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Extra cranial Schwannomas of Head and Neck

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Abstract:

Schwannomas are benign slow growing tumors that arise from the Schwann cells of any nerve (peripheral, cranial or autonomic). It also called nerilemmona, neurinoma spindle cell tumor etc. Extra cranial Head and Neck schwannomas usually present as solitary and well demarcated lesions. The lesion can cause secondary symptoms such as nasal obstruction, dysphagia and hoarseness, depending upon the location of the lesion. All patients with head and neck schwannomas treated in our department from April 2008 to dec.2010 were reviewed. Most of cases (3 cases) presented with neck swelling. Preoperative diagnosis may be aided by FNA, computed tomography and MRI. The main stay of treatment is complete intracapsulor excision with preserving the nerve of origin.

Key Words: Schwannoma, nerilemmona

#### Introduction:

Benign nerve cell tumors have been given various names like schwannoma, nerilemmona, neuroma, neurofibroma, spindle cell tumors etc. Schwannoma of head and neck region are not uncommon but they are rarely reported. About 25-45% of all the extra cranial schwannoma have been reported in the head and neck region(1). Head and Neck region is the site of origin in more than one third of all solitary neurilemmoma as and they occur more often the lateral part of neck(2). These are reported in the parapharyngeal space,retrophargeal space, posterior pharyngeal wall,paranasal sinuses, nasal cavity, sub mandibular region, oral cavity (3,4.5). Four cases of extracranial schwannoma presented to ENT Dept. Govt Medical College, Hassan Karnataka state during the period April 2008 to 2010. These presented with lateral neck masses, lateral pharyngeal wall and nasal masses are reported and discussed. The incidence of age between 20 years to 50 years.

### Case; No:1

A 46 yrs old female patient presented with right neck mass and foreign body sensation since 1 year. There was no h/o loss of appetite, loss of weight or trauma.

On examination the Neck there was smooth mass on right carotid triangle which moves in horizontal plane. But there is restriction of the movement on vertical plane. Computed tomography showed a well-defined mass with anterior displacement of the common and internal carotid artery. On contrast there is heterogeneous enhancement.

Surgical excision was planned under general anesthesia through transcervical route. A horizontal incision was put mass was separated from the surrounding structure. Mass was excised in to Blunt dissection was done with the finger. No significant bleeding was seen. Post operative period was uneventful. Histopathological report showed interlacing bands of Spindle cells and palisade of nuclei at places. Suggestive of neurilemmonas.

#### Case No.2

A 20yrs old female presented with an asymptomatic swelling in the left side of the neck since 1 year. Examination showed 4X3 cm swelling in upper 1/3<sup>rd</sup> of Sternocleidomastoid muscle which was non tender, firms and mobile in the transverse direction. On computed tomography there was encased heterogeneous mass left side of neck below the carotid triangle Major vessels pushed medially FNA was inconclusive.

Surgical excision was done by horizontal incision. Mass was excised in to the Nerve of origin could not made out. Post operative period was uneventfull. Specimen showed on histopathological examination as Schwannoma.

#### Case no.3:

A 25 years old female presented with swelling in the left Submandibular area since 1 yr 4 months. History of dysphagia was since 6 month. Change in choice since 3 months. On examination, there was smooth mass in the left Submandibular area measuring about 6mX3cm, non tender, firm in consisteny, bidigitally not palpable. On oral cavity examination, there is bulge from the lateral pharyngeal wall of oropharynx(fig no 1), which was extending up to the left AE fold.

CT scan showed a wall defined oval shaped post styloid Para pharyngeal solid mass lesion(fig no 4).

On per oral FNA was done, It showed features of spindle cell lesion.

Surgical excision was planned by external route. Transverse cervical excision was put, tumor was separated laterally inferiorly from the surrounding structure, tumor was separated superiorly by blunt dissection, tumor was excised in toto(fig no3), Post-operative period was uneventful. Histopathological examination showed schwannoma.

#### Case No: 4

A 36 years old male patient presented with right side progressive nasal obstruction since 2 years.

On examination there was polypoidal mass seen Rt nasal cavity. Which was extending up to the inferior turbinate. On probe examination mass was arising from the lateral wall of nose. CT scan records:-soft tissue density polypoidal lesion at right maxillary sinus extending into ipsilateral, nasal cavity with extension into controlateral nasal cavity and Rt orbit.

With suspicious of inverted papilloma, biopsy was taken endoscopically and sent for HPE. Histopathological examination showed features suggestive of Schwannoma with degenerative change.

With extension of the tumour, we planned for medical maxillectomy, Mass was excised in toto through the medical maxillectomy. Post operatively the eye movements were normal Histopathological examined confirmed the diagnosis.

