Mikulicz Syndrome

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Mikulicz syndrome is characterized by diffuse bilateral enlargement of lacrimal, parotid and other salivary glands. It can be secondary to many diseases like lymphoma, tuberculosis, leukemia, sarcoidosis and syphilis. We report a case of primary non-Hodgkin's lymphomapresenting withthe features of Mikulicz syndrome. As this entity is controversial, literature is reviewed with particular emphasis on historical perspective and recent developments.

Case Report

A 13 years old girl presented with 3 weeks history of painless swellings in both parotid and submandibular regions accompanied by facial asymmetry. There was no history of fever, weight loss, night sweats or recent upper respiratory tract infection or contact with a person with tuberculosis or having similar complaints. Apart from mild headache there was no histoiy of neurological symptoms. The patient was admitted three months prior to this presentation when she was referred from a peripheral hospital for work-up of anaemia. Complete blood counts had revealed pancytopenia with a white cell count of 1.8x10 9/liter, hemoglobin 30 grams/liter with normal red cell indices and platelet count of$35x10^9$/liter. There was no history of recent viral illness or exposure to drugs. Physical examination was normal apart from severe pallor. Autoimmune screening and serological studies for hepatitis B and C were negative. Bone marrow examination had shown severely hypoplastic marrow, without evidence of infiltration. She received supportive treatment in the form of antibiotics, blood andplatelet transfusions and steroids. While inthe hospital, blood counts started to improve and white cells and platelets rose to normal level within a week while haemoglobin started to improve rather slowly. The cause of this transient bone marrow suppression was presumed to be viral in origin. She was discharged on tapering doses of steroids. Physical examination during the current admission revealed symmetrical enlargement of both parotid, submandibular and lacrimal glands with left lower motor neurone 7th nerve palsy (Figure 1).

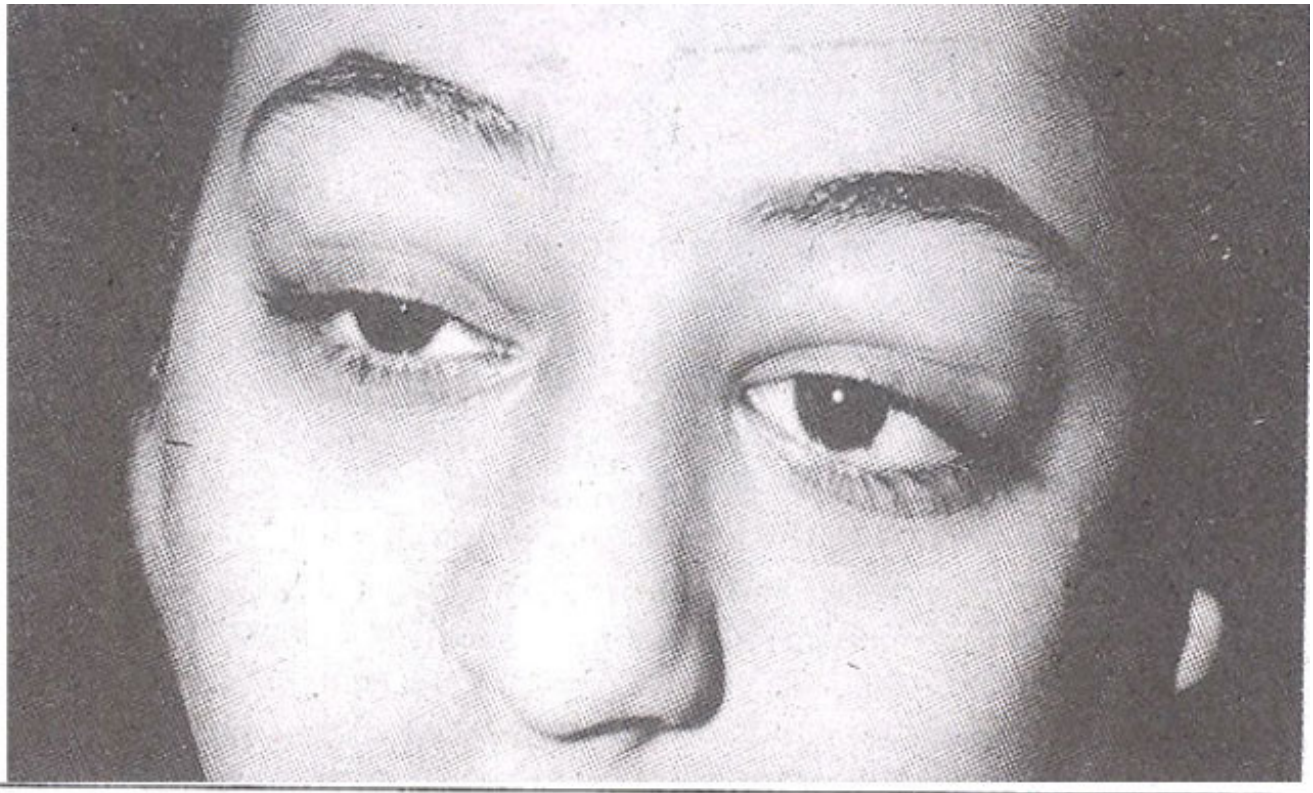

Figure 1. Picture of the patient showing bilateral enlargement of parotid and lacrimal glands.

There was no lymphadenopathy orhepatosplenomegaly, rest of the physical examination was normal. Initial investigations including complete blood count, liver function test, urea, creatinine, electrolytes, serum calcium, uric acid and chest x-ray were normal. There was a mild rise of alkaline phosphatase and serum lactate dehydrogenase above the normal value. Serology for hepatitis B, C, Epstein-Barr virus and HIV was negative as were antinuclear antibodies. A fme needle aspiration of the left parotid gland showed non-Hodgkin's lymphoma. While still in hospital the patient developed multi-palpable cervical lymph nodes. So a biopsy was carried out and confirmed the diagnosis of non-Hodgkin's lymphoma of diffuse large cell type (Figure 2).

