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Hypoglossal Nerve Schwannoma: A rare case presentation

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Abstract:
Hypoglossal nerve schwannoma is a very rare slow growing encapsulated tumour. We would like to present a case report of a 51 years old man presented with painless, slow progressively swelling in the right submandibular region with history of giddiness and syncope off and on for more than one year.
Histopathology exam of post-operative specimen shows Schwannoma with degenerative changes
Keywords: *Hypoglossal nerve schwannoma, submandibular swelling.*

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I. Introduction

Schwannoma is a benign slow growing encapsulated tumour that originate from the myelin producing schwann cells or nerve fibre sheet cells at the glial-Schwann cell junction. Schwannoma is diagnosed with Antoni A and Antoni B areas on histopathology exam. Hypoglossal nerve schwannoma develops in the intracranial and extracranial segment or both forming dumbbell shape tumour. Benign schwannomas represent 35% of the head and neck district tumours. 90%of schwannoma are acoustic schwannoma. Only 5% of non-acoustic Schwannoma accounts for hypoglossal schwannoma.

II. Case report

A 51 years old male presented to ENT OPD with painless, slow progressivelyswelling of the right submandibular region without any other head and neck complain for one year associated with the history of giddiness and syncope attack off and on. Clinical examination revealed the presence of an ovoid, non-tender mass in the right submandibular triangle with no neurological deficient.

Routine investigations like CBC, LFT, KFT, RBS, BT, CT, CXR-PAV, ECG were all within normal limit. FNAC shows features of chronic sialadenitis, X-ray soft tissue neck AP and Lateral views were within normal limit (fig:1), Ultrasonography of neck shows right submandibular well defined, encapsulated hypoechoic mass adjacent to posterior border of right submandibular gland with minimal vascularity (fig:2), CECT of neck shows right submandibular cystic swelling containing high attenuating collection with no evidence of enhancement in contrast study with no evidence of periglandular fat stranding (fig:3).

