DIDATIC IMAGE
LACRIMAL GLAND SWELLING IN A YOUNG AFRICAN WOMAN

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ABSTRACT

We describe an African patient with sarcoidosis involving the lacrimal glands. Gallium scintigraphy showed the so-called "panda sign".

KEY WORDS

Lacrimal gland, Mikulicz, Panda sign, Sarcoidosis

CASE REPORT

A previously healthy 25-year-old African woman presented with a 2 months history of bilateral swollen upper eyelids. The patient had no systemic complaints. She grew up in West-Africa and lived in Belgium 2 years.

Beside the pronounced but painless swelling of the lacrimal glands, physical examination revealed discrete bilateral swelling of both the parotid and submandibular glands (Figure). Visual acuity was 20/20 in both eyes. Fundus examination was unremarkable. Intra-ocular pressure, gum and Schirmer tests were within normal limits.

Blood analysis showed normal liver, biliary tract, pancreas, kidney, and thyroid functions. Inflammatory parameters (C-reactive protein, leucocytes) were within normal ranges. Rheumatoid Factor, antinuclear and antineutrophil cytoplasmic antibodies were negative. The total immunoglobulin dosage (19 g/L; normal value: 7-15), and serum angiotensin-converting enzyme level (85 U/L; normal value 15-68) were elevated. IgG4 dosage was within normal limits. The immublot for Sjögren syndrome antibodies was clearly positive for type A antibodies, for type B. Serologic testing for *Mycoplasma pneumoniae*, *Chlamydia pneumoniae*, *Treponema pallidum, Borrelia burgdorferi, Bartonella*, Human immunodeficiency virus, Cytomegalovirus, Epstein-Barr virus, Parvovirus, and was negative.

Echographic examination of the lacrimal and salivary glands showed hypoechoic areas with relatively high vascularisation. Computed tomographies of the neck, lungs and abdomen

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did not show any abnormality beside the enlarged glands in the facial region found by clinical examination. Gallium scintigraphy revealed increased bilateral lacrimal and parotid glands uptake (Figure). Radiogallium was not observed to be taken up by the lungs.

Biopsy of an enlarged lacrimal gland showed mononuclear cells. Culture for (myco)bacteria was negative. A labial biopsy was undertaken but unfortunately no salivary gland tissue was harvested.

The diagnosis of Mikulicz syndrome with underlying sarcoidosis was made and the patient was treated with high-dose intravenous methylprednisolone (125 mg per day for 10 days). The systemic corticosteroid treatment was tapered over several weeks to a maintenance dose of 16 mg per day to keep the inflammation under control. Methotrexate was tried as a steroid sparing agent but was discontinued because of severely impaired liver function tests.

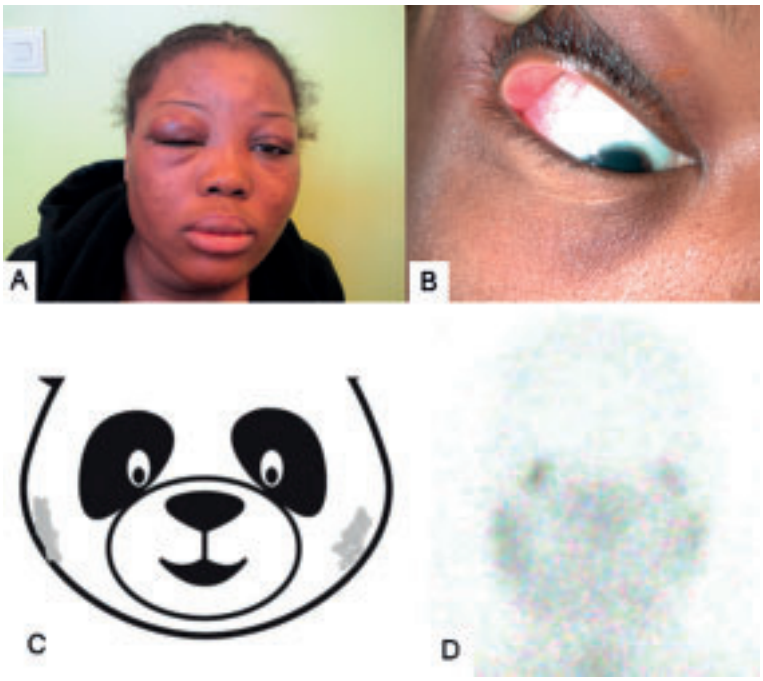

Fig.: A-B: swelling of the lacrimal glands; C-D panda sign: the panda's black nose correlates with the normal nasopharyngeal uptake; the black fur around the eyes with the intense lacrimal uptake; and the gray blush on the cheeks with the abnormal parotid uptake.

DISCUSSION

Mikulicz syndrome is a chronic condition characterized by the abnormal enlargement of the lacrimal and parotid glands. Other glands or tonsils in the head and neck may also be involved. (1) Although the disorder is almost always described as benign, it always occurs in association with another underlying disorder such as tuberculosis, leukemia, syphilis, Hodgkin's disease, Sjögren syndrome, lupus, IgG4 related disease (or Mikulicz disease), or sarcoidosis as in our case (2). Since the term "Mikulicz syndrome" is a confusing term describing heterogeneous pathology, it has been proposed to no longer use it and to only describe the underlying pathology (2). Sarcoidosis is a granulomatous disorder of unknown etiology typically affecting young adults (3). The disease is three to four times more common in blacks. Moreover, the disease presentation in this racial group tends to be more severe (3). The lung is the mostly affected organ (90%), but the disease may affect nearly all other body parts together or separately. The lacrimal and/or salivary glands are, in most cases asymptomatically, affected in approximately 5% of cases (4).

Definitive diagnosis of sarcoidosis is made by demonstration of non-caseating granulomas on tissue biopsy together with compatible clinical features (3). The combination of raised serum angiotensin convertin enzyme levels with abnormal gallium scanning a specific and sensitive tool (6). Nevertheless, clinical entities with a similar disease pattern have to be excluded before ultimately establishing diagnosis of sarcoidosis. For bilateral painless lacrimal gland swelling, this includes viral infections, tuberculosis, malignancy, malnutrition and alcoholism, acromegaly, diabetes melli-

tus, IgG4 related disease, and Sjögren syndrome (3). Although Sjögren syndrome antibodies in our case were elevated, the criteria from the American-European study group on the classification criteria for Sjögren syndrome were not fulfilled (7). These antibodies may in fact also be elevated in a number of other diseases, including sarcoidosis (3).

In summary, we report an unusual case of the panda sign on gallium scintigraphy. It is important for physicians to consider diagnosis of sarcoidosis in any patient presenting with bilateral lacrimal and/or parotid glands swelling.

REFERENCES

(1) Mikulicz JH – Uber eine eigenartige symmetrische Erkrankung der Tranen- und Mundspeicheldrusen. Beitr. Chir. Fortschr. Stuttgart, 1892; 610.
(2) Ihrler S, Harrison JD – Mikulicz's disease and Mikulicz's syndrome: analysis of the original case report of 1892 in the light of current knowledge identifies a MALT lymphoma. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2005; 100: 334-9.
(3) Thomas KW, Hunninghake GW – Sarcoidosis. JAMA 2003; 289: 3300.
(4) Mavrikakis I, Rootman J – Diverse clinical presentations of orbital sarcoid. Am J Ophthalmol 2007; 144: 769-775.
(5) Kurdziel KA – The panda sign. Radiology 2000; 215: 884-85.
(6) Rothova A – Ocular involvement in sarcoidosis. Br J Ophthalmol 2000; 84 110-116.
(7) Vitali C, Bombardieri S, Jonsson R et al – Classification criteria for Sjogren's syndrome: A revised version of the European criteria proposed by the American-European Consensus Group. Ann Rheum Dis 2002; 61: 554-558.

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