CASE REPORT

Küttner's tumour: an unusual cause of salivary gland enlargement

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Abstract

Enlargement of the submandibular gland is a condition that raises the alarm for further investigation as the risk of an underlying malignancy is higher in comparison to parotid gland enlargement. Chronic sclerosing sialadenitis or Küttner's tumour is usually mis-recognised as it is only after excision of the gland that the correct diagnosis is made.

We present a case of a 47-year-old male patient who presented with one year history of firm non-tender enlarged right submandibular gland which was removed surgically and histology showed to be sclerosing sialadenitis.

Küttner's tumour is a rare disease, which mimics malignancy. There is not enough evidence to support any diagnostic means that could help in the differential diagnosis of this benign condition. Given the high rate of malignancy in firm, painless lesions of the submandibular gland, surgical excision is often advocated and Küttner's tumour is usually diagnosed by the histopathologist. Hippokratia 2008; 12 (1): 56-58

Key words: Kóttner's tumour, chronic sclerosing sialadenitis, submandibular gland.

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Introduction

Enlargement of the submandibular gland is a condition that raises the alarm for further investigation as the risk of an underlying malignancy is higher in comparison to parotid gland enlargement. Chronic sclerosing sialadenitis is a condition that is perhaps more common than thought but usually mis-recognised as it is only after excision of the gland that the correct diagnosis is made. We present a case of a male patient with chronic sclerosing sialadenitis otherwise known as Küttner's tumour in order to raise awareness of this interesting and not well-known entity.

Case Report

A 47-year-old male patient presented with twelve months' history of right submandibular painless swelling. He denied any alteration in his taste or other symptoms. On examination he had a very firm non-tender enlarged right submandibular gland confirmed by CT scan. Examination of the rest of the salivary glands was unremarkable. A salivary gland malignancy was suspected and the lesion was removed surgically and histology (Figures 1 and 2) showed preservation of its architecture, marked septal fibrosis, varying acini atrophy, accompanied by a moderate to marked lobulocentric inflammatory infiltrate of lymphocytes, plasma cells and reactive lymphoid follicles. No lymphoepithelial islands were identified. Immunohistochemistry demonstrated a reactive lymphoid population; B cell markers (CD20 and CD79a) highlighted the B-lymphocytes with no CD43 co-expression and with kappa and lambda showing no light chain restriction. CD3 and CD43 stained the T lymphocytes with a preponderance of CD4 over CD8 lymphocytes. Cyclin D1 did not stain the lymphocytes. This report was consistent with the diagnosis of sclerosing sialadenitis otherwise known as Kuttner's tumour. The patient had an unremarkable recovery.

Discussion

Küttner's 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[1,2]. It was first described in 1896 by H. Küttner, a German physician, who described a series of patients with a uni-

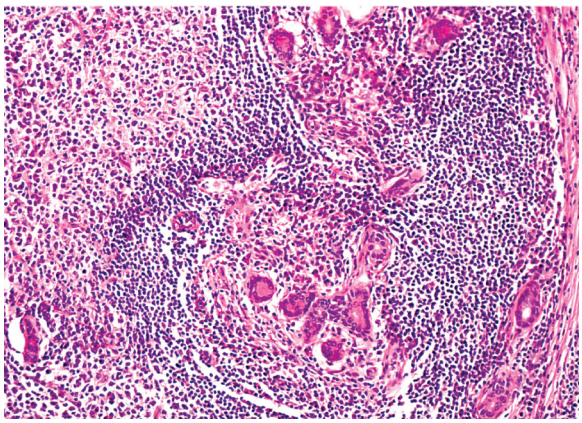

Figure 1. Medium power histological view of the tumour showing residual salivary ducts with areas of chronic inflammation including follicle formation and peripheral fibrosis.

