ORIGINAL ARTICLE

Kuttner's tumour (chronic sclerosing sialadenitis) of the submandibular gland: a clinical perspective

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Objective To study clinical perspectives pertaining to chronic sclerosing sialadenitis, which is also known as Kuttner's tumour.

Design Retrospective medical chart review.

Setting Regional hospital, Hong Kong.

Patients From February 2005 to February 2007, nine cases with Kuttner's tumour were identified from our hospital electronic database.

Interventions They underwent submandibular sialadenectomy under either local (n=6) or general (n=3) anaesthesia.

Main outcome measures The results of preoperative ultrasonography, fine-needle aspiration cytology, and intra-operative frozen section examination were correlated with the final diagnosis. Operative morbidity was also evaluated.

Results The mean age of the patients at diagnosis was 61 years; three were females. Three had bilateral submandibular swellings. Following preoperative ultrasonography in six of the patients, tumours were suspected in two, an enlarged lymph node in one, and diffuse enlargement was visualised in the other three. Six patients had preoperative fine-needle aspiration cytology; five yielded scanty acini with normal-looking ductal cells, variable degrees of infiltration by chronic inflammatory cells without granuloma admixing fibrosis. In the sixth patient, only bland-looking epithelial cells, indicative of ductal differentiation suspicious of neoplasm were noted. Intra-operative frozen section examination was conducted in three patients: chronic inflammation without evidence of carcinoma was visualised in each. Operations performed under local anaesthesia were well tolerated; only one patient endured a transient, marginal facial nerve palsy.

Conclusions Kuttner's tumour is by no means rare. When supported by ultrasonography and fine-needle aspiration cytology, an accurate diagnosis can be made preoperatively and surgery can be reserved for symptomatic cases. Submandibular sialadenectomy is a safe and effective means of treating Kuttner's tumour, and can be accomplished under local anaesthesia.

Key words
Autoimmune diseases; Sclerosis; Sialadenitis; Submandibular gland

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Introduction

Chronic sclerosing sialadenitis, also known as Kuttner's tumour (KT), is a benign tumour-like lesion predominately affecting the submandibular gland first recognised by Kuttner in 1896.[1] As a result of the chronic inflammation, the presenting masses are often indurated and masquerade as carcinoma.

Although described more than a century ago, this clinical entity is still under-reported and preoperatively it remains unrecognised by many surgeons. The submandibular masses are often removed following a preliminary diagnosis of carcinoma, thus inciting substantial worry to both patients and clinicians. This state of affairs is attributed to the under-reporting of KT in the English literature,[2] which mainly focuses on its pathological aspects. From our experience, KT is not infrequent. We describe herein a cohort of KT, in order to draw the attention of clinicians to this unfamiliar disease. The results of preoperative investigations and intra-operative frozen section (FS) examinations could therefore be correlated with the final diagnosis and the latest concepts about its pathogenesis discussed.

Methods

Twenty-one patients who underwent submandibular sialadenectomy from February 2005 to February 2007 were identified from our hospital electronic database. Among these, nine (43%) were confirmed to be KT, based on the clinicopathological testing, and represented our study group. The pathological diagnoses of the remaining 12 patients were: pleomorphic adenoma (n=7), Warthin's tumour (n=1), mucoepidermoid carcinoma (n=1), adenoid cystic carcinoma (n=1), and acute inflammation (n=2).

The nine patients all presented with submandibular swellings that clinically resembled discrete lumps, suspected to be tumours. The final histopathological examination of these nine patients revealed the characteristic preservation of the lobular architecture of the salivary gland admixing marked lymphoplasmacytoid infiltration, periductal fibrosis, and varying degrees of acinar atrophy.[3]

The clinical features of these patients were studied by reviewing their case charts. The results of the preoperative imaging investigations, fine-needle aspiration cytology (FNAC) and FS examination reports were evaluated for any suspicion of neoplasm or KT.

The submandibular sialadenectomy was performed under either local anaesthesia (LA) or general anaesthesia (GA). A 4- to 5-cm transverse neck incision was made about 2-finger breadths below the mandible. Subplatysmal flaps were elevated. After the marginal facial nerve was preserved, the superficial lobe of the submandibular gland was mobilised. Vessels from both the distal and proximal facial vessels were controlled. The mylohyoid muscle was then retracted to expose the deep lobe. When the lingual nerve was identified, its ganglionic branch to the submandibular gland was divided. The submandibular duct was then divided after ligation. The whole submandibular gland was removed. A drain was usually not inserted. If enlarged adjacent lymph nodes were encountered, they were also removed.