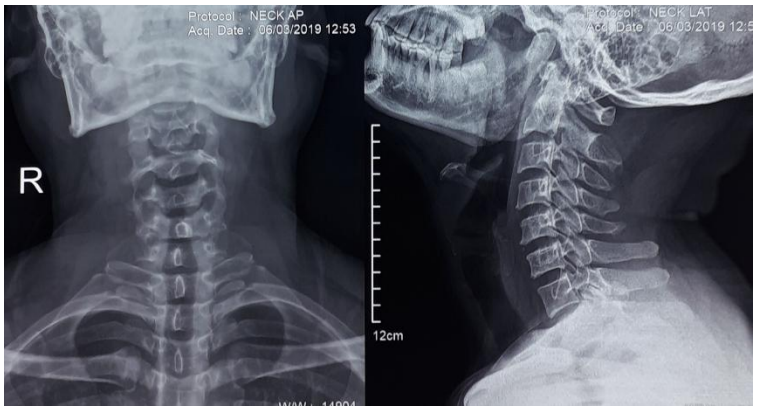


Fig: 1 X-ray soft tissue neck AP and Lateral view

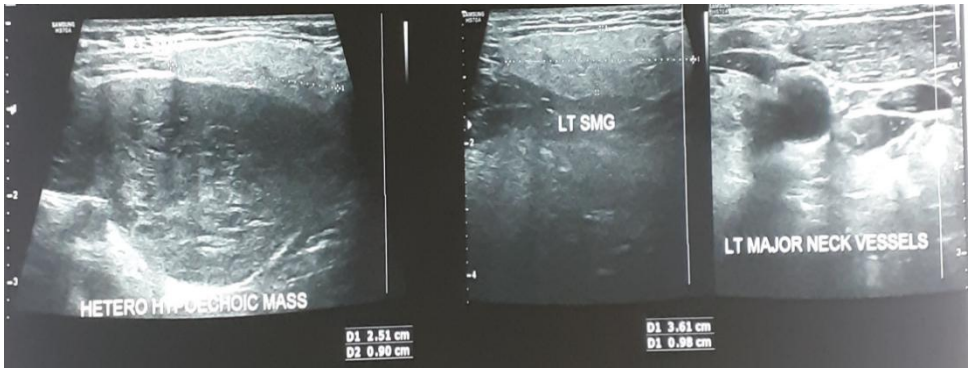


Fig: 2USG of Neck

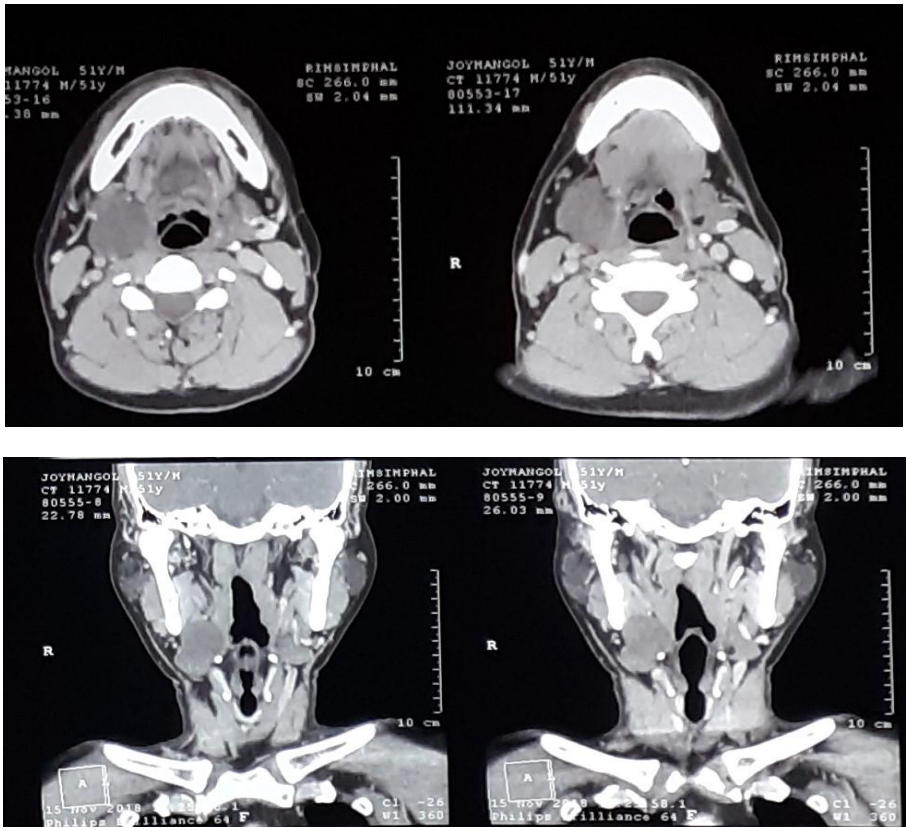
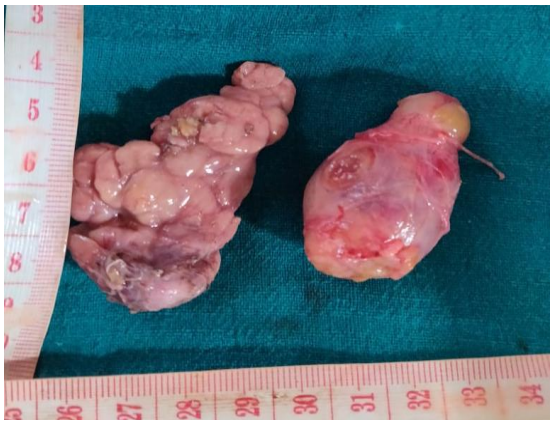


Fig: 3 CECT Neck

Surgery: after obtaining informed written consent, the patient underwent surgical excision of the submandibular gland under general anaesthesia and intra-operatively another voluminous ovoid mass underneath the submandibular gland was found pressing the carotid sheath. Hypoglossal nerve was identified and found embedded in the mass. After obtaining another consent from the patient party the mass 2nd mass was excised and alone with it the hypoglossal nerve has to be sacrificed

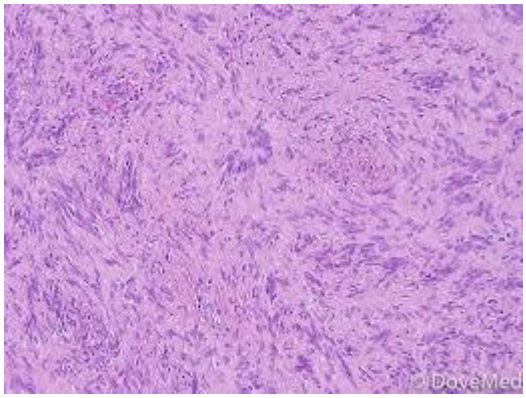
III. Histopathology

Specimen: Excision biopsy, swelling, right submandibular triangle for HPE
Gross: 1. Specimen labelled right submandibular gland measures 4.5x3x2 cm, C/S unremarkable
2. Specimen labelled container 2, consist of a soft tissue mass measuring 4x2.5x2 cm, external surface is unremarkable,C/S – firm grey white and glistening with areas of haemorrhage