#### Discussion.

The study of literature showed that the neurogenic tumors arise from the neural crest which differentiate into the Schwann cells and sympathicoblasts. The Schwann cells give rise to neurofibroma and neurileumoma(Schwannoma)(6). A Schwannoma is a slow growing solitary and encapsulated tumour attached to a nerve. A Schwannoma shows degenerative changes such as cystic alterations and hemorrhage necrosis whereas changes are not seen in neurofibroma(7). Schwannoma may arise from any cranial or spinal nerve that has a sheath i.e. any motor or sensory nerve other than the optic and olfactory nerves which do not have the Schwann cell sheath. Schwannoma was first established as a pathological entity by verocay in 1908 (WHO lateral called it neurinoma in 1910) later the term neurilemmona coined by Stout in 1935(8). Approximately 25-45% of all the reported neurilemmomas occur in the head and neck and most of them are in the eight nerve(9). The most common site of the extracranial Schwannoma in the head and neck region in the parapharyngeal space(10,11). The first case of parapharymgeal space was reported in 1933. Other site in the head and neck like submandibular space, paranasal sinus, oral cavity etc are rare (12).

The size of tumor may varies from few mm to over 24 cm. The clinical sites and symptoms may vary according to the anatomic site of the tumor in the head and neck. Majority of the patients presents with painless mass. Other symptoms may be difficulty in breathing (Nasal), dysphagia(pharynx).hoarseness(larynx). The swelling is most often freely mobile in soft tissues, but when it is connected to a large nerve or trunk there is restriction of the neck swelling, one patient presented with nasal obstruction.

The Schwannoma may arise at any age and there is no gender and race prediction (8). But in our cases, majority were female. Most of tumors nerve of origin is usually not identifiable (2). In our cases we identified nerve in two cases.

In the nose and Para nasal sinuses, these tumors arise from the intranasal nerves(7). In the neck, Schwannomas are divided into medical and lateral groups on the basis of nerve origin. The medical group arises from the last four cranial nerves and cervical sympathetic chains, the lateral group arises from the cervical, neck trauma, cervical plexus and the brachial plexus. Most of tumors in the parapharyngeal space arises from vagus nerve (13).

The pre-operative diagnosis of Schwannomas in the head and neck region is difficult. FNAC should be recommended after computed tomography. Compute tomography advised in all cases of Schwannomas (2, 7) CT will differentiate between vascular and non vascular tumor. After confirmation of non vascular tumors, FNA is advisable; FNAC is very effective in differentiating benign and malignant tumors of soft tissue. All though FNAC is very useful in most neck masses, it has low accuracy in the diagnosis of neural tumors.

Computed tomography with contrast enhancement should be done it differentiate, Schwannomas from carotid body and Glomus vagale tumors, because the distinction may influence treatment planning. Carotid body tumors arise at the carotid bifurcation, splaying the external and internal carotid arteries, whereas Glomus vagus tumors usually displace the internal carotid artery anteriority or medially or both. Both tumors enhance both CT and MR images and reveal a characteristic 'salt and pepper 'appearance on enhanced T1-weighted MR images became of flow voids frequently noted within the mass. This salt and pepper appearance is not a feature of Schwannomas.

Complete surgical excision is the treatment of choice for all, Schwannomas. Recurrence after successful enblock removal of the tumor is very rare (14). Macroscopic appearance shown in fig no 5.

Histological it exhibits two main patterns, Antoni A and Antoni B.Antoni A tissue is represented by a tendency towards palisade of the nuclei about a central mess of

cytoplasm (verocay bodies) in contrast, Antoni B tissues is a loosely arranged stromas in which the fibers and cells form no distinctive pattern other typical features include necrosis, hemorrhage and cystic degeneration.

The tumor is radio resistant and the possibility of the malignant degeneration of the benign tumor is extremely rare. Radiotherapy should be reserved for palliation when surgical management is impossible (2).

In our study the male: female ratio has been 3:1 but this may not been significant as numbers of cases has been less. The nerve of origin could be traced in two cases. The tumors cervical approaches is better approach in all neck. Schwannoma as in our cases than oral approach, because Trans oral approach offers direct route to tumours presenting in the oropharynx but provided no control of the great vessels(15). Trans oral FNA is better option when tumors has got more oropharyngeal presentation than neck, but FNA should be done after CT scan to rule out vascular tumors.

### Conclusion:

Extracranial Schwannomas which most often present as which often present as asymptomatic solitary masses, are rare tumors. The pre-operative diagnosis may be different and it is often made after the surgery. These Schwannomas should be differentiated from vascular tumors like carotid body tumors, glomus vagale pre-operatively, become treatment plan depends upon the diagnosis.

Complete surgical excision with appropriate approaches is the treatment of head and neck Schwannomas. The possibility of nerve injury should be kept in mind, local recurrence is extremely rare.



Fig 1 showing the mass pushing the lateral pharyngeal wall on the left side



Fig 2 showing swelling on exposure



Fig 3 showing swelling being removed

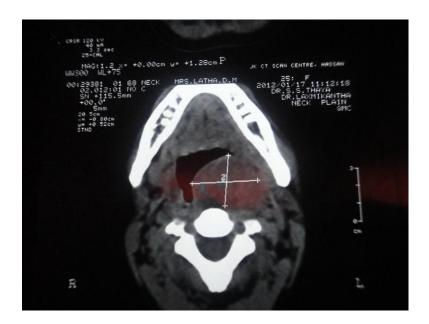


Fig 4 Showing soft tissue mass occupying the parapharyngeal space



Fig 5 Showing the excised specimen

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