact that our patient was getting worse she was administered infliximab, an anti TNF-a agent, as a third line treatment for Cogan's syndrome and systemic steroids. Ocular pain and visual acuity were improved (Snellen visual acuity: 0.2 on the right and 0.3 on the left eye) and inflammation regressed, while bilateral papilledema was still present (Fig. 3). Audiovestibular, general symptoms and skin manifestations were moderately improved.

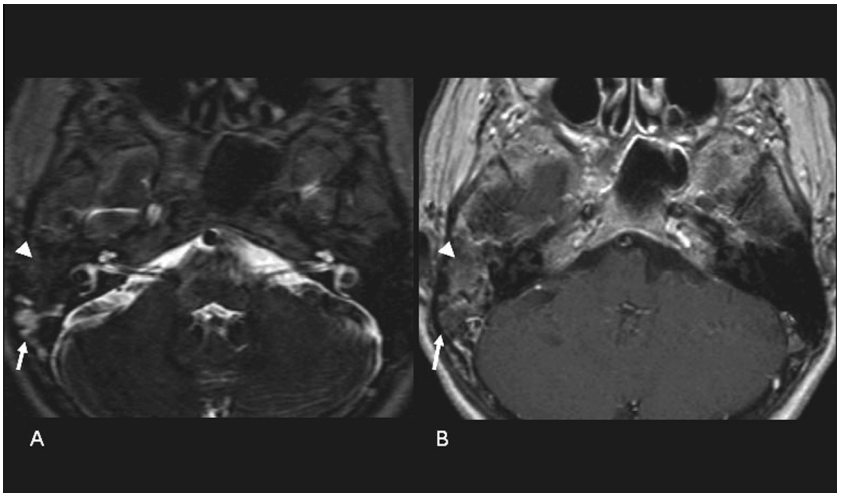

Fig. 1 (A) Axial T2-weighted scan (TR/4000 ms, TE/250 ms) demonstrating a low signal intensity tissue (white arrowhead) occupying a large part of the right mastoid. Mastoiditis at the periphery of the lesion appears with high signal intensity (white arrow). The inner ear components appear normal with the expected high signal. (B) Axial contrast enhanced T1-weighted scan (TR/500 ms, TE/20 ms) same level with (A) demonstrates an enhancing tissue (white arrowhead) occupying a large part of the right mastoid. Mastoiditis at the periphery of the lesion appears with intermediate signal intensity (white arrow). No contrast enhancement was observed at the inner ear.