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Solitary plexiform neurofibroma of the submandibular salivary gland

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Abstract

Neurofibroma affecting the major salivary gland is uncommon. This tumour is particularly rare in the submandibular and sublingual gland.

Here, a case of solitary plexiform neurofibroma of the submandibular gland without any other manifestations of von Recklinghausen's disease is presented. It is probably the first case report of this tumour invading the submandibular gland in a solitary form.

Key words: Submandibular gland neoplasms; Neurofibroma; Neurofibromatosis 1

Introduction

Neurofibroma of the major salivary gland is unusual. Seifert *et al.* (1986) noted 120 patients with nonepithelial tumours amongst 2,700 patients with salivary gland tumours. These tumours consisted of angiomas (52.5 per cent), lipomas (18.5 per cent), sarcomas (7.5 per cent), and several other benign tumours. Patients with a neurofibroma numbered only 12, regardless of whether they had an underlying diagnosis of neurofibromatosis or not. They represented 10 per cent of 120 patients with nonepithelial tumours, and 0.4 per cent of all patients with salivary gland tumours. In another series, Castro *et al.* (1972), listed two plexiform neurofibromas among 300 cases of major salivary gland tumours in children.

Seifert *et al.* (1986), also stated that almost 90 per cent of nonepithelial tumours were in the parotid gland, with the rest in the submandibular gland. Several cases of a neurofibroma affecting the parotid gland have been reported (Byars *et al.*, 1957; Reiquam, 1963; Karlan and Snyder, 1968; Brasfield and Das Gupta, 1972; Osebold and Moore, 1979; Weitzner, 1980; Sullivan *et al.*, 1987; Kayem *et al.*, 1995), but we could trace only one case report of this tumour in the submandibular gland (Weitzner, 1980), with the diagnosis of von Recklinghausen's disease.

In this paper, a rare case of solitary plexiform neurofibroma of the submandibular gland is presented.

Case report

A 29-year-old female consulted our hospital because of repeated swelling of her left submandibular gland for 10 years. It had occurred one to two times per year and was accompanied by an attack of fever and tenderness. Usually, she recovered within several days with antibiotics.

Her left submandibular gland was mildly enlarged in comparison with the right one. Her left sublingual fold and caruncle were markedly swollen with hyperaemic change. No signs or family history of von Recklinghausen's disease

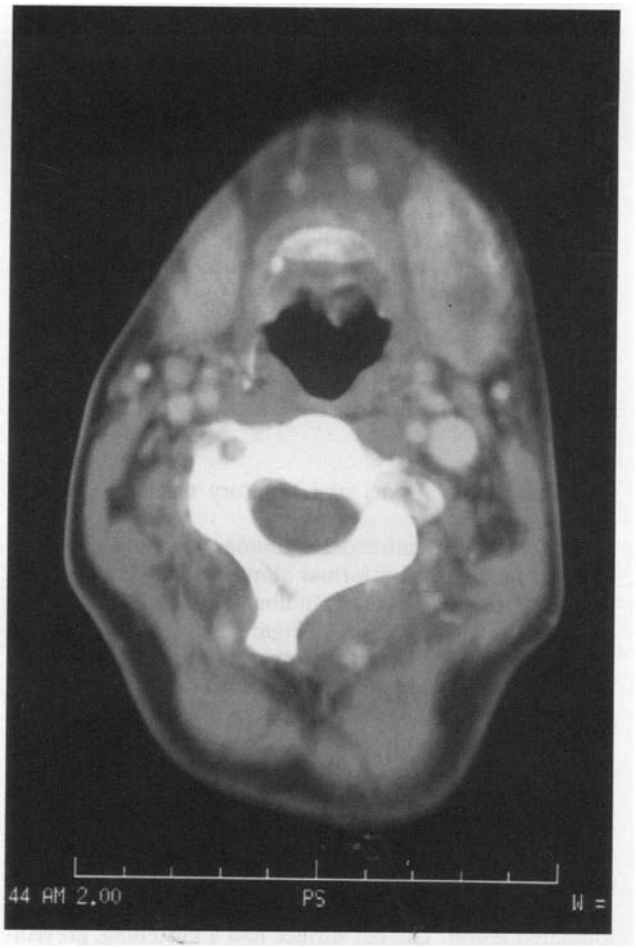

FIG. 1

CT scan showing the left submandibular gland with low density area.

