KUTTNER TUMOUR – A RARE CLINICAL ENTITY WITH DIAGNOSTIC DILEMMA

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ABSTRACT: Kuttner tumour (chronic sclerosing sialadenitis) is a rare, chronic inflammatory disease of the salivary glands. It occurs more commonly in the submandibular gland. Hence it presents as a firm to hard swelling, it mimics malignancy clinically. The diagnosis is confirmed only by histopathological examination. We report a case of Kuttner tumour in a 55-years-old male who presented with a painful submandibular mass.
KEY WORDS: Kuttner tumour, submandibular gland, sialadenitis.

INTRODUCTION: Kuttner tumour was first described in 1896 by H. Kuttner, a German physician, who described a series of patients with a unilateral, hard, tumour-like mass of submandibular gland, which histologically showed features of chronic sclerosing sialadenitis[1]. Compared to parotid gland, the risk of malignancy is more in submandibular gland enlargement. Hence Kuttner tumour presents as a firm to hard mass, clinically it gives diagnostic dilemma in favour of malignancy. In order to raise the awareness of this condition, we present the case of Kuttner tumour in a middle aged man. To the best of our knowledge there are only few cases have been reported in Indian literature.

CASE HISTORY: A 55 year old male patient presented with a history of mild pain and swelling in the right submandibular gland since 6 months. He had no other specific complaints. On examination the swelling was firm to hard and measured 4 X 2 cm in size and confirmed as a salivary gland mass by CT scan. Examination of other salivary glands was unremarkable. There were no lymphadenopathies noted. The basic blood investigations were within normal limits. Patient was HIV negative. FNAC was done, but did not yield any diagnostic material. Excision biopsy was advised.

After a week the patient had undergone surgery and the specimen was sent for histopathological examination.

Histopathological examination:
Gross features: The specimen consisted of grayish white, firm mass of tissue measuring 3.5 x 1.5 x 1cm. Cut section revealed few grey white areas (Fig.1).
Microscopy: Multiple sections studied showed partial loss of architecture of the gland with areas of dense lymphoplasmacytic infiltrate and lymphoid follicle formation. Ducts showed dilatation and periductal concentric fibrosis. Acinar atrophy was noted. There was no evidence of granuloma or lymphoepithelial lesion (Figs.2-4). The diagnosis of Kuttner tumour was made.

DISCUSSION: Kuttner tumour, otherwise known as chronic sclerosing sialadenitis, is a chronic inflammatory disease of the salivary gland characterised by progressive periductal fibrosis, dilated ducts with a dense lymphocyte infiltration and lymphoid follicle formation and acinar atrophy[1]. It usually presents more commonly in males[2].

The characteristic clinical presentation is a firm or hard, painful or painless swelling in the submandibular gland of middle aged adults. It is rarely reported to occur in adolescents. More commonly there is unilateral involvement of the gland, but less frequently both glands as well as the parotid and minor salivary glands can be involved. Unusual presentation includes bilateral involvement of submandibular gland and lacrimal gland. The exact cause of this entity is uncertain. Several mechanisms have been postulated, including sialolithiasis secretory dysfunction with ductal inspissation, duct abnormalities, infectious agents and an autoimmune reaction[3].

The overall appearance is similar to that of sclerosing autoimmune pancreatitis, and this process now appears to be a member of the growing family of immunoglobulin (Ig) G4-related sclerosing diseases[4]. Tiemann et al[5] support that Kuttner tumour shows the features of an autoimmune process, where the quantity of CD 4 and CD8 positive cells was higher than that of CD3 cells and the number of cytotoxic T cells and macrophages decreased with increasing sclerosis.

The histological and cytological features of Kuttner tumor show various characteristics, according to stage in the progressive process and severity of inflammation. According to
Seifert, Kuttner tumor may evolve through four different histological stages as follows:

Stage 1: Focal chronic inflammation with nests of lymphocytes around salivary ducts, which are moderately dilated and contain inspissated secretion.

