Case Report

Schwannoma of 7th Cranial Nerve: A Case Report.

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Abstract: Schwannomas are benign tumors that arise from the nerve sheath. Facial nerve schwannoma also known as facial nerve neuroma/neurilemoma, is a schwannoma that arises from the Schwann cells, originating from the surface of the facial nerve. The vestibular schwannoma, also known as the acoustic neuroma, is the most frequent type of nerve cell tumor of the head and neck region, though facial neuromas are extremely rare. The most common clinical presentation is facial neuropathy (42%). Schwannomas associated with the parotid gland are rare neoplasm. In this brief case report, we have described a rare case of schwannoma of the parotid gland in a young female aged 12 yrs.

Keywords: Facial nerve; Schwannoma; Schwann cell; Parotid gland; Endoneurium; Tumor.

INTRODUCTION

The tumors of the peripheral nerves are called neuromas, defined as growths / swellings on nerves. Neuromas may arise extrinsically or intrinsically. Primary intrinsic nerve cell tumors include schwannomas, which arise from Schwann cells that support the endoneurium, as well as perineuromas, which arise from cells that line the perineurium. Fifty percent of all neuromas are found in the head and neck region.[1] Schwannoma is an ectodermal benign encapsulated tumor, which are extremely rarely seen along the facial nerve and can arise anywhere along the course of this nerve. Schwannomas of the parotid gland are rare neoplasms.[2]

Approximately, 25-30% of all reported schwannomas occur in the head and neck and most of these in the eighth nerve.[3] Eneroth and Hamberger[4]could demonstrate two cases with neurogenic origin and in a review of 700 parotidectomies, amongst 802 parotid tumors, Nussbaum[5]found only one case of neurilemmoma of the facial nerve.

CASE REPORT

A 12 year-old female patient, presented with a gradually progressive, painless swelling in the right preauricular region. Patient denied any facial weakness, twitching or pain. On palpation, there was a well-defined 15-20 mm, fusiform, oval, firm, nontender, mobile swelling in right parotid region. The history of duration was 3yrs, and slowly progressive in nature. On palpation there was a non tender firm swelling and not fixed to underlying structures. Excisional biopsy was done without damaging the facial nerve. During surgical excision the tumor mass seemed like eccentric mass arising from the facial nerve. The tumor mass was submitted to the department of Oral Pathology, Rama Dental College, Hospital and Research Centre.

The gross specimen was oval to fusiform in shape with a smooth surface and whitish to creamish in colour, measuring about 5 X 2.5cm (Figure 1). The cut surface showed the capsule lining, with nodular or whorl-like appearance (Figure 2).

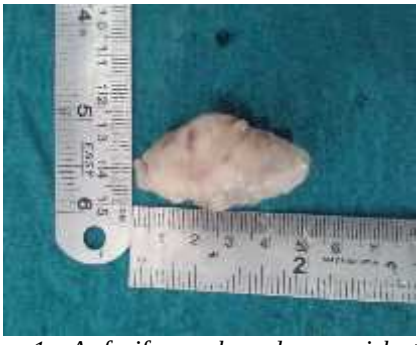

Figure 1: A fusiform shaped creamish tumor mass.

Figure 2: The cut surface showing capsule and whorled lobules.

On histopathologic examination, the H & E stained section revealed the presence of a capsulated tumor with palisaded spindle cells in the form of whorled structures forming typical Antony type A structures along with Verocay bodies. Haphazardly arranged Schwann cells in the form of Antony B were also seen. The features were appropriate for the diagnosis of Schwannoma (Figure 3).

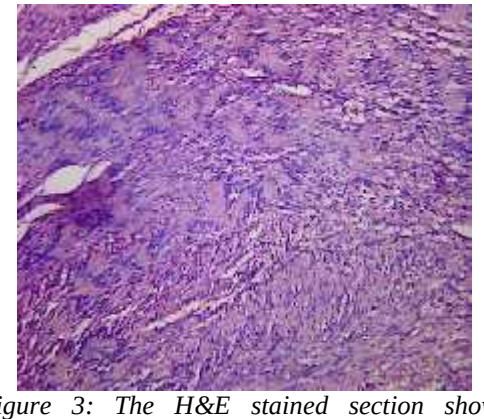

Figure 3: The H&E stained section shows palisaded spinlle cess in Antoni type A arrangement and hapazard distribution os spindle cells in Antoni B areas (100X).