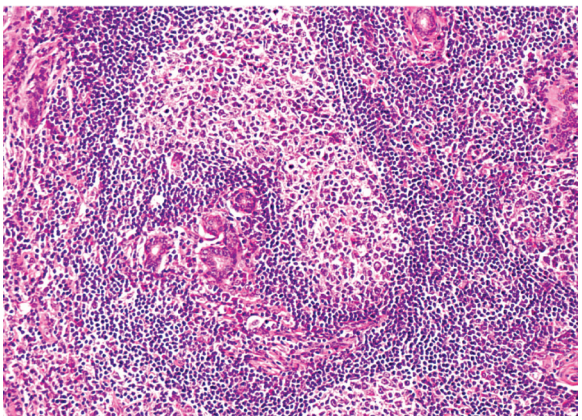

Figure 2. High power histological view of the tumour displaying salivary ducts surrounded by follicles of lymphoid tissue.

lateral, hard, tumour-like mass of submandibular gland, which histologically showed features of chronic sclerosing sialadenitis[3].

Küttner's tumour creates an interesting diagnostic dilemma since, although it is a benign lesion, it resembles closely the clinical characteristics of malignancy, i.e. it presents as a painless hard mass in the submandibular gland. The known fact that 80% of tumours presenting in this organ are malignant tumours complicates the management of Küttner's tumour even further. Despite the fact that it has been known for more than a century, it is still a rare lesion, but one which must be taken into account in the differential diagnosis of submandibular tumours.

There are several theories of the disease's cause: sialolithiasis is said to induce the inflammation and fibrosis [1, 4] although this is not clear and other authors state that the formation of calculi is secondary to the sialadenitis[5]. Other causes include an active local immune reaction, [1] secretory dysfunction with ductal inspissation, duct abnormalities, infectious agents or autoimmune reaction[6]. Kitawaga et al [2] suggest the involvement of IgG4 antibodies and its disturbance in the pathogenesis of sclerosing sialadenitis. Tiemann et al[7] support that Küttner's 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.

It usually presents more commonly in males[1] although other authors report an equal sex distrubution. Its clinical characteristics include presentation as a firm or hard painful or painless mass usually in the submandibular gland. Most commonly there is unilateral involvement of submandibular gland, but less frequently both glands as well as the parotid and minor salivary glands can be involved[8-10].

Sonographic appearances resemble those of "cirrhotic liver", diffuse involvement with multiple hypoechoic lesions against a heterogeneous background together with duct dilatation and calculi. Doppler shows prominent intraglandular vessels, with no evidence of displacement. Focal lesions are seen as hypoechoic, heterogeneous masses with a radial branching vascular pattern within[11].

The differential diagnosis includes sialadenitis[2], Mikulicz's syndrome[2], benign lymphoepithelial lesion, Kimura's disease, extranodal marginal zone B-cell lymphoma of MALT[12], inflammatory pseudotumour[13], fibrohistiocytic tumours, sclerosing lymphoma, sarcoidosis and neoplasms of the salivary glands[4]. Several associations have been reported, such as sclerosing cholangitis or sclerosing pancreatitis[2]. In the latter the presence of IgG4 is again another indicator of the relation between the two entities[2].

Histological characteristics include septal fibrosis and acini atrophy in a bed of chronic inflammatory infiltration and follicle formation. IgG plasma cell infiltration is another characteristic sign[2].

Management can be conservative by adopting a "watch and wait" approach in the case where the mass is otherwise asymptomatic and the patient is comfortable, provided there is adequate evidence that the lesion is benign. Chou et al[6] suggested colour Doppler sonography and sonographically guided needle biopsy as an adequate means of diagnosis, which avoids operative excision of an otherwise benign condition, but their results are based on a small number of patients. Most commonly the mass is surgically excised, as it is difficult to differentiate it from a malignancy due to the fact that a mass in the submandibular gland in particular has a high likelihood of being malignant. It is true that if it was possible to pre-operatively differentiate this lesion from a malignant tumour we would most likely avoided performing an operation for a mass which was painless and was otherwise causing no symptoms. However the suspicion of a salivary gland carcinoma is not one to be taken lightly and could therefore not be ignored.

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 state in which a malignant lesion can arise[13].

Conclusion

Küttner's tumour is a rare disease, which mimics malignancy. There is not enough evidence to support any diagnostic means that could help in the differential diagnosis of this benign condition. Given the high rate of malignancy in firm, painless lesions of the submandibular gland, surgical excision is often advocated and Küttner's tumour is usually diagnosed by the histopathologist.

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