KUTTNER TUMOR – A RARE AND UNDERDIAGNOSED ENTITY: A CASE REPORT
Prakash S.B¹, Nishan²

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ABSTRACT: Kuttner tumor also known as Chronic sclerosing sialadenitis (CSS). It is a rare, benign, fibro-inflammatory disease of the salivary gland most commonly affecting submandibular gland¹-⁴, characterized by progressive fibrosis of periductal area, dilated ducts with a dense lymphocytic infiltration, lymphoid follicle formation, and acinar atrophy and ultimately leading to marked sclerosis of the gland. We are reporting this case to increase the awareness about this rare entity which masquerades as malignancy and usually under diagnosed because of the diagnostic confusion.

KEY WORDS: kuttner tumor, benign, sclerosis, submandibular gland.

INTRODUCTION: Kuttner tumor is a rare, benign, inflammatory condition commonly affecting submandibular salivary gland. It was first described in 1896 by German physician¹, H. Kuttner. He explained it as hard tumor, usually unilateral, most commonly affecting submandibular gland, which histologically showed features of chronic sclerosing sialadenitis. In new WHO classification it is classified as a tumor like lesion of salivary gland⁵.

The pathology behind it is still uncertain. Many causes have been identified like sialolithiasis, infection of chronic duration, secretory dysfunction with ductal inspissation, abnormalities of duct, autoimmune reaction, involvement of IgG4 antibodies⁶ and its disturbance in the pathogenesis of sclerosing sialadenitis (Kitawaga et al)⁶. Tieman et al⁷ found that there was an intimate relationship between the T-cell dominated inflammatory infiltrate and the acinar and ductal cells.

CASE SUMMARY: A 50 year old male, non-diabetic, non-hypertensive presented with a painless, gradually enlarging, firm to hard nodule in the right submandibular region since 18 months. There was no history of dryness of mouth, no history of trauma.

Fig. 1: Solitary swelling in the right submandibular area
Local examination on inspection revealed about 3.0x1.5x2.0 cm, single swelling in the right submandibular region with normal appearing skin over it. On palpation there was a single, mobile, non-tender, firm to hard swelling with well-defined borders and irregular surface. No palpable cervical nodes were present. The examination of the oral cavity, left submandibular gland and both the parotid glands did not reveal any abnormality. Submandibular duct openings were normal. Swelling was bidigitally palpable and mobile. Systemic examination was normal and blood investigations revealed normal parameters. He was negative for both HBsAg and HIV. To know the pathology we advised him for fine needle aspiration cytology (FNAC). FNAC was done for two times, but both the time it showed features of chronic reactive lymphadenitis. X ray showed no evidence of calculi. Sialography was inconclusive. The CT and USG neck showed the mass was confined to submandibular gland and there was no involvement of adjacent structures. So we planned for excision biopsy after doing all imaging studies.

**Intraoperative findings:** Whole gland was enlarged with intact fascia over it. It was firm to palpate and mobile. Submandibular gland along with one lymph node excision was performed and specimen was sent for histopathological examination.

Macroscopically the specimen was irregular, firm, and tan-brown with cut surface showing areas of fibrosis. No stone was palpable.

**Histopathological findings:** Microscopically the lobules showed dense lymphoplasmacytic infiltrate with acinar atrophy and periductal concentric fibrosis and lymphoplasmacytic inflammation with lymphoid follicle formation. There was no evidence of lymphoepithelial lesions, granulomas, ductal atypia or malignancy.
Post-operative follow up: No further treatment was advised. He was given one course of antibiotics and was assured about the benign nature of the disease. Wound healing was good and there was no sign of recurrence and the patient is under good follow up.

