

Kuttner's tumor of bilateral submandibular and parotid glands

Abstract

Chronic sclerosing sialadenitis, known as Kuttner tumor (KT), is a chronic inflammatory disease of the salivary glands. KT is a benign condition but presents with a hard mass that clinically mimicks salivary gland neoplasms. It is an underdiagnosed entity among pathologist and otolaryngologist even though it was first reported by Kuttner more than a century ago. It might be due to rare occurrence and not to have malign histologic features. KT primarily affects one of the submandibular glands. Herein, we report a case of KT presenting with time-serial involvement of the bilateral submandibular and parotid glands. This case would be of particular interest to head and neck surgeons and pathologists, in order to create more awareness with regard to KT which is a not well-known entity.

Keywords: salivary gland, submandibular gland, kuttner tumor, chronic sclerosing sialadenitis

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Abbreviations: KT, kuttner tumor; ANA, anti-nuclear antibody; IgG4, immunoglobulin g4

Introduction

Chronic sclerosing sialadenitis, known as KT, was first described in 1896 by Kuttner. KT is clinically characterised by a firm mass of submandibular gland in middle-aged adults. This is an uncommon tumor-like lesion that typically involves one of the submandibular glands.¹⁻³ The treatment of choice is complete removal of the affected gland. The definitive diagnosis is made by histopathological examination. *Histologically*, KT is characterized by lymphoid follicle, acinar atrophy, periductal fibrosis and dilated ducts. it is a benign and relatively uncommon condition. Therefore, it may be a little difficult for a pathologists to distinguish KT from other forms of sialadenitis. Multiglandular involvement of KT rarely occurs and it might be a manifestation of IgG4-related systemic sclerosing disease.^{4,5} We experienced a rare case of KT that presents with a time-serial involvement of the bilateral submandibular and parotid glands. This report described the histological, clinical features and management of this patient. We aimed to raise awareness among otolaryngologists and pathologists about a not well-known and under-recognized entity.

Case report

A 60-year-old woman was admitted to our clinic with the complaint of a *slow-growing mass* in the left *submandibular region* for 4 months. She had no history of drying eyes or mouth. She was with normal general health and no other personal history of disease. Physical examination revealed a very firm, *slightly tender*, 4×3 cm sized left submandibular mass. Ultrasonography showed diffuse enlargement with impaired parenchymal structure in the left submandibular gland. But there was no sign of sialolithiasis. No other masses or adenopathy were noted in the head and neck in physical examination and ultrasonography. Flexible endoscopic examination of the nasopharynx, hypopharynx, larynx was normal. *Fine needle aspiration cytology* showed a *non-specific inflammation*. Routine laboratory tests including full blood count, white cell count, liver function tests, erythrocyte sedimentation rate were within normal range. Left submandibular gland excision was performed

under general anesthesia. Light microscopic examination of tissue stained with hematoxylin and eosin revealed remarkable fibrosis, accompanied by the inflammatory infiltrate and lymphoid follicles (Figure 1A). Acinar atrophy, concentric *periductal sclerosis* (“*onion-skin*” appearance), and ductal ectasy were also observed (Figure 1b). But there were no neoplastic features. All these findings were consistent with chronic sclerosan sialadenitis. The patient recovered well after operation. But she presented with a prominent right submandibular mass and mild bilateral *symmetric* parotid swelling three months later (Figures 2A & 2B). Right submandibular mass had the same clinical features of the left. Computed tomography of the neck with contrast was also performed. It showed diffuse enlargement of the right submandibular and bilateral parotid gland which enhance moderately (Figure 3). The patient underwent operation again and right submandibular gland was surgically removed. Histological features of the excised right submandibular gland same as the left submandibular gland. Specimens of the bilateral submandibular glands displayed the histopathologic features of the chronic sclerosing sialadenitis. Bilateral submandibular gland specimens predominantly showed the stage 2 KT according to Seifert's histopathologic classification (Table 1). *We also performed immunohistochemical staining*. Inflammatory infiltrate were composed of CD4+ T cells (helper T cell) and CD8+ T cells (cytotoxic T lymphocyte) in periductal area (Figures 4A&4B). A large number of CD 138+ cells(plasma cell) were also observed in interfollicular area (Figure 4c). Lymphoid follicles predominantly consisted CD 20+ cells (B lymphocyte) in the germinal center (Figure 4D). Patient was also *positive for anti-nuclear antibody(ANA) in blood*. These findings indicated that it may be an *immune-mediated process* and the patient was referred to an immunologist for excluding the autoimmune diseases. Other autoimmune antibodies were not detected. IgG and IgG4 level that might be elevated in IgG4-related systemic sclerosing disease were also normal range. Immunologist reported to have no evidence of otoimmun diseases in the patient because there was no sign of autoimmun disorder except the positive antinuclear antibody that considered as non-specific. The patient was followed for 6 months without further therapy. No other neck masses detected in this period except bilateral mild parotid swelling. Patient did not mentioned dry mouth or any other symptom after surgery.