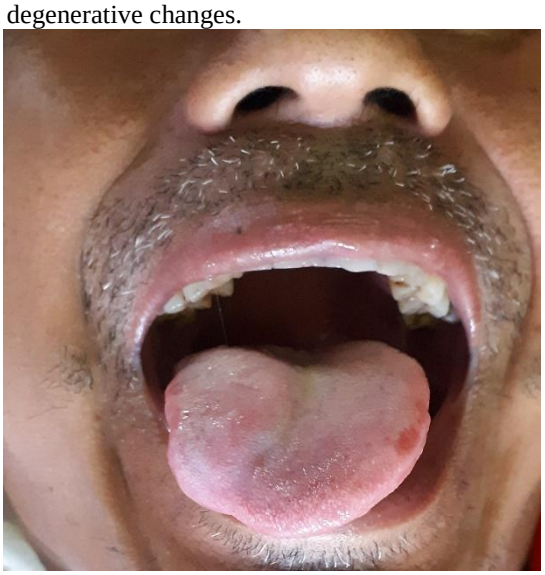


Microscopy: Multiple section studied from the entire cut surface of the specimen labelled right submandibular gland shows well preserved lobular architecture. No evidence of inflammatory cell infiltration or granulomas seen

Multiple study from the entire cut surface of the specimen 2 shows well capsulated tumour displaying hypercellularity Antoni A and hypocellularity Antoni B areas. Numerous verocay bodies are seen. Interspersed in between are many blood vessels with thick hyalinised walls and areas of haemorrhage. No evidence of nuclear atypia is seen. Features suggestive of Schwannoma with degenerative changes.



Impression: Schwannoma with degenerative changes.



Post-operatively patient had deviation of tongue to ipsilateral side and there is no other associated complaints. Patient was discharged with advice for regular follow up every weekly for a month and then monthly.

IV. Discussion

Clinical features: a specific diagnosis of schwannoma is difficult as it is slow-growing discrete lump and is usually asymptomatic. Only when they reached certain dimensions (~2-3cm) may cause pain and paraesthesias. They are able to increase their volume up to 4-5cm and cause compression and dislocation without infiltration of the nerve of origin.

Location of the lesion: Hypoglossal nerve Schwannoma develops in the intracranial and extracranial segment or in both intra and extracranial segment forming a dumbbell shape tumour. As observed from a study of the literature and as confirmed by Lee et al (1), the incidence of isolated extracranial lesions appears to be extremely rare.

Symptomatology: depending on the dimensions of the lesion, the clinical picture may vary greatly. F Rosario at el (2) statedin almost all the cases there are varying degrees of paresis of the Hypoglossal nerve, ipsilateral deviation, hemiatrophy and fasciculation of the tongue. These symptoms may be accompanied by hoarseness, intermittent headache, dysarthria slur speech, swallowing difficulties and throat cramps.

90 % of Schwannoma are Acoustic Schwannoma, of which only 5% on non-acoustic Schwannoma accounts for Hypoglossal Schwannoma. (3,4)

Patients with extracranial tumours may present with a mass in the upper neck or parapharyngeal area, in addition to wasting of the tongue. (3)

Imaging and diagnosis: Pre-operative imaging like CT scan and MRI is useful, it is impossible to make a definitive diagnosis without a histological examination of the excised specimen. (2)

Grading: Nonaka et al classified the lesion involving the hypoglossal canal: Type A, intradural tumour; Type B, transdural and extradural dumbbell shaped tumors; Type C, Extracranial base tumour. (5)

Treatment: Microsurgical resection is the main aim of management. Complete microsurgical resection is often associated with a high rate of morbidity. Subtotal and near-total resection followed by stereotactic radiosurgery or observation now offer an alternative approach. (3)

Despite the rarity of schwannomas, this condition should be considered in differential diagnoses for masses localised in the neck, as in cases where they reach considerable sizes >3 cm in diameter. (6)

V. Conclusion

Though hypoglossal nerve Schwannoma is very rare it should be considered in differential diagnosis for mass localised in the neck.Surgical Resection of the mass is the treatment of choice.Diagnosis is confirm by HPE with Antoni A and Antoni B areas

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Conflicts of interest
There is no conflicts of interest

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