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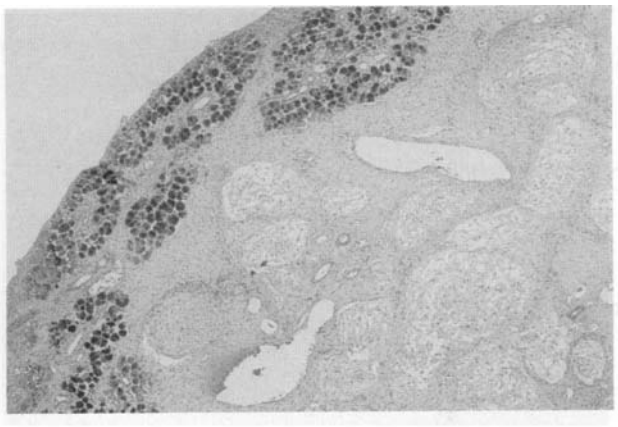
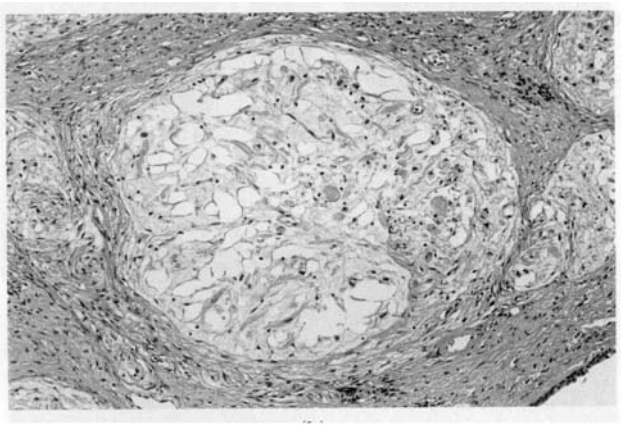


(a)



(b)

FIG. 2

Photomicrograph of the resected submandibular gland. (A): There were irregularly distorted, enlarged nerve bundles, which appeared to be multiple nodules of different sizes. These were embedded in a background of neurofibtomatous tissue. Normal tissue of the gland was seen in the circumference (H & E; × 40). (B): All nodules were well circumscribed but not encapsulated (H & E; × 100).

was found. The results of laboratory tests were within normal limits.

She underwent ultrasound tomography and CT with contrast medium. The former showed a solid and somewhat heterogeneous submandibular gland with mildly dilated ducts. The CT scan (Figure 1) showed a swollen left submandibular gland surrounding a low density area. This gland did not demonstrate enhancement with contrast medium. Fine needle aspiration of the mass was performed, but the result was nondiagnostic.

Resection of the left submandibular gland was performed under general anaesthesia. The surface of the gland was easily exposed, but anteriorly the gland invaded the layer superficial to the mylohyoid muscle. There was excessive bleeding during excision of this part.

Macroscopic view of the surgical specimen showed a lobulated lesion. The cut surface had a glistening, greyish-white coloured fleshy texture in its central portion, with lobulated edges. Histopathological examination revealed a characteristic plexiform neurofibroma permeating the whole gland (Figures 2a and b). Immunohistochemical studies showed positive staining of anti-S100 protein and antibodies to neurofilament. Normal glandular tissue was found on the periphery.

Discussion

This case may be the first of a solitary neurofibroma affecting the submandibular gland. However, the pathological diagnosis in our case was plexiform neurofibroma, which is said to be indicative of von Recklinghausen's disease even though it may be the only manifestation (Weitzner, 1980; Megahed, 1994).

Besides, neurofibroma tends to bleed when excised (McMaster, 1972; Mukherji, 1974), and incomplete excision may lead to recurrence. Littlewood and Stilwell (1983) mentioned that there is a variant of plexiform neurofibroma with a plethora of abnormal vessels which bleed excessively when incised. Our patient was thought to have this variant.

Further, sarcomatous transformation of a neurofibroma is recognized especially after surgical intervention (Hosoi, 1931; Chaudhuri *et al.*, 1980), though this has also been disputed (Preston *et al.*, 1952; D'Agostino *et al.*, 1963). So, from these points of view, careful follow-up observation will be necessary to note any recurrence or any other manifestations which may appear.

The origin of the tumour was not obvious at operation. However, there were no signs of impairment of the hypoglossal and lingual nerve and this may indicate that the tumour might have originated from the secreting nerve.

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