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Peripheral nerve sheath tumors arising in salivary glands: A clinicopathologic study

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ABSTRACT

Primary salivary gland peripheral nerve sheath tumors (PNST) are uncommon. This study is a retrospective, clinicopathologic review of 9 cases of PNST (5 neurofibromas, 3 schwannomas and 1 malignant peripheral nerve sheath tumor (MPNST)) arising from the salivary glands, encountered between 1990 and 2015. All patients with neurofibromas were male (ages 1–62 years) and had a single parotid lesion of which 2 were diffuse, 2 plexiform and one mixed diffuse/plexiform. Four had a history of neurofibromatosis I. Four of 5 presented with symptoms related to mass effect including facial swelling, facial drooping, and dysphagia. All underwent de-bulking surgery and recurred due to continued growth. Of the 3 patients with schwannomas, 1 was male and 2 were female (ages 19, 44 and 56 years). One tumor each arose in the sublingual, submandibular, and parotid glands. Two of 3 presented with soreness and swelling local to the affected gland, especially while chewing. There was no recurrence of these tumors after resection. An MPNST in a male presented as a tender mass in the patient's left parotid; the tumor was resected. There was no evidence of tumor elsewhere in the body. The tumor did not recur in 12 years of follow-up. The most common tumor type in the current series was neurofibroma; most arose in the background of neurofibromatosis type I and all of which recurred after initial subtotal resection. Most PNST arose in the parotid gland.

1. Introduction

Peripheral nerve sheath tumors (neurofibromas, schwannomas, and malignant peripheral nerve sheath tumors (MPNST)) are rarely encountered as primary neoplasms within the salivary glands. Due to their rarity in these locations, their diagnosis is usually not expected preoperatively and is only made on microscopic evaluation after resection. A single institution retrospective analysis by Cho et al found that neurofibromas and schwannoma comprised 0.2% (n = 1) and 1.1% (n = 6), respectively, of 527 mesenchymal tumors found in salivary glands [1]. Of the 22 neurofibromas [2], 9 [3–11] MPSNT, and approximately 150 schwannomas [12–22] in the literature that have been found to arise in the salivary gland, the vast majority have been reported as single case reports or small series [13].

Presented herein is a 9-case series of 5 neurofibromas 3 schwannomas and a single MPNST, which arose from salivary glands. Within this series is the first ever reported mixed morphology neurofibroma arising from the salivary gland, the second and third reported diffuse-type neurofibromas arising from a salivary gland (including the first in an adult), and the second reported sublingual schwannoma. The purpose of this report is to review the clinical and pathological features of peripheral nerve sheath tumors that arise from the salivary glands.

2. Methods and materials

Institutional Review Board approval was obtained prior to commencement of the study. Surgical pathology records were searched from 1990 to 2015 for cases of neurofibroma, schwannoma, and MPNST associated with a salivary gland. Twelve cases were discovered. Three patients of this group were excluded, since their tumors did not directly involve the salivary gland, or arose from outside of the gland. The remaining 9 patients composed the study group and had their clinical history and other pathologic findings extracted from the medical records and pathology reports. Clinical information was unavailable in 1 patient.

3. Results

3.1. Neurofibromas

Five patients in this series had neurofibromas (Table summarizes the clinicopathological features of all cases in this series). This group was exclusively composed of men, 4 of whom had a history of Neurofibromatosis I (1 patient did not have a medical record available for review).

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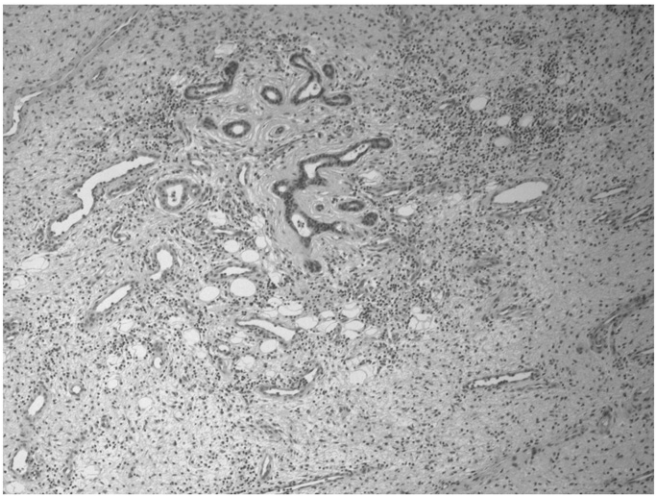

Fig. 1. Case 3. Low magnification appearance of a diffuse neurofibroma associated with chronic inflammation, replacing most of the salivary gland (hematoxylin and eosin, original magnification ×100).

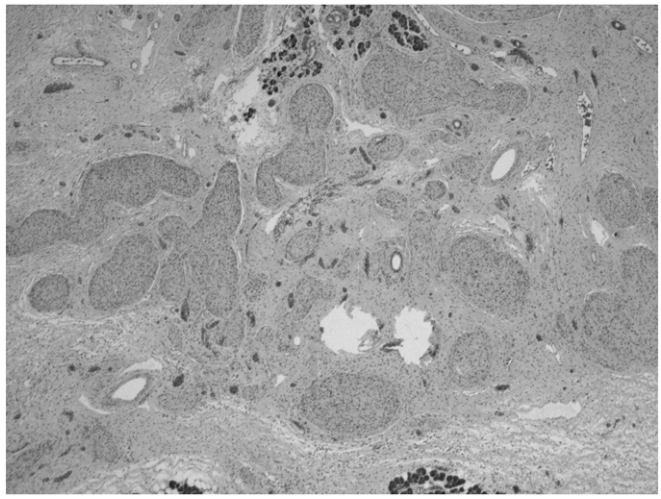

Fig. 3. Case 5. A plexiform neurofibroma marked by multiple nodules of tumor within the parotid gland (hematoxylin and eosin, original magnification ×50).

The patients' ages ranged from 1 to 62 years and all tumors arose in the parotid gland, with 3 affecting the left side. In 4 cases, the patient's presented with significant facial deformity and associated mass effect symptoms such as pressure headache, dysphagia, and difficulty speaking. In all patients, imaging revealed a unilateral lesion within the parotid gland with sizes ranging from 3.8 to greater than 10 cm.

Four patients had a subtotal resection to relieve their symptoms, and the final patient underwent incisional biopsy. Surgery resulted in immediate alleviation of symptoms in the 4 patients with known history. No postoperative complications were reported, and all of these patients are currently alive (follow-up ranging from 5-26 years). However, in all cases, the mass continued to grow following the initial surgery, resulting in recurrence of facial droop. Two patients in this group required a revision subtotal resection of the same lesion due to the new mass effect symptoms, with 1 patient also experiencing invasion of the mass to a new area of the face. A single patient underwent incisional biopsy at 1 year of age followed by annual computed tomographic (CT) screening of his intraparotid neurofibroma; in the intervening 15 years of follow-up, this mass grew at a rate proportional to the patient's body, causing greater facial deformity, difficulty chewing and speech impediment. Subsequent speech therapy successfully resolved the latter symptom. At last follow-up, this lesion seems to have a slowed growth trajectory.

Upon microscopic evaluation, 2 cases in this group showed a nonencapsulated mass containing interlacing bundles of elongated cells with wavy nuclei consistent with a diffuse type neurofibroma (Figs. 1 and 2). Two patients' lesion contained nodules of tortuous, expanded nerve bundles consistent with plexiform neurofibroma. The final patient had pathological features of both morphologies (Fig. 3). Patients 1 and patient 4 lesions also contained evidence of organizing hemorrhage with hemosiderin.

3.2. Schwannomas

Three patients in this series had schwannomas that arose within various salivary glands: a 19-year old female with 5.0 cm lesion in the left submandibular gland, a 44-year-old male with a 4.5 cm lesion in the left parotid gland, and a 56-year-old female with a 0.2 cm lesion in the

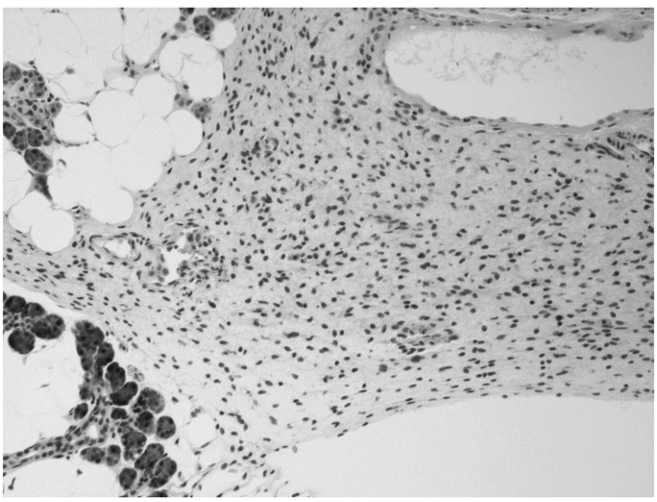

Fig. 2. Case 4. Higher magnification appearance of a diffuse neurofibroma infiltrating into the parotid gland (hematoxylin and eosin, original magnification ×200).

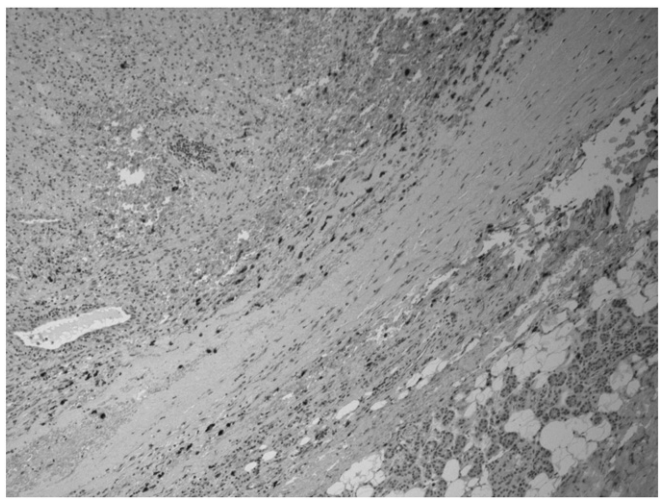

Fig. 4. Case 7. Low magnification showing an encapsulated schwannoma (upper left) arising in the left parotid gland (lower right) (hematoxylin and eosin, original magnification ×100).