Results

The mean age of these nine patients was 61 (range, 48-76) years; three were female. Four (44%) patients complained of painful submandibular swelling, while the other five had painless masses. Three patients had bilateral submandibular swellings; notably, a left parotid mass was also identified in one of them. The median duration of the presenting masses was 2 months (range, 0.5 months-10 years). The median diameter of the submandibular masses was 2.3 cm (range, 1-3 cm), and in three patients stones were present in the Wharton's duct (Table).

Preoperative ultrasonography (USG) was performed on six patients; tumours were suspected in two, an enlarged lymph node in one, and diffuse enlargement was visualised in the other three (in two of whom stones were displayed). Two patients underwent computed tomographic imaging; diffuse parenchymal disease was diagnosed in one, and a stone was noted in the other. Only one patient had magnetic resonance imaging; diffuse glandular enlargement with one stone in the Wharton's duct was detected.

Six patients had preoperative FNAC; the other three were spared this investigation as stones were detected during imaging. In the cytological findings of five of these patients, FNAC revealed scanty acini and normal-looking ductal cells, with a variable degree of chronic inflammatory cell infiltration but no granuloma admixing fibrous tissue. In one patient, only bland-looking epithelial cells of ductal differentiation suspicious of neoplasm were present.

Intra-operative FS examination of the specimens was conducted in three patients; chronic inflammation

以臨床角度探討下頜下腺Kuttner腫瘤（慢性硬化性頜下腺炎）

目的 從臨床角度探討又稱為Kuttner腫瘤的慢性硬化性頜下腺炎。

設計 對病歷作回顧研究。

安排 香港一所分區醫院。

患者 在2005年2月至2007年2月期間，從香港醫院電子數據庫中識別出9宗Kuttner腫瘤的病例。

療法 局部麻醉（n=6）或全身麻醉（n=3）下施行頜下腺摘除術。

主要結果測量 計算術前超聲波掃瞄、細針穿刺細胞學檢查、術中冰凍切片檢查的結果與最後診斷的相關性，並評估術後併發症。

結果 患者接受診斷時的平均年齡為61歲，3人為女性。3人有雙側頜下腫脹。對其中6人進行術前超聲波掃瞄檢查後，懷疑其中兩人有腫瘤，一人有腫大淋巴結，其餘三人屬彌漫腫大型。6人接受術前細針穿刺細胞學檢查，發現5個檢查結果有極少腺泡，含外觀正常的導管細胞，並有不同程度的炎細胞浸潤，但無摻雜纖維組織的肉芽腫。在第6位患者的結果，只發現有外觀無異樣的上皮細胞，表示有導管分化，懷疑有腫瘤。3位病人接受術中冰凍切片檢查，全部結果顯示慢性炎症但沒有證據顯示患癌。局部麻醉下進行的手術無排斥反應，只有1位病人出現極短暫而輕微的面癱。

結論 Kuttner腫瘤並不是罕見的疾病。利用超聲波掃瞄和細針穿刺細胞學檢查，可在術前作出準確的診斷，因而可以只為有症狀的患者施行手術。治療Kuttner腫瘤其中一種安全有效的方法就是頜下腺摘除術，而且只需局部麻醉已可施行。

TABLE. Salient demographic and clinical features of the patients with Kuttner's tumour and information related to their surgery*

Case No.	Sex/age (years)	Pain (+/-)	Duration† (months)	Bilateral or unilateral	Size (cm)	Stone (+/-)	Operation	
							GA/LA	Complication
1	F/55	+	0.5	Bi	3.0	-	GA	Nil
2	F/64	+	2.0	Uni	NR	+	LA	Nil
3	M/64	-	2.0	Uni	2.0	-	GA	TFP
4	F/58	-	12	Uni	1.5	-	LA	Nil
5	M/76	-	2.0	Uni	1.0	-	LA	Nil
6	M/57	+	3.0	Uni	2.5	+	LA	Nil
7	M/48	-	1.0	Uni	2.5	-	GA	Nil
8	M/72	-	3.0	Bi	1.5	-	LA	Nil
9	M/59	+	120	Bi	3.0	+	LA	Nil

* NR denotes not recorded, GA under general anaesthesia, LA under local anaesthesia, and TFP transient facial paresis

† Duration refers to pain, swelling, or either clinical feature at diagnosis

was visualised in each instance. Carcinoma was not suspected in any of them.

Of the nine surgical procedures, six and three were performed under LA and GA, respectively. The LA was well tolerated by the patients and none were converted to GA. Regarding surgical complications, only one patient (having GA) suffered a transient marginal facial nerve palsy, which recovered spontaneously.