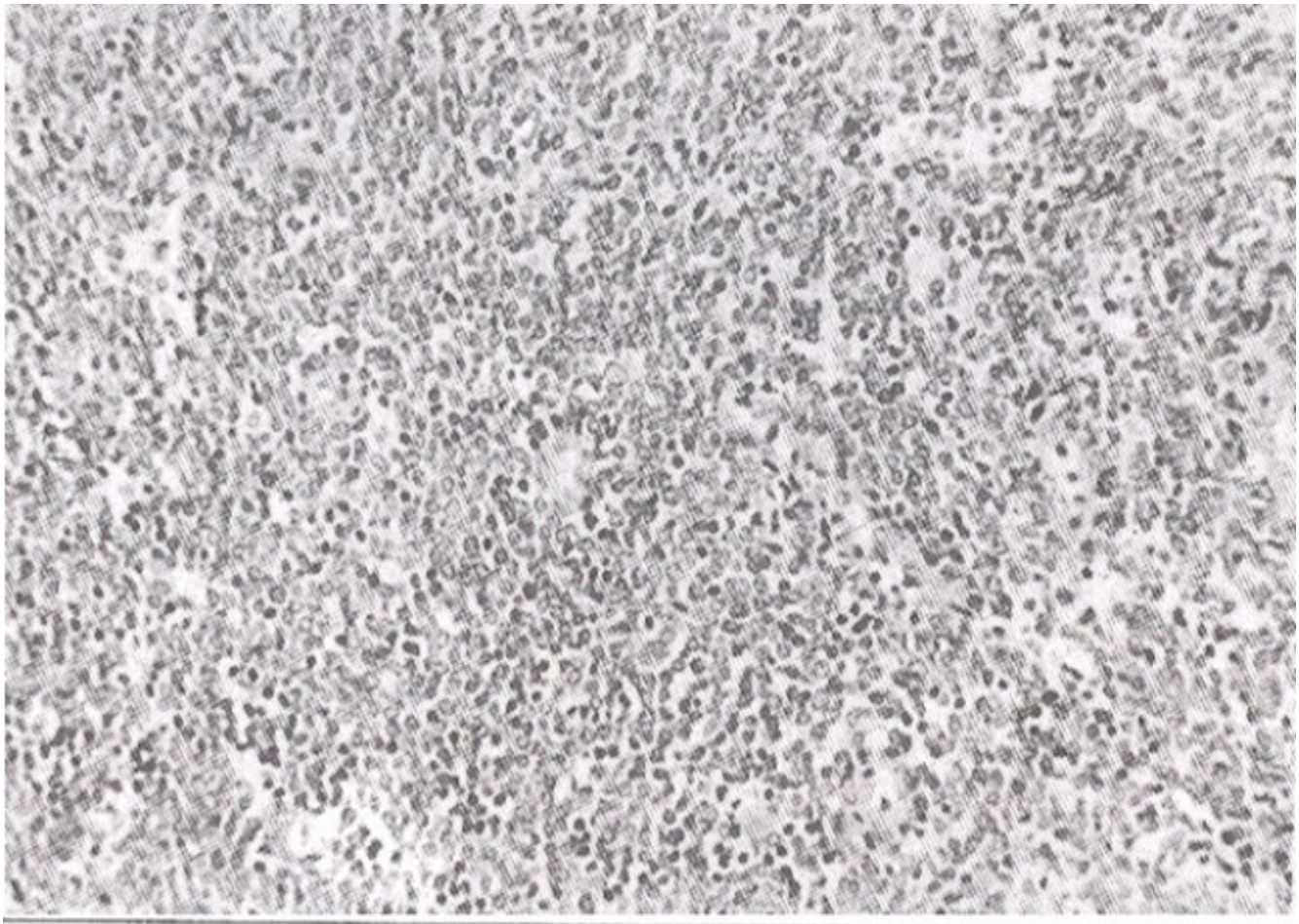

Figure 2. Photomicrograph of lymph node biopsy showing non- Hodgkin's lymphoma of diffuse large cell type (Hematoxylin and eosin, x100).

Staging procedures including CT-scan of chest and abdomen, bone marmw biopsy and whole body Gallium scan did not reveal any evidence of lymphoma in the rest of the body. Cerebrespinal fluid examination and CT-scan of brain were also normal. The patient was started on CNOP chemotherapy consisting of Cyclophosphamide 750 mg/m^2 intravenously (IV) ^{2}. Mitoxantrone 10 mg/m^2 IV, Oncovin(Vincristine) 1.4 mg/m^2 IV and Prednisolone 100 rng per oral daily for 5 days, (each cycle repeated at 3 weekly intervals). The 1st cycle of chemotherapy was followed by complete resolution of swellings in the neck and face. The facial nerve palsy also resolved without any residual defect. Central nervous system (CNS) prophylaxis (intrathecal chemotherapy) was considered but it was refused by the patient and her parents. The patient achieved complete remission after 8 cycles of chemotherapy. Three months later she presented with severe headache and right upper motor neurone 7th nerve palsy. Cerebro-spinal fluid examination showed large number of lymphoma cells. CT-scan of brain was still normal. Intraventricular chemotherapy consisting of methotrexate 10 mg,cytosine arabinoside 50 mgandhydrocortisone 50 mgwas initiated thmugh Omayya reservoir. A total of 10 cycles of intraventricular chemotherapy were required to achieve CNS remission. She however, died few months later due to progressive systemic and CNS disease.

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