Original Research Paper Medical Science

# A Case of Mikulicz's Disease


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**ABSTRACT**

*Mikulicz's disease is a rare, bilateral, symmetrical, painless, idiopathic, swelling of lacrimal and major salivary glands. It was considered as a subtype of Sjogren's syndrome previously but latest reports show evidence of differences between the two. Mikulicz's disease is considered as idiopathic whereas Mikulicz's syndrome is found to be associated with TB, sarcoidosis, lymphoma or leukemia. We report a case of Mikulicz's disease in a 34 year old female with bilateral enlargement of lacrimal and parotid glands. There was no associated systemic conditions and a good response to corticosteroids was observed.*

**KEYWORDS : Mikulicz's disease, Sjogren's syndrome, lacrimal glands.**

**CASE REPORT:**

A 34 year old female reported to our out patient department with complaints of bilateral enlargement of lacrimal and parotid glands which was painless and symmetrical for a duration of one month. She also had associated fever for 4 days. There was no history of previous similar complaints. On ocular examination, a swelling was noted in lateral aspect of upper lid giving an S shaped deformity with mechanical partial ptosis. On eversion of upper lid, enlarged lacrimal gland was visualized (figure 1). Ocular surface appeared normal with normal tear film meniscus. Anterior and posterior segment examination using slit lamp bio-microscopy and 90D was normal. Visual acuity in both eyes were 6/24 with astigmatic correction improved to 6/6. Schirmer's 1 showed 6 mm at 5 minutes and schirmer's 2 was recorded as 12 mm at 5 minutes showing mild decrease in tear secretion. TBUT was found to be 8 seconds. A lacrimal gland biopsy was done which showed diffuse lymphocytic infiltration (figure 2).

**Figure 1:clinical photograph of parotid (a,b) and lacrimal (c,d) gland enlargement.**

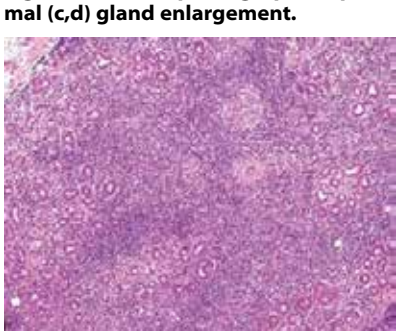

**Figure 2: HPE of lacrimal gland with lymphocytic infiltration.**

Bilateral parotid gland enlargement was noted with dryness of mouth. Systemic examination was done to rule out associated conditions. Peripheral blood smear showed no abnormal cells and differential count showed lymphocytes 50%. Blood sugar was normal(FBS-76mgs, PPBS- 123mgs) Mantoux was negative, chest X ray was within normal limits. Serum ACE was 10microlitres. Thyroid profile was within normal limits (FT4 =1.15 ng/dl , TSH =1.784mIU/L ). Blood immunology showed negative anti SS-A, SS-B, and ANA. MRI was done which showed smooth, homogenous, diffuse and nearly symmetrical enlargement of bilateral lacrimal and parotid glands with homogenous enhancement (figure3)

**Figure 3: MRI showing lacrimal gland enlargement.**

The patient was started on oral prednisolone 1 mg/kg/day OD for a week which was tapered as 10mg/week, along with topical lubricants 4TID. Steroids were continued as 10mg/day for a month and stopped .Plenty of oral fluids was advised. After 6 weeks the swelling disappeared and tear film parameters improved. TBUT was 15 sec and schirmer 1 was 13mm, schirmer 2 -15mm at 5 minutes.

**Discussion**

In 1888, Johann von Mikulicz-Radecki reported a case of bilateral, painless, symmetrical swelling of lacrimal, parotid and submandibular gland. Schaffer in 1927 described Mikulicz's disease if it was idiopathic

and Mikulicz's syndrome if there was associated lymphoma, sarcoidosis or TB.

Mikulicz's disease is a rare, bilateral, symmetrical, painless, idiopathic, benign lymphoepithelial lesion of lacrimal and major salivary glands. It was considered as a subtype of Sjogren's syndrome previously, but latest reports show evidence of differences between the two. Diagnosis was based the following- 1. Symmetrical / persistent enlargement of more than two lacrimal glands and major salivary glands. 2. Prominent mononuclear cell infiltration of lacrimal and salivary glands. 3. Exclusion of other diseases with glandular swelling such as sarcoidosis and lymphoproliferative diseases.

It is more common in mid adult age group and 60-80% distribution among females. MD has a normal ocular surface with retention of some lacrimal gland function with preservation of reflex tearing. HPE shows lymphocytic infiltration with lack of apoptotic features in acinar cells. There is a good response to glucocorticoids and recovery of gland function on treatment. There is an elevated Ig G4 in serum with prominent plasmacytes expressing Ig G4 in lacrimal and salivary glands. Anti SS-A and SS-B antibodies were negative. Complications of MD include autoimmune pancreatitis, tubulointerstitial nephritis, retroperitoneal fibrosis, Reidel's thyroiditis, autoimmune hypophysitis where Ig G4 was seen in pathogenesis.

Sjogren's syndrome exhibit intermittent swelling which might resolve even without treatment. Keratoconjunctivitis sicca is severe with squamous metaplasia of ocular surface . Inflammatory components like interferon gamma produced by the infiltrated lymphocytes worsen the ocular condition. Reflex tearing is poor. Acinar cell apoptosis is present with positive staining with APO 2.7 and salivary gland function donot recover on treatment. Anti SS-A and SS-B will be detectable.

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