DISCUSSION: Kuttner tumor is a fibroinflammatory disease of the submandibular gland characterized by progressive periductal fibrosis, dilated ducts with a dense lymphocytic infiltration, lymphoid follicle formation, acinar atrophy and eventually marked sclerosis of the gland. Usually it present as unilateral swelling but very rarely it can involve both the glands. It can present as a firm to hard, painless (painful rarely) swelling in the submandibular gland of middle aged adults. In adolescents it is rarely reported. It can involve parotid and lacrimal gland in rare cases.

To rule out malignancy USG /CT/MRI neck can be done. But the ultimate method of confirmation is tissue biopsy.

Ahuja et studied the sonographic findings in case of Kuttners tumor and found liver like pattern, i.e. multiple hypoechoic lesions against a heterogeneous background as the most common findings. That is why it is also known as 'cirrhosis' of submandibular gland.

Kuttner tumor has some characteristic features on fine needle aspiration cytology,

- Relatively low cellularity,
- Scattered ductal structures with acinic atrophy
- Ducts intimately surrounded by fibres or lymphoid cells
- Small isolated fragments of fibrous stroma
- Moderate to large number of lymphoid cells lacking atypia

Fig. 3: Histological view of the tumor showing lobular disarray with periductal and intralobular chronic inflammation including follicle formation. (100X)

Fig. 4: Histological view of the tumor displaying dilated salivary ducts surrounded by concentric fibrosis and lymphocytic and plasmocytic inflammatory cell infiltration. (200X)
The histopathologic features of Kuttner tumor may evolve through four histologic stages:

1. Mild, focal chronic lymphocytic and plasmacytic cell infiltration, usually periductal with periductal fibrosis. The lobular architecture of the gland is preserved.
2. Loss of lobular architecture with dense lymphocytic infiltration and severe periductal fibrosis.
3. More prominent lymphoplasmacytic infiltrate with reactive lymphoid follicle formation, extensive fibrosis with acinar atrophy, periductal hyalinization and ductal dilatation.
4. Destruction of lobular architecture with sclerosis with parenchymal loss.

The differential diagnosis of Kuttner tumor includes 3-6
- Salivary gland neoplasms.
- Chronic sialadenitis
- Sjogren’s syndrome
- Kimura ‘s disease
- Inflammatory pseudotumour etc.

The presence of a heavy lymphoid infiltration can raise the suspicion of other lymphoproliferative processes such as intraglandular reactive lymph node, benign lymphoepithelial lesion and low-grade lymphoma.

Immunohistochemical studies of the lymphoid population in Kuttner tumor show the presence of activated B cells in the lymphoid follicles, while the T cells in the perifollicular zone exhibit great predominance of helper/ inducer T cells over suppressor/cytotoxic T cells.

Kuttner tumor is a benign process which has no tendency for recurrence. However there are reports suggesting that this condition may provide a state in which malignancy may arise. A case report of marginal zone B cell lymphoma arising in a Kuttner tumor has been published by Ochoa et al in 2001 13.

Surgery is the standard and best treatment for Kuttner tumor. However in patients with small and asymptomatic tumor observation and regular follow up is enough. This entity has to be considered in the differential diagnosis of the salivary gland tumors in patients presenting with firm to hard swelling of the salivary gland.

CONCLUSION: Kuttner tumor is a rare entity may be because it is underdiagnosed most of the time. It’s a benign entity. Because of the CT /MRI scan and FNAC one can suspect such benign lesions of salivary glands but confirmation by histopathological examination remains the gold standard modality.

REFERENCES:

AUTHORS:
1. Prakash S.B.
2. Nishan

PARTICULARS OF CONTRIBUTORS:
1. Assistant Professor, Department of ENT, MMC & RI, Mysore.
2. Post Graduate, Department of ENT, MMC & RI, Mysore.

NAME ADDRESS EMAIL ID OF THE CORRESPONDING AUTHOR:
Dr.Prakash S.B.,
Door No. 136,
Vijayanagara 4th Stage, 3rd Phase,
Mysore – 18.
Email-prakashsb.mmc@gmail.com

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