

Mikulicz Syndrome - A Cytological Diagnosis in An Adolescent Male

Piyush Anand¹, Hiremath S S², Thippeswamy M T R²

¹ Post Graduate, ² Professor,

Department of Pathology, JJM Medical College, Davangere

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Abstract :

We report Mikulicz Syndrome diagnosed by fine needle aspiration cytology, in a 14 year old boy. He presented with dryness of mouth, difficulty to swallow solid food & bilateral non-tender parotid and lacrimal glands enlargement. Degenerated epithelial cells with lymphocytic attachment, mixed population of lymphoid cells, neutrophils and endothelial cells in a hemorrhagic background. Were characteristic of Mikulicz Syndrome along with lymphoepithelial islands. Thus aspiration cytology correlated with clinical features is an important tool for diagnosis of mikulicz syndrome.

Key words: Mikulicz syndrome, FNAC

Introduction:

Mikulicz syndrome refers to a rare, idiopathic, bilateral, persistent painless and symmetrical swelling of the lacrimal and salivary gland seen more commonly amongst women. It is found to be associated with prominent infiltration of plasma cells and lymphocytes. Though Salivary gland involvement is more common, other organ involvement has also been reported. Due to its histological similarities, it has been considered a subtype of Sjogren's syndrome.

Differential diagnosis include Extranodal Marginal zone B cell lymphoma, chronic sialadenitis, Warthins tumour and simple lymphoepithelial cyst.

Case Report: A 14 year old boy presented with dryness of mouth, difficulty in swallowing solid food and bilateral progressive parotid swelling and lacrimal gland swelling since 6 months. On examination, a Non-tender swelling of both Parotid glands and lacrimal glands has been made out.

Fnac From Parotid: revealed degenerated epithelial cells with attached lymphocytes in the background of RBCs, mixed population of lymphoid cells, neutrophils and endothelial cells, suggesting a diagnosis of sialadenitis of autoimmune origin, most probably Mikulicz syndrome.

Discussion: In 1888, DR. johan mikulicz laid down the characteristics of this disease as "a symmetrical enlargement of the lacrimal and salivary gland of chronic inflammatory nature caused by unknown organism" in a 42 year old male, ever since then there is an ongoing

debate on origin, causative agent and classification of these lesions. Front et al¹ & Godwin et al in 1967, recommended replacement of term Mikulicz disease by Mikulicz syndrome for the clinical enlargement of lacrimal and salivary glands during the course of systemic disease. Parotid glands are the most commonly affected salivary glands (54.9%) cajualis et al². and Frable et al³ and it correlates well with the study conducted by Sukes, Pallipady A and Murthy N⁴.

FNAC features from the Parotid swelling in this case revealed degenerated epithelial cells with attached lymphocytes in the background of RBC'S, mixed population of lymphoid cells, neutrophils, and endothelial cells consistent with the characteristic description of Mikulicz syndrome. Apart from this, it also shows large cohesive sheets of pale, overlapping ductal-type cells infiltrated by lymphocytes called as lymphoepithelial islands⁵.

All suspected cases of Mikulicz syndrome have to be differentiated from Extranodal Marginal zone B cell lymphoma, Chronic Sialadenitis, Warthins tumour and simple lymphoepithelial cyst.

In Extranodal Marginal zone B cell lymphoma, there is monotonous population of B lymphocytes whereas in Mikulicz syndrome, along with lymphocytes there are degenerated epithelial cells, plasma cells and sometimes plump acinar cells. In chronic Sialadenitis, the aspirate is sparsely cellular with fewer lymphocytes, germinal centre fragments and lacking the characteristic lymphoepithelial islands⁵ whereas in Mikulicz syndrome, it is more cellular with plenty of lymphocytes, degenerated epithelial cells, plasma cells and a lymphoepithelial island consisting of large cohesive sheets of pale, overlapping, ductal-type cells infiltrated by lymphocytes. Warthin's tumour shows predominantly lymphocytes, oncocytes, granular debris and mast cells⁵.