Stage 2: More marked diffuse lymphocytic infiltration, and more severe peri-ductal fibrosis. The ductal system shows inspissated secretion and focal metaplasia with proliferation of ductal epithelium. Peri-ductal lymphoid follicles are well developed. There is fibrosis in the centers of the lobules, accompanied by atrophy of acini.

Stage 3: Even more prominent lymphocytic infiltration, with lymphoid follicle formation, parenchymal atrophy, peri-ductal hyalinization, and sclerosis. Squamous and goblet cell metaplasia in the ductal system.

Stage 4: (end-stage): Cirrhosis-like, with marked parenchymal loss and sclerosis (the "burnt out" phase) [6].

Our case corresponds to stage 2.

The differential diagnosis includes sialadenitis, benign lymphoepithelial lesion, Kimura's disease, extra nodal marginal zone B-cell lymphoma of MALT, inflammatory pseudo tumour, fibrohistiocytic tumours, sclerosing lymphoma, sarcoidosis and neoplasms of the salivary glands[1]. Prognosis is very good as these are benign lesions that do not tend to recur. There have been reports nevertheless supporting the opinion that this condition may provide a state in which malignancy can arise[7].

CONCLUSION: Kuttner tumour is a rare disease that clinically mimics malignancy. It should be considered in the differential diagnosis of any firm to hard swelling of the salivary gland. Histopathological examination is a must to confirm this lesion.

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Fig1: Cut section of the right submandibular gland.

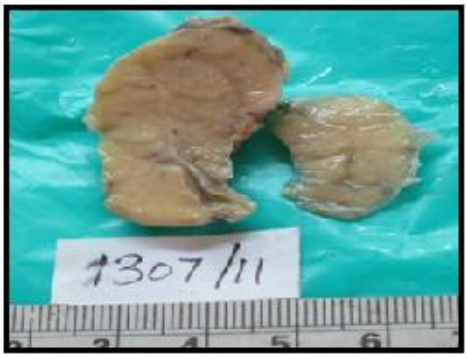


Fig 2: Scanner view shows chronic inflammatory cell infiltrate, lymphoid follicle formation, acinar atrophy and ductal dilatation.

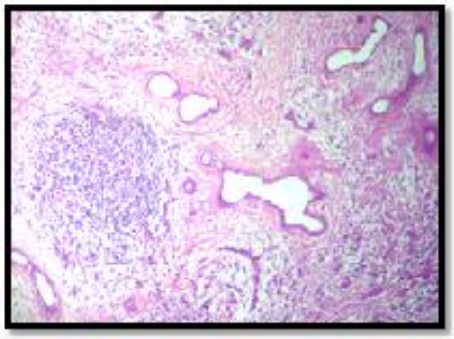


Fig.3: Periductal concentric fibrosis and inflammation (HEX20)

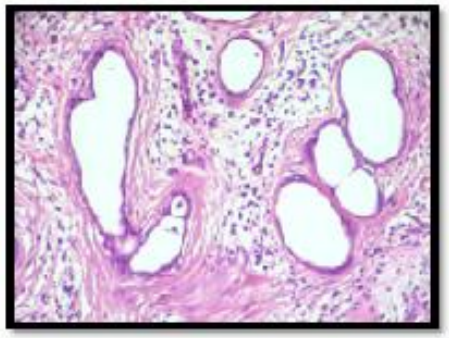
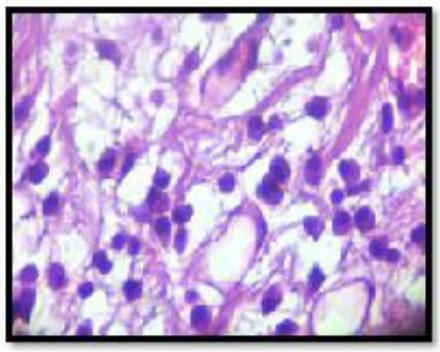


Fig. 4: Presence of plasma cells amongst the inflammatory cell infiltrate (HEx40)



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