DISCUSSION

Facial nerve schwannoma (FNS), also known as facial nerve neuroma/neurilemoma, is a schwannoma that arises from the facial nerve. It is a slow growing encapsulated tumor of neuroectodermal derivation that originates from the Schwann cells of the neural sheath. They are rare lesions that can arise anywhere along the course of the facial nerve, from its origin in the cerebellopontine angle to its extracranial ramifications in the parotid space of the extracranial head and neck,[6,7] that is from either the extratemporal or intratemporal course of this nerve. Most of these tumors are intratemporal, whereas 9% are located extracranially and usually appear as an asymptomatic parotid mass. Intraparotid facial nerve schwannomas account for only two of 142 parotid tumors.[8] As schwannomas elsewhere, they originate from the surface of the nerve, and displace and splay the nerve fibres over their eccentric growth. They are generally uncommon, and when involving the temporal bone, make up less than 1% of all temporal bone tumours. Schwannomas are relatively slow growing, mostly benign and less than 1% show malignant transformation into a form neurofibrosarcoma. The risk of malignant transformation of head and neck schwannoma varies from 8 to 10%.[9]

The vestibular schwannoma, also known as the acoustic neuroma, is the most frequent type of nerve cell tumor of the head and neck. Vestibular schwannoma (or acoustic neuroma) affects the nerve that connects the brain to the inner ear, affecting sense of balance. Facial neuromas are extremely rare and tend to involve multiple facial nerve segments. Some studies report the geniculate ganglion as the most frequently involved portion of the nerve. Unlike vestibular schwannomas, no genetic locus has been implicated in the pathogenesis of facial schwannomas.[2]

Schwannomas can arise from a genetic disorder called neurofibromatosis. In a population based study of schwannoma, 90% were sporadic, 3% cases occurred with NF2, 2% with schwannomatosis, 5% in association with meningiomas in patients with or without NF2. About 60% of sporadic and NF2 associated schwannomas have inactivating mutations of the NF2 gene. These mutations are accompanied by inactivation of the remaining wild type allelle on 22q. Nevertheless, all schwannomas, whether sporadic or syndromic, have the protein product, merlin.[10]

These tumors can present with a wide variety of symptoms. Most symptoms of facial nerve dysfunction due to a neoplasm are caused by nerve compression, secondary to tumor growth. Therefore, a relatively small tumor can become symptomatic if it arises within a narrow bony canal, while a more proximal tumor within the cerebello pontine angle can become quite large before causing symptoms.

There is no gender predilection, and these tumors may arise at any age, the peak incidence is between the third and sixth decades.[11] The present case is rare in terms

of age of occurrence, as this a young female of 12 yrs only. Patients may present with a painless, palpable facial mass. The presence of facial paralysis is variable, frequently with gradual, and often incomplete, facial nerve palsy. Rapid onset mimicks somewhat to Bell palsy. Mass effect on adjacent nerves may cause sensory neural hearing loss (SNHL) or even conductive hearing loss if growth into the middle ear impairs the normal function of the ossicles. In a minority of cases (10%) the tumour is extra-cranial, where it presents as an asymptomatic parotid mass. The difficulty in establishing a correct preoperative diagnosis has been pointed out by Conley and Janecka[12] because this tumor is infrequent and generally unsuspected as preoperative facial nerve paresis is unusual. The patient in our case was also asymptomatic. She did not complain of any pain or paresthesia.

A study by Doshi et al on 28 patients with facial nerve schwannomas found19 patients (68%) had tumor most affecting the facial nerve segment running through the internal auditory canal. They also found that 46% of patients had multisegmental schwannomas. Hearing loss and facial weakness were the most common presentations, with the latter being most frequently linked to labyrinthine segment involvement (89%).[2] Schwannomas are homogeneous tumors, consisting only of Schwann cells. The tumor cells always stay on the outside of the nerve, but the tumor itself may either push the nerve aside and/or up against a bony structure (thereby possibly causing damage).