Address Correspondence to :
Dr. Piyush Anand

JJM Medical College, Davangere
E-mail : drpiyushshe@gmail.com
Mob. : 08892396686



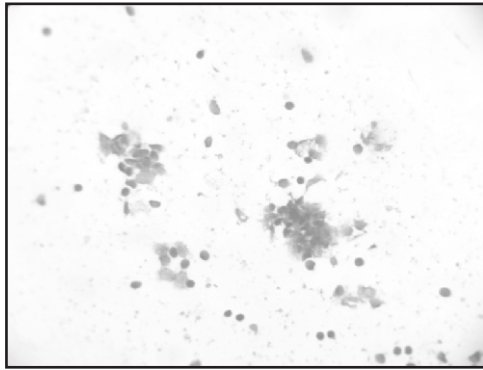


Fig1&2 Degenerated epithelial cells with clinging of lymphocytes,10x H & E stain

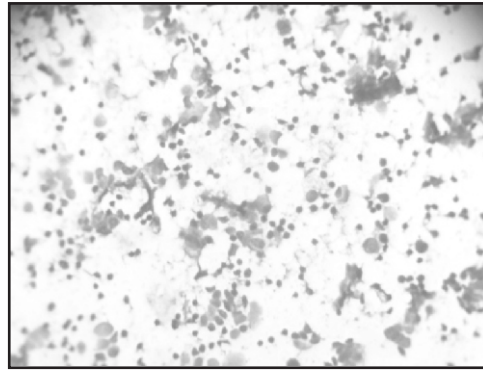


Fig. 4 Population of lymphoid cells, 40x Giemsa stain

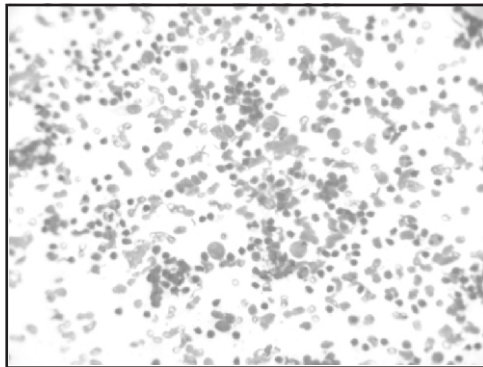


Fig. 3 Mixed population of lymphoid cells and epithelial cells, 10x H &E stain

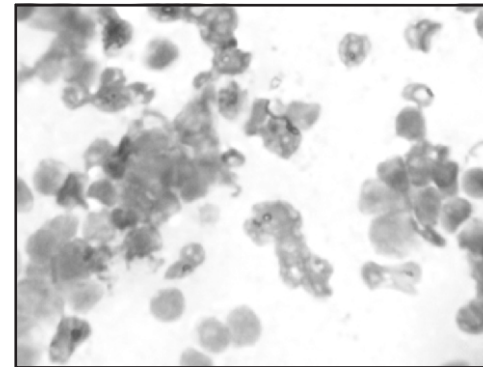


Fig. 6 Degenerated epithelial cells in the back ground of lymphoid cells & plasma cells, 10x H&Estain

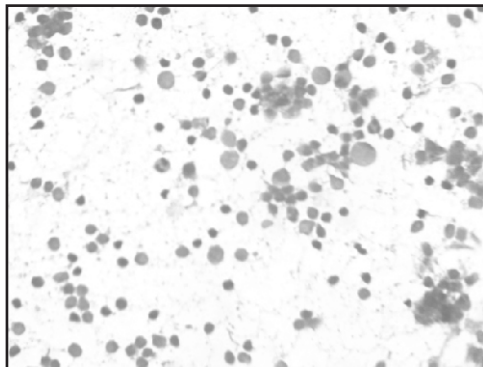
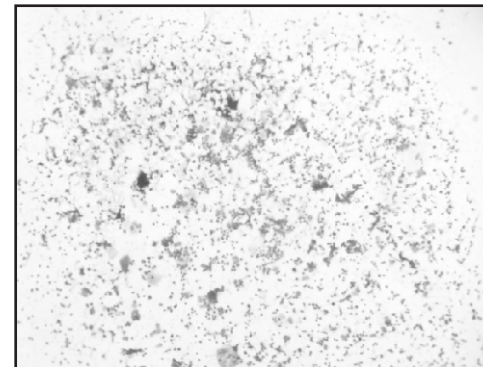


Fig. 5 Mixed population of lymphoid cells and plasma cells, 40xH&E stain



Conclusion : FNAC of salivary gland is an important diagnostic tool in conjunction with clinical correlation and back ground of differential diagnosis there by helping appropriate and timely management. High index of suspicion, careful evaluation and judicious use of diagnostic modalities would result in early and accurate diagnosis of Mikulicz syndrome.

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