Sarcoidosis appeared as Mikulicz’s syndrome: a Case report

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No conflict of interest

Written consent for use of the information in these case was obtained from the patient

Resume:
A 25-year-old woman with symmetrical bilateral swelling of the parotid, submandibular, and lacrimal glands - also called Mikulicz’s symptoms - showed erythema nodosum and arthralgia. Serum ACE was elevated. Chest CT showed hilar and mediastinal adenopathy and micronodules with lymphatic distribution. Flexible bronchoscopy showed a mucous membrane with granulations, the bronchoalveolar lavage was lymphocytic. Bronchial and labial biopsies showed tuberculoid granulomatous inflammation without caseous necrosis. The respiratory function tests were abnormal.

Based on these findings, the definitive diagnosis was Mikulicz’s syndrome associated with sarcoidosis. The administration of corticosteroid, caused parotid, submandibular, and lacrimal glands swelling to disappear also the respiratory function tests to be normal.

Keywords: Mikulicz syndrome; Salivary gland; Lacrymal gland; Granuloma; Sarcoidosis

Introduction:
Mikulicz Syndrome (MS) is a rare chronic condition characterized by the abnormal enlargement of glandular tissue in the head and neck. While this can be a benign self-limiting condition, other complex systemic diseases, such as sarcoidosis, may represent other underlying etiologies.[1]

We report a case of MS in a 25-year-old female that was found to be secondary to sarcoidosis

Observation:

A 25-year-old woman initially presented with a chief complaint of dry cough with facial swelling and inflammatory arthralgias of large joints. Historical evaluation was unremarkable for any systemic illness. The patient did not complain of pain, and her vision and tearing were not affected. No xerostomia was noted.

The examination was notable for bilateral parotid and submandibular glands swelling that were mobile and non-tender (figure 1). It was also noted that he had bilateral non-tender lacrimal gland swelling with ptosis (figure 2), which he stated had been increasing over a period of 3 months. Skin examination showed erythema nodosum and abdominal examination revealed hepatomegaly (14 cm liver arrow). No abnormalities of thoracic examination were present.

Chest X-ray showed bilateral hilar lymphoma with bilateral micronodular infiltrates (Figure 3). On Chest CT, there were hilar and mediastinal adenopathies (Figure 4) associated with lymphatically distributed micronodules with septal and nonseptal thickening (Figure 5). Cervical ultrasound confirmed hypertrophy of the parotid and submandibular glands without other abnormalities. Abdominal ultrasound revealed hepatomegaly without splenomegaly.

Flexible bronchoscopy showed whitish granulations, bronchoalveolar lavage was lymphocytic (lymphocytes: 30%), BK test was negative. The bronchial biopsies noted tuberculoid granulomatous inflammation without caseous necrosis.

The blood count showed a lymphopenia (870/mm3). The liver, kidney, blood and urine phosphocalcic tests, 24-hour proteinuria, tuberculin test, protein electrophoresis, HIV and HCV serology and antinuclear antibodies were negative. However, ACE was elevated to 191 IU/l (N: 20 and 70 IU/l). There was no uveitis. Rhinocavoscopy and nasal biopsy were unremarkable. Labial biopsy showed tuberculoid granulomatous inflammation without caseous necrosis.

Based on a radio-clinical and histological presentations, the diagnosis was sarcoidosis.

Her dry cough worsened and she developed dyspnea stage II on the mMRC scale.

Her plethysmography showed peripheral bronchial obstruction without restrictive ventilatory disorder, DLCO was 64%, there was a desaturation at 6 min TIM of 92% at 6 min, No abnormality was noted on arterial blood gas. cardiac ultrasound and electrocardiogram were unremarkable.

The treatment was corticosteroid therapy (dose of 1 mg/kg/day, decreasing by 5mg every 6 weeks), combined with adjuvant therapy.

After one year, the evolution was marked by regression of cough and dyspnea as well as arthralgias, disappearance of Mikulicz syndrome, hepatomegaly with normalisation of ACE (65 IU/l vs 191 IU/l). On the CT scan, there was a regression of adenopathies and micronodules (Figure 6).

Discussion:

Johann Mikulicz first presented Mikulicz Disease (MD) in 1888 at a meeting of the Society for Scientific Medicine in Königsberg. He later published a report of this sentinel case detailing a 42 year-old man with initial symptoms of lacrimal gland swelling followed by submandibular and parotid swelling [1]

Classic symptoms of MD are xerostomia and parotid enlargement. Dry eyes and lacrimal gland enlargement are also commonly reported symptoms. Parotid and lacrimal gland enlargement has been reported with both bilateral and unilateral distribution, in concurrence, or in isolation [2]
The term Mikulicz Disease should be reserved for cases that follow a benign course and are of unknown etiology. Mikulicz Syndrome should be used in cases with concomitant disorders [3].

Multiple etiologies account for Mikulicz syndrome that includes Sjogren syndrome, sarcoidosis, lymphoma, and tuberculosis.

Sarcoidosis usually presents with painless bilateral parotid enlargement. The imaging features of sarcoidosis are nonspecific but can show multiple non-cavitary masses representing enlarged intra-parotid lymph nodes. Alternatively, affected glandular tissue may be diffusely enlarged with homogenous enhancement [4]. The lacrimal gland is affected bilaterally and asymptomatically in 60- 87% of cases [5]. The lacrimal gland is affected bilaterally and asymptomatically in 60- 87% of cases [5]. Glandular enlargement is present in 8-32% of cases [5] and may be palpable as a painless bilateral superolateral mass. It is responsible for a dry syndrome in 0-42% of cases [5]. If the lacrimal gland is affected, sarcoidosis is systemic in the majority of cases [6]. Rapid regression is the rule under systemic corticosteroid treatment [7].

Sarcoidosis treatment depends on patient symptoms. Most patients (>75%) require only symptomatic therapy with NSAIDs. Approximately 10% need treatment for extrapulmonary disease and 15% are treated for persistent pulmonary disease. Oral prednisone given daily is the mainstay of treatment for people with chronic disease [7].

Mikulicz syndrome is a very rare mode of revelation of sarcoidosis, which is the particularity of our case

**Conclusion:**

Mikulicz Syndrome is a rare chronic condition characterized by inflammatory process of lacrimal, parotid and sub-mandibular glands and lymphocytic infiltrations. It rarely reveals the diagnosis of sarcoidosis.

**References:**


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