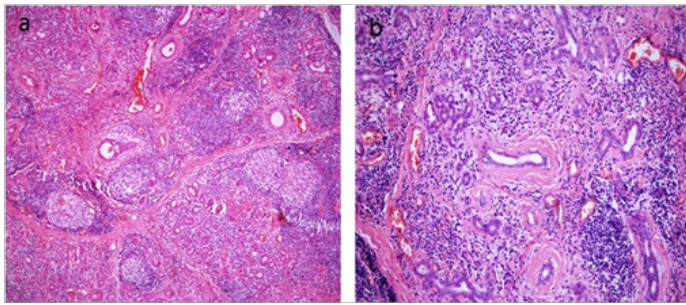


Figure 1 (a) Remarkable fibrosis, inflammatory infiltrate and lymphoid follicles, x40 H&E (b) Acinar atrophy, ductal ectasia and concentric periductal sclerosis, x100 H&E.



Figure 2 Prominent right submandibular mass.

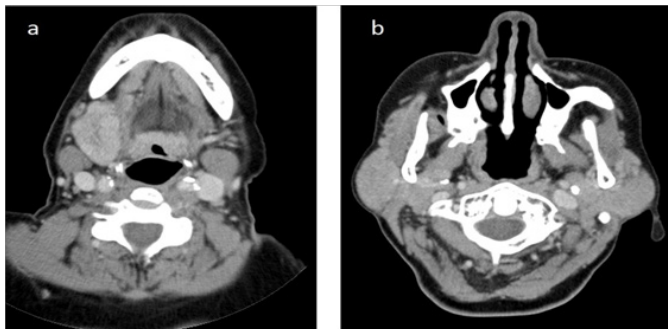


Figure 3 Axial CT scan showing the enlargement of the right submandibular gland (a) and bilateral parotid glands (b).

Discussion

KT is a chronic inflammatory process that involves the salivary glands. It is a rare entity and typically affects one of the submandibular glands. KT presents with a slow-growing, hard mass in the submandibular region and clinically mimics a neoplasm of salivary gland. This disease is classified as a tumor-like lesion of the salivary glands by the World Health Organization.⁶ The diagnosis of KT is based on histologic findings. Acinar loss, dense fibrosis, ductal dilatation and lymphoid follicle formation are the characteristic histologic features.

Several hypotheses have been suggested to explain the origin of

the KT. Sialolithiasis were demonstrated in 29%-83% of cases. Some authors state that stones cause the obstruction of salivary flow. Stasis of secretions induce the inflammation and fibrosis which resulted with chronic sclerosing sialadenitis. Whereas, others state that the formation of calculi is secondary to the sialadenitis. Seifert and Donath have proposed a theory of obstructive electrolyte sialadenitis.⁷ This hypothesis claims that a secretion disorder produce mucus plugs obstructing the small ducts and also cause changes in the electrolyte content which leads to immune process. Nevertheless, etiology is still unclear.

