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Case Report

Schwannoma arising from hypoglossal nerve in the submandibular triangle: a case report

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ABSTRACT

Hypoglossal schwannomas usually develop in the intracranial portion of the brain. The incidence of hypoglossal schwannomas of the submandibular region is extremely rare. Here we report a case of schwannoma arising from the extracranial hypoglossal nerve in the left submandibular region.

Keywords: Schwannoma, Submandibular region, Hypoglossal nerve

INTRODUCTION

Schwannomas are also known as neurilemmomas or neurinomas are benign neoplasms derived from schwann cells. Schwannomas have a predilection for sensory nerves.[1] They may arise from any peripheral, spinal or cranial nerves except optic and olfactory nerves. Approximately one third of cases of schwannomas present in the head & neck region[2] and in that lateral cervical region being the most common site outside the cranium.[3] Most common cranial nerve from which schwannoma arises is vestibulocochlear nerve, others being trigeminal, facial, glossopharyngeal and vagus nerves.[4] Schwannomas arising from hypoglossal nerve are rare because it consists only motor component. Schwannomas if arise from hypoglossal nerve are usually develop in the intracranial portion or both in the intracranial & the extracranial components forming a dumb-bell shape.[5] Schwannoma arising from peripheral segment of hypoglossal nerve is extremely rare.[6,7] Here we are reporting a case of scwannoma arising from peripheral segment of hypoglossal nerve in the left submandibular space.

CASE REPORT

A 50 year old female presented to our OPD with complaint of swelling in the left submandibular region since 1 year. It was insidious in onset, gradually increased in size. There were no symptoms of pain, no purulent discharge from the floor of mouth, odynophagia, dysphagia, weight loss or night sweats. Her past medical, personal and family history were unremarkable. On examination 2 x 2 cm globular swelling present in the left submandibular region with smooth surface and firm in consistency. It was free from skin and underlying structures. It was bidigitally palpable. On USG imaging 3 x 1.5 x 1.5 cm homogenous lobulated well defined mass in the left submandibular gland consistent with benign submandibular gland neoplasm. FNAC gave differential diagnosis of myoepithelioma / myoepithelial adenoma / monomorphic adenoma of submandibular salivary gland. Intraoperatively a well-defined mass of size 3 x 2 x 2 cm present in the left submandibular triangle. Excision was done by identifying and securing hypoglossal and lingual nerves.

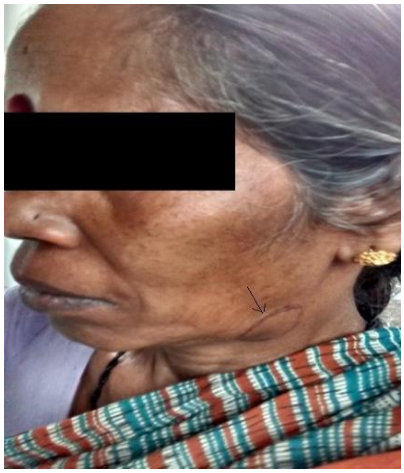

Figure 1: Pre-operative.

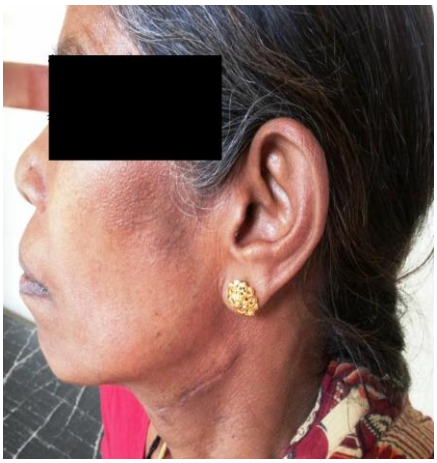

Figure 2: Post-operative.

Post operatively patient developed transient tongue deviation to left side which was subsided in 6 weeks. Histological examination shows predominant Antoni B areas, blood vessels with foci of myxoid stroma, perivascular hyalinisation and Antoni A pattern features are consistent with schwannoma.

DISCUSSION

Schwannomas are usually solitary, slow growing, well encapsulated tumours composed of neoplastic schwann cells in a collagenous matrix. Schwannomas which are non-encapsulated, pedunculated form is also recognized in the oral cavity.[8] Schwannomas are a component of Neurofibromatosis 2 and even sporadic ones are also commonly associated with inactivating mutations in the NF2 gene on chromosome 22. Loss of expression of the NF2 gene product merlin, is a consistent finding in all schwannomas.[9] Recently a condition known as schwannomatosis was described in which multiple schwannomas in peripheral, spinal and cranial nerves are present in the patient. It is associated with mutation of INI1 gene located on chromosome 22q at a separate locus from the NF2 gene.[10] Schwannomas arise from a small number of fascicles with in the nerve and as it grows it pushes the nerve of origin.

Schwannomas can present at any age but most commonly between 20-50 years. Both sexes are equally affected. They are usually asymptomatic but focal neurological symptoms and signs may be associated with nerve compression. They are usually less than 4cm diameter in the head and neck region, although tumours as large as 10 cm have been reported a finding usually associated with neglect and late presentation or intra lesional hemorrhage.[11] Malignant transformation is extremely rare in schwannomas. Only a single case was documented in the scientific press.[12]

Schwannomas form firm, gray masses that may have areas of cystic and xanthomatous change. On microscopic examination tumors show a mixture of two growth patterns. In the Antoni A pattern of growth, elongated cells with cytoplasmic processes are arranged in fascicles in areas of moderate to high cellularity and scant stromal matrix; the “nuclear-free zones” of processes that lie between the regions of nuclear palisading are termed Verocay bodies. In the Antoni B pattern of growth, the tumor is less densely cellular and consists of a loose meshwork of cells, microcysts and myxoidstroma. In both areas the individual cells have an elongated shape and regular oval nuclei. Electron microscopy shows basement membrane deposits encasing single cells and collagen fibers. Because the lesion displaces the nerve of origin as it grows, silver stains or immunostains for neurofilament proteins demonstrate that axons are largely excluded from the tumor, although they may become entrapped in the capsule. The Schwann cell origin of these tumors is borne out by their S-100 immunoreactivity and schwannomas will have strong and homogenous uptake of S-100. A variety of degenerative changes may be found in schwannomas, including nuclear pleomorphism, xanthomatous change, and vascular hyalinization.

Diagnostic investigations include Computed Tomography (CT), Magnetic Resonance Imaging (MRI), ultrasound scan and FNA. MRI is the best choice in detecting the extent of the tumor and correlates well with the operative findings.[13] Schwannomas have specific MRI properties, including specific signs (split-fat sign, fascicular sign, target sign) and signal patterns that is, isointense T1 signal relative to skeletal muscle; increased and slightly heterogeneous T2 signal.[14] Surgical excision is the treatment of choice since they are encapsulated and require little more than enucleation to ensure adequate clearance margins. Non encapsulated forms however require a cuff of normal tissue if recurrence is to be minimized.[15]

Nerve preservation is important since a neurilemmoma is benign and slow growing, and further surgical exploration may not be required. It should be noted that radiotherapy is not effective in these tumors.

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