Distinctive pathologic features of schwannoma includes a dimorphic growth pattern comprising of cellular (Antoni A) and loose-textured (Antoni B) areas, Verocay bodies and hyaline blood vessel. Antoni A regions consist of palisading trabeculae of spindle-shaped Schwann cells.[13]The structure as a whole, including a central stromal region, is known as a Verocay body. By comparison, the Antoni B regions are loose and hypocellular, and the Schwann cells appear polymorphic. The present case showed the typical Antony A and B arrangements.

The schwannomas exclusively consist of Schwann cells. Ultra structurally, these cells have attenuated cell processes that emanate from the cell body and lie in undulating layers adjacent to cell body. The cytoplasm contains flattened invaginated nucleus, microfibrils, occasional lysosomes and scattered mitochondria. Antoni B areas show increased number of lysosomes and myelin figures with fragmentd basal lamina, suggesting these are degenerated Antoni A areas.[14] On IHC, S-100 protein shows strong positivity for neurilemmomas, reflecting their propensity of Schwann cells[13]. They also show positivity for vimentin, Leu-7 antigen and glial fibrillary acidic protein.[15]Nuclear staining for SOX10, a marker for neural crest differentiation, has proven to be an excellent marker for schwannomas.[14] On IHC, S-100 protein shows strong positivity for neurilemmomas, reflecting their propensity of Schwann cells.[13] They also show positivity for vimentin, Leu-7 antigen and glial fibrillary acidic protein.[14]

The tendency to misdiagnose schwannomas and to overestimate their grade makes schwannomas noteworthy. Considering their morphology, clinical associations and behavior, peripheral nerve sheath tumors are among the most varied of human neoplasm and very often, are subject to frequent misdiagnosis.

This is particularly true of the spectrum of schwannomas which include: a) conventional schwannoma, a histologically benign tumor which, on occasion, is destructive of surrounding osseous structures, b) the relatively recently described cellular schwannoma, a tumor that histologically simulates malignant peripheral nerve sheath tumor (MPNST), c) plexiform schwannoma which, particularly in cellular form and when occurring in childhood, simulates MPNST, and d) melanotic schwannoma which is often mistaken for melanoma.

The psammomatous form of the latter is often associated with Carney complex, a rare heritable disorder that includes cutaneous lentigines, myxomas of skin, subcutaneous

tissue, and heart, and endocrine neoplasms.[16] Long standing cases show degenerating changes like cyst formation, calcifications and haemorrhage. They show marked nuclear atypia. Preoperative diagnosis of this tumor in the parotid gland is generally difficult because of the low frequency of the disease and few typical signs associated with it.

The estimated frequency of parotid tumors originating in the facial nerve ranges from 0.2% to 1.5%.[17] In addition, preoperative diagnosis of a parotid tumor as a schwannoma is difficult in the absence of facial nerve dysfunction,[18] which is an uncommon feature when the tumor arises from the facial nerve trunk. Asymptomatic cases may gradually lead to extensive involvement and serious complications. Hence, it is difficult to establish a correct preoperative diagnosis for facial nerve schwannoma. With early diagnosis of parotid schwannoma, management of patient can be planned and ultimately, facial nerve function can be optimized. Enucleation with nerve preservation is preferred choice to offer better facial function, whereas nerve excision with cable graft may be performed with satisfactory results. Facial nerve integrity may be spared in rare occasions, but more frequently nerve reconstruction is required. Final facial function recovery is mainly dependent on the preoperative presence of facial nerve deficit and its duration.[19]

CONCLUSION: In brief, schwannoma of the parotid gland is rare and may be mistaken as pleomorphic adenoma, a common parotid tumor.[20] Although most patients with intraparotid schwannomas do not present with facial nerve palsy, postoperative facial nerve paresis or palsy is common, and these patients can be better informed of this complication before surgery.

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