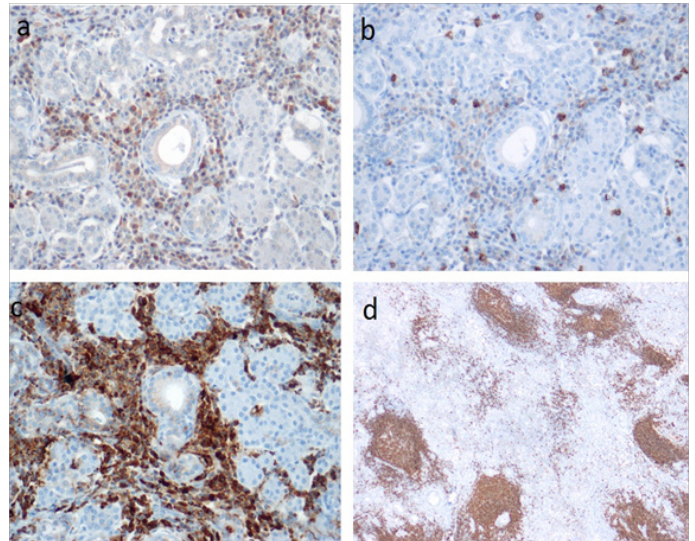


Figure 4 (a) Prominent periductal CD4+ lymphocytic infiltration, x200 CD4 immunostaining (b) Mild periductal CD8 + lymphocytic infiltration, x200 CD8 immunostaining (c) Dense plasma cell infiltration in stroma, x200 CD138 immunostaining (d) B lymphocytes in the lymph follicles, x40 CD20 immunostaining.

A histological diagnosis is necessary because KT usually present with the progressive enlargement of the submandibular gland that mimicking a true neoplasm. Histological features of KT are marked fibrosis, inflammatory infiltration, lymphoid follicles, loss of acini, periductal fibrosis and dilated ducts. Lymphocytes and plasma cells are predominant in the inflammatory infiltrate. Dilated ducts usually contain inspissated secretion. Fibrosis begins in the periductal region in an onion-skin arrangement and extends into the interlobular septa. According to Seifert,⁷ severity of inflammation in KT may progress through 4 different histological stages (Table 1).

Multiple involvement of salivary glands is an extremely rare condition and suggests that KT is closely related to an immune process.⁵ In literature, it is reported that KT may be related with the Immunoglobulin G4(IgG4)-related systemic sclerosing disease which is a chronic autoimmune disorder with diverse clinical presentation such as retroperitoneal fibrosis, sclerosing cholangitis, and sclerosing pancreatitis. Therefore, we performed investigations for the autoimmune disease. Immunocytochemical investigations on sections from the glands revealed predominately CD4+ lymphocytes and plasma cells in the inflammatory infiltrate. There was a preponderance of CD4 over CD8 lymphocytes. Prominent B-cell reaction was noticed in the lymph follicles. These findings suggested a sialadenitis related with immune process. Autoimmun antibody except the ANA were negative. IgG and IgG4 levels were in normal range. Patient was referred to an

immunologist with these findings. *But no otoimmun disorder reported by the immunologist.*

Table 1 Seifert's histological staging of KT

Stage 1	Focal chronic inflammation with accumulation of lymphocytes around moderately dilated salivary ducts that contain inspissated secretion
Stage 2	Severe periductal fibrosis and diffuse lymphocytic infiltration are more noticeable. The ductal system shows inspissated secretion and focal metaplasia with proliferation of ductal epithelium. Periductal lymphoid follicles are well developed. There is fibrosis in the centres of the lobules along with the atrophy of acini.
Stage 3	Even more prominent lymphocytic infiltration with lymphoid follicle formation, parenchymal atrophy, periductal sclerosis in addition to squamous and goblet cell metaplasia in the ductal system.
Stage 4	Diffuse sclerosis and prominent parenchymal loss cause cirrhosis-like appearance (end-stage, the "burnt-out" phase).

Possible differential diagnoses of KT include other benign lesions of salivary glands such as Kimura's disease, Sjogren's syndrome, simple chronic sialadenitis, granulomatous sialadenitis. KT is differentiated from others with prominent sclerosis and lack of epimyoe epithelial islands.

Treatment of the KT is the removal of the affected salivary gland. However, management can be conservative if the mass is asymptomatic and there is enough evidence that the lesion is benign. Cheuk et al suggested that sonography and sonographically guided needle biopsy may indicate for KT and prevent operative excision of an otherwise benign condition.⁸ However, it is usually difficult to differentiate KT from a true neoplasm preoperatively. A hard mass in the submandibular gland is highly suggestive of tumor. Therefore, the submandibular gland is most commonly surgically removed.

Kuttner's tumor is a rare benign disease that mimics true neoplasm due to presentation with unilateral firm submandibular mass. *Final diagnosis* of KT is established by *the histopathological examination.*

KT is generally *treated with surgical removal of the affected salivary gland and no further treatment is required.* However, in some cases, KT may be a manifestation of a systemic IgG4-related autoimmune disease. *Therefore,* patients diagnosed with KT should be referred to an immunologist to exclude an autoimmune disorder. Both surgeon and pathologist KT must be aware of this benign condition for the correct diagnosis of KT.

Acknowledgments

None.

Conflicts of interest

The authors declare that there are no conflicts of interest.

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