CASE REPORT

Retropharyngeal Schwannoma excised through Smith Robinson approach: a rare cause for dysphagia

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Introduction

Neurofibromatosis (NF) is a neurocutaneous disorder that leads to tumor formation in the central and peripheral nervous systems, affecting the brain, spinal cord, organs, skin, and bones. It is classified into three types: NF1, which constitutes approximately 96% of cases; NF2, occurring in about 3%; and schwannomatosis (SWN), which accounts for less than 1%.[1]. Neurofibromatosis type 2 (NF2) is defined by the formation of multiple benign tumors in the nerve sheath, known as schwannomas, which primarily impact the vestibular nerve [2]. Schwannoma has an extended clinical latency and a late and difficult diagnosis characterize it. Space-occupying lesions in the retropharyngeal region are uncommon. Several types of masses have been identified in this area, including angiomyomas, ganglioneuromas, malignant mesenchymomas, and neuroblastomas, with hamartomas being the most frequently observed. Clinical presentations vary, but common symptoms include difficulty breathing (dyspnea), trouble swallowing (dysphagia), and obstructive sleep apnea. Here, we present a rare case of retropharyngeal neurofibroma that was managed surgically.

Case Report

A 31-year-old female patient with a background history of thalassemia trait, NF type 2- multiple neurofibromatosis, was referred to our neurosurgery unit in February 2024 for evaluation of spastic quadriparesis. The patient presented with progressive weakness over both the upper limbs and lower limbs. But she denied other local pressure effects neither loss of weight nor loss of appetite. The patient also complained of non-progressive painless dysphagia at the throat.

On clinical examination, the patient had increased tone but reduced power over both upper and lower limbs (power of 2/5) but had no objective sensory deficit. There was no obvious swelling or any palpable lymphadenopathy. Her ESR value was within the normal range. The patient underwent an MRI which showed four intra-spinal masses/neurofