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CASE REPORT

MIKULICZ'S SYNDROME

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A rare case of Mikulicz's syndrome with underlying tuberculosis in a 65-year-old male with bilaterally symmetrical, asymptomatic enlargement of parotid, submandibular and lacrimal glands is reported.

Key Words : Mikulicz's syndrome, Mikulicz's disease

Introduction

In 1888 Mikulicz described a syndrome consisting of enlargement of the parotid, submandibular and lacrimal glands.[1] Mikulicz's disease and Mikulicz's syndrome produce the same clinical picture. Mikulicz's disease is due to an autoimmune process in the glands themselves and is generally looked upon as a clinical variant of Sjögren's syndrome.[2] Mikulicz's syndrome is a broad nonspecific term incorporating a wide variety of diseases[3] like sarcoidosis, lymphoma, leukemia, tuberculosis, syphillis, lead or iodide poisoning, gout,[4] lupus erythematosus,[5] recurrent parotiditis of unknown aetiology and actinomycosis.[1]

Case Report

A 65-year-old farmer presented with bilaterally symmetrical, asymptomatic enlargement of the parotid and submandibular glands (Fig. 1) of 2 years duration; and of the lacrimal glands of 3 months duration. He also had pain and photophobia in both eyes, dryness of the throat, breathlessness, productive cough and evening rise of fever all since 2 months.

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Systemic examination revealed generalised, mobile, nontender lymphadenopathy (inguinal, femoral, axillary and cervical), bilateral crepitations, hepatomegaly (2 cm below the subcostal margin) and ichthyosis. Ophthalmological examination showed mild proptosis; supercillary madarosis with eyebrow elevation; a fusiform, 3x2 cm, nontender, firm lacrimal gland swelling (Fig. 1);

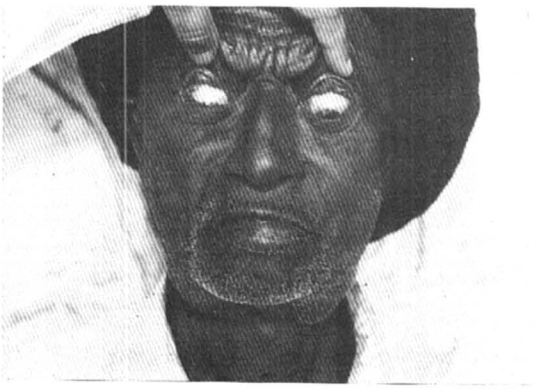

Fig. 1. Symmetrical enlargement of parotid, submandibular and lacrimal glands.

mechanical ptosis with poor levator palpebrum superioris function; superficial punctate keratitis with deficient precorneal tear film; normal anterior chamber, iris and pupil; bilateral immature senile cataract; and primary open angle glaucoma.

Investigations revealed microcytic hypochromic anaemia; lymphocytic leucocytosis; raised ESR; positive AFB in

the sputum; bilateral consolidation with left sided pleural effusion; positive Mantoux test; positive Schreimer's test; normal skiagram of orbit; and negative peripheral blood smear, blood VDRL and LE cell phenomenon. Patient refused to undergo a glandular or lymph node biopsy.

Discussion

Mikulicz's disease consists of (1) symmetrical enlargement of all the salivary glands, (2) narrowing of the palpebral fissures due to enlargement of the lacrimal glands, and (3) parchment-like dryness of the mouth.[2] It is characterised by a dense infiltration of lymphocytes, occasionally arranged in follicles, throughout the salivary tissues, accompanied by atrophy and disappearance of acinar tissue. The more popular and modern term for these lesions is 'benign lymphoepithelial lesion'. The diffuse lymphocytic infiltrate suggests the possibility of an autoimmune disease.[6]

The highest incidence of the disease is in patients between 31 and 40 years of age.[6] The onset may be sudden, but is usually painless and of long duration.[5] While several of or all the major salivary glands may be involved, a single parotid gland is the most frequent site (80%).[6] It appears resistant to treatment and is progressive.[3]

The peculiarities of this rare case of Mikulicz's syndrome were late presentation; presence of enlargement of parotid, submandibular and lacrimal glands of much longer duration than the chest complaints, and the underlying aetiology of tuberculosis.

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