Discussion

Preoperatively, KT is often misdiagnosed as a malignant neoplasm due to its indurated consistency. Fortunately, as the index of suspicion for this disease increases, an accurate preoperative diagnosis can be made. Kuttner's tumour should be seriously considered in patients with bilateral submandibular involvement, and the diagnosis is reinforced by the absence of malignant cells in FNAC specimens. Cheuk and Chan[4] reported the cytological features of KT to be paucicellular to moderately cellular, scattered ductules enveloped by collagen bundles or a lymphoplasmacytic infiltrate, isolated fragments of fibrous stroma and scanty acini.

Interestingly, KT can also affect the parotid gland.[5] In our series, one patient had bilateral submandibular swelling co-existing with left parotid swelling. Thus, multi-glandular presentation is indicative of KT.

The ultrasonic features of KT have been described as a diffuse appearance simulating liver cirrhosis with or without duct dilatation or calculus.[6] For equivocal cases, FS examination can disclose the diagnosis,[7] and therefore reassure the surgeon immediately. Otherwise, simultaneous neck dissection might appear necessary, as metastatic lymph nodes are often found in the same vicinity. The following is a summary of the diagnostic clinicopathological features of KT:

1. Indurated swelling arising from submandibular gland;
2. Diffuse heterogenous echogenecity and multiple hypoechoic shadows mimicking liver cirrhosis on USG examination;
3. FNAC findings showing paucicellularity with scattered tubular ductal structures surrounded by lymphoplasmacytic infiltration in a background of fibrous stroma; and
4. Histopathology showing preservation of the lobular architecture, dense lymphoplasmacytic infiltration, periductal fibrosis, and loss of acini.

The aetiology of KT is not well-elucidated. Postulated factors important in its pathogenesis comprise sialoliths and intrinsic ductal abnormality resulting in inspissated secretions, which can evoke chronic inflammation.[2] More recently, an autoimmune aetiology has been postulated. Sekine et al[8] reported a patient afflicted by simultaneous KT and idiopathic retroperitoneal fibrosis. Both pathologies shared similar histological and immunohistochemical features, possibly mediated by macrophages. Tsuneyama et al[9] described a case of sclerosing cholangitis associated with KT. The histopathological findings of both biliary and salivary ducts were quite analogous. They hypothesised that similar immune reactions take place in both organs. Kitagawa et al[10] in 2005 classified KTs into two types—class 1 associated with extra-salivary sclerosing lesions, and class 2 confined to salivary glands. Abundant immunoglobulin G4 (IgG4)–positive plasma cells were present in both types. In 2006, Kamisawa et al[11] reported four patients with KT, two of whom also manifested autoimmune pancreatitis. They suggested that the serum IgG4 level was useful in diagnosing KT. Because our series was evaluated retrospectively,

serum/tissue IgG4 levels were not available.

Controversies exist regarding the relationship between Sjogren's syndrome, Mikulicz's disease and KT. Kamisawa et al[12] demonstrated that IgG-positive plasma cells are present in sclerosing sialadenitis but not Sjogren's syndrome or sialolithiasis. By contrast, raised serum IgG4 concentrations and IgG4-positive plasma cells were found in Mikulicz's disease, which often responds to steroid treatment.[13] Because of these differences, Sjogren's syndrome should be regarded as distinct from the other two entities. Based on the systemic nature of IgG4-related autoimmune disease,[14] Mikulicz's disease and IgG4-positive KT can therefore be considered as different manifestations of this new clinicopathological entity.

Surgery is the standard therapy for KT. However, based on immunological findings recently implicated in its development, administration of steroids has been shown to be effective in shrinking such salivary swellings.[11] From a surgeon's perspective, we believe submandibular sialadenectomy offers a better treatment option. Our results show that operative morbidity is minimal; only one patient suffered a temporary marginal facial nerve palsy. Despite the presence of chronic inflammation and adhesions, the operation can even be performed as a day case under LA, without undue difficulty. Moreover, after surgery the mass vanishes immediately. By contrast, steroids can induce severe side-effects and take several weeks for full resolution of the submandibular swelling. Steroids should be reserved for patients with extra-salivary involvement (pancreas/biliary tract) or those who refuse surgery. Of course, for patients with painless and non-progressive KT in whom the preoperative diagnosis is unequivocal, observation should also suffice.

Conclusion

Kuttner's tumour is by no means rare; it is just under-recognised. According to our experience, it is present in a significant proportion (44%) of patients undergoing submandibular excision. With increased awareness of this disease and the support of USG and FNAC findings, an accurate preoperative diagnosis can be made. Moreover, submandibular sialadenectomy can be accomplished under LA, which is a safe and effective means of therapy for symptomatic KT without extra-salivary involvement.

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