Abstract

Solitary neurofibroma arising from facial nerve of parotid glands are very uncommon. These tumours are known to arise from schwann cells. This report is a rare case of a 35-year-old male patient who complained of swelling in parotid gland region without any symptoms of pain, trismus and facial paralysis. Fine needle aspiration cytology (FNAC) was performed on the patient, followed by ultrasonography, and a provisional diagnosis of pleomorphic adenoma was rendered. Superficial parotidectomy with en bloc resection of the tumour mass conserving the facial nerve was done. Histopathologically, a diagnosis of neurofibroma of parotid gland was given. In this paper, the incidence, presentation of the tumour, histopathology and the surgical aspects of the tumour has been discussed. This case is presented due to the rarity in site and its presentation without any clinical features of neurofibroma. A provisional diagnosis of benign nerve sheath tumours should be considered for patients with parotid mass and pain.

Key words: Facial nerve, neurofibroma, parotid gland

Case report

Introduction

The incidence of solitary tumours at intra-parotid involving facial nerve are rare, and comprise of 0.2 to 1.5 %.1 Of all the salivary gland lesions, neurofibromas (NF) are rare and constitute to only 0.4 %.2 They are conventionally presented as slow growing, painless or a painful mass in the parotid gland with local infiltration. Most of the neoplasms are of long standing duration with or without paresis, paralysis or spasm involving the facial nerve.3 Magnetic resonance imaging (MRI) and computed tomography (CT) scan imaging helps us in diagnosing the extent of the tumour. However, histopathology remains to be the gold standard for diagnosing neurofibromas.

Case description

A 35-year-old male reported with a chief complaint of swelling in the right parotid region, which was slowly progressing in size. The patient did not report of pain or increase in size of swelling. A firm well defined swelling in the parotid region of size 3.5×3 cm was noted on examination. Ultrasound scan revealed a well circumscribed hypoechoic solid nodule measuring about 3.3x3.0x3.0 cm at the periphery of the right parotid gland with minimal vascularity. Correlating the findings of the ultrasound with FNAC, a provisional diagnosis of pleomorphic adenoma involving the parotid gland was given. A right superficial parotidectomy was performed, which revealed the main trunk of the

facial nerve (Figure 1). Post-operative transient period of neuropraxia was seen; however, the patient recovered completely after three months. Grossly, the specimen appeared nodular, firm in consistency and cut surface showed tan yellow-white appearance (Figure 2). Histopathologically, the section showed both hypo and hyper cellular areas with predominantly spindle cells arranged in sheets and fascicles, having elongated wavy nucleus and moderate cytoplasm. No cellular atypia was present (Figure 3). Intervening stroma showed chronic inflammatory cell infiltrates, myxoid areas and thick hyalinized blood vessels (Figure 4). Salivary gland component in another block appeared unremarkable. Sections were negative for granulomas or malignancy. Tumour cells demonstrated positivity for S-100 immunohistochemically (Figure 5). Features were suggestive of NF of parotid gland.

Figure 1: Intraoperative view - The main trunk of the facial nerve, which could be traced to the buccal branch. The tumour was located within the parotid gland at the level of the facial nerve.

Figure 2: Gross specimen of Neurofibroma showing yellow-white appearance on cut surface

Figure 3: Neurofibroma showing hypo and hyper cellular areas with tumour cells surrounding the ductal component

Figure 4: Neurofibroma showing myxoid areas with neoplastic cells and ropey collagen bundles

Figure 5: S-100 protein immunostain of neurofibroma showing positivity in the tumour cells
Discussion

Nerve sheath tumours involving salivary glands are usually rare and comprise of 0.2–1.5 % of intraparotid tumours. NF can develop as solitary or multiple lesions associated with neurofibromatosis I or II syndromes in 10 % of cases. These lesions can occur anywhere along the path of the facial nerve, from the cerebellopontine angle to its branches. They are mostly painless, slow growing nodules present as localized, diffuse or plexiform lesions, but occurrence at intraparotid region, are usually noted for their rarity. Malignant transformation of NF occurs in 2%-5% of patients with NF 1 as compared to an incidence rate of 0.001% in the general population. The first case of intraparotid NF was reported by Maxwell in the year 1951, following which Katz et al in 1971 reported another case. A review done by Nussbaum et al on approximately 700 parotidectomies, two cases of neurofibromas were evident in his survey. NF was seen in one case out of the 56 primary neurogenic neoplasms, which were involved the facial nerve in a study done by Sullivan et al.

NF displays a spectrum of cell types ranging from the Schwann cell to the fibroblast. Definitive diagnosis requires mainly histological evaluation, but it is not advisable to perform incisional biopsy in major salivary glands. Adhesive nature of cells in the tumour, cell poor or collagen fiber rich nature of NF, and the position at which the needle is inserted in the tumour may vary, and hence one may find it difficult to get a positive cytology, making the use of FNAC rare for diagnosis of such lesions. MRI and CT are usually the recommended diagnostic tests. Histopathological examination of NF shows proliferation of elements of nerve, which includes Schwann cells, axons and fibroblasts. Neurofibromas are mostly characterized by spindle shaped cells arranged in whorls, short fascicles or storiform pattern. In between, the cells appears as a loosely textured collagenous matrix which has a “shredded carrot” appearance, and may also assimilate the nerve fibres within the tumour matrix. Sometimes, these tumours are highly myxoid, and resembles myxomas. S-100 protein can be identified in these tumours by immunohistochemistry, but is focally positive.

Management of benign facial nerve sheath tumours generally depends on the age of patient and its association with facial nerve function. Simple excision of the tumour is considered as adequate therapy. NFs assimilate nerves and are generally resected by enbloc with the involved nerve. Prognosis of the patient is good, but reoccurs if not excised completely.

Conclusion

Solitary NF of the parotid gland involving the facial nerve is a rare tumour. The preoperative diagnosis of this tumour is very difficult due to its rarity, type of presentation and behaviour, and the involvement of the facial nerve. Surgical excision with facial nerve grafting is an option for patients with large and aggressive tumours. Recurrence occurs in incomplete excision of tumour, and complications like neuropraxia and facial nerve paralysis can be seen. Although not common, these benign neurogenic tumours of the facial nerve should be considered during provisional diagnosis in a patient involving a parotid mass.

References


**Acknowledgments and disclosure statements**

Acknowledgments: None

Conflict of Interest: None