Kuttner's tumor: The salivary gland tumor mimic

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Abstract
Kuttner tumor is a rare tumor like lesion of the salivary gland characterized by chronic inflammatory disease of the submandibular salivary gland. It is under diagnosed entity since 1896, when Kuttner reported four cases of chronic sclerosing sialadenitis (Kuttner's tumor) for the first time.
Diagnosis of chronic sclerosing sialadenitis is made only on histopathology; hence it is challenging entity for the clinician as well as for the pathologist in view of its presentation as hard palpable mass and mimics of malignant neoplasm.
We report case of 35-yrs- old male presented with left submandibular Kuttner tumor. The aim of the case report is to highlight the role of histopathology in early diagnosis and stress on this rare, under diagnosed tumor mimic.

Keywords: Kuttner tumor, Sclerosing, Sialadenitis, Submandibular gland.

Introduction
In 1986,series of patients with unilateral hard submandibular gland mass was diagnosed with chronic sclerosing sialadenitis by Kuttner.[1] It is misdiagnosed as malignancy and behaves as tumor mimics and classified as tumor like lesion of the salivary gland by WHO.[2] Kuttner tumor was under reported entity over the years and it is un-recognized by ENT surgeons preoperatively.[3] It is commonly presented as hard palpable mass in submandibular region and frequently excised following a provisional diagnosis of Carcinoma.[3] Hence its worrisome for the patient as well as the for the clinicians. Regarding the etiological factors the principal cause is sialolith and mucus plugs in 29-83 percentage of the lesions.[4] The other factors are ascending bacterial infections of the oral cavity and duct obstruction by foreign bodies.[4] Recently autoimmune role in the Kuttner tumor was postulated.[5,6]

In this case report, we highlighted the role of histopathology in the early and final diagnosis with specific features and stress on such rare and underdignosed tumor mimic that is Kuttner tumor (Chronic sclerosing sialadenitis).

Case Report
A 35-year-old-male presented to the ENT Surgeon's OPD with left submandibular mass since 2 months. The mass was gradually increasing and slightly painful. The personal and family history was not significant. Local examination revealed a firm, nodular and mild tender mass in submandibular region measuring 5x4 cms. No cervical lymphadenopathy noted. In view of its gross appearance, provisional diagnosis of malignancy was suspected and FNAC was advised. The FNAC was paucicellular with only lymphocytic infiltration and no epithelial cells seen in view of firm mass. Excisional biopsy was advised. All hematological, biochemical and serological investigations were within normal limits. Surgical excision in toto was done with care of facial nerve under general anesthesia. The post operative period was uneventful. We received single, irregular, nodular grey white firm mass measuring 4x3x1.5 cms. E/S-was grey whitish with multiple nodules of varying sizes. C/S-whitish with nodular and nodules appears to be separated by thin grey whitish firm areas with thick fibrous areas were more on periphery.

Light Microscopy: Multiple sections studied show salivary gland with partial effacement of the architecture by dense inflammatory infiltrate (Fig. 1). The varying sized nodules separated by fibrous bands were evident with diffuse and dense infiltration by lymphocytes and plasma cells (Fig. 2). Large areas of storiform fibrosis and acinar atrophy was noted in the center of the lesion (Fig. 3). The acinar atrophy with dense lymphocytic infiltrate encircling the glands was noted (Fig. 4). Focal areas showed ductal hyperplasia and apocrine changes due to chronic irritation in the periphery of the lesion was seen. There is no evidence of tuberculosis or malignancy in the sections studied. Final histopathological impression was given as-Chronic sclerosing sialadenitis (Kuttner tumor).After 6 months of follow up, the patient has been well without evidence of recurrence or any complaints.

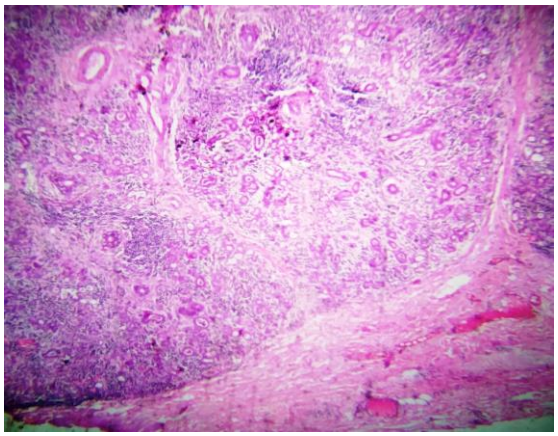

Fig. 1: Photomicrograph showed partial effacement of the architecture of salivary gland by dense inflammatory infiltrate and fibrosis (H &E stain, x40)

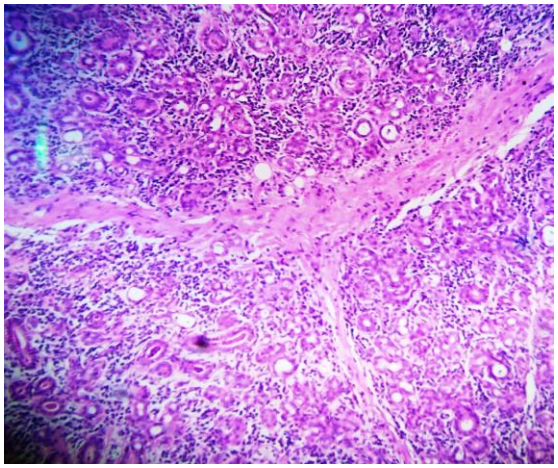

Fig. 2: Various nodules separated by fibrous bands with diffuse and dense infiltration by lymphocytes and plasma cells (H &E stain, x100)

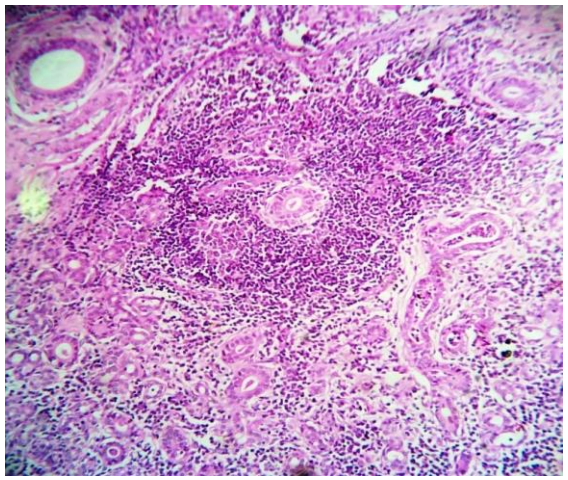

Fig. 3: Photomicrograph showed large areas of fibrosis, acinar atrophy and dense lymphocytic infiltrate (H &E stain, x100)

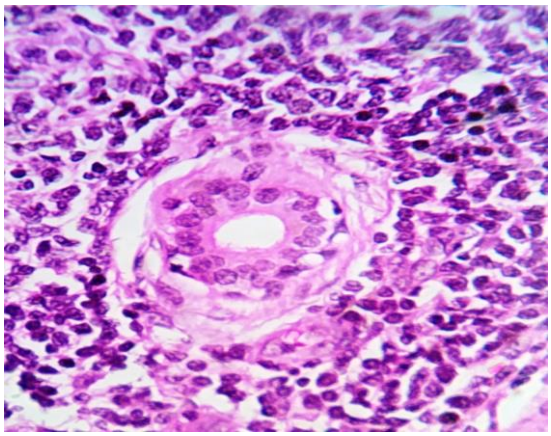

Fig. 4: The acinar atrophy with dense lymphocytic infiltrate encircling the glands on high power. (H &E stain, x400)

Discussion

Kuttner tumor is often misdiagnosed as malignancy preoperatively due to its hard and indurated consistency.[3] High index of suspicious and accurate diagnosis by histopathology is the key to success to diagnose Kuttner tumor (Chronic sclerosing sialadenitis).

The main clinicopathological feature to diagnose Kuttner tumor is hard and indurated swelling arising from the submandibular gland. Diffuse heterogeneity and multiple hypoechoic shadows mimicking liver cirrhosis on USG.[3] Regarding diagnosis, FNAC was very scant cellular and showed only lymphocytic infiltrate and fibrosis. Light microscopy showed preservation of lobular architecture, dense lymphocytic infiltrate, periductal fibrosis and loss of acini.[3]

The main precursors for the etiology are sialoliths and mucus plugs.[3] Ascending bacterial infections of the oral cavity with duct obstruction by foreign bodies are the other factors.[4] Recenlty, plasma cells are usually positive for IgG4 in most of the cases, given a clear relation with IgG4 related Kuttner tumor. 90% of Chronic sclerosing sialadenitis have been found to be IgG4 related.[5,6] In view of obliterate phlebitis in 1/3 of cases, elastin stain may have role in diagnosis of Kuttner tumor.[6]

The differential diagnosis of Kuttner tumor is varies from benign –simple chronic sialadenitis, granulomatous sialadenitis, necrotizing sialometaplasia, inflammatory pseudo tumor, lymphoepithelial tumor to carcinomas. Pre-operative steroid therapy may be tried to shrink the lesion.[5,6] Surgical excision is the standard treatment for the Kuttner tumor with evidence of recurrence or malignancy.[3]

Conclusion

Kuttner tumor is tumor like, benign and under –recognized entity over the years. We highlight the role of the histopathogy due to its clinical and gross feature mimic of malignancy. Surgery is the main treatment

modality of the Kuttner tumor. It should be kept in mind in the differential diagnosis hard mass in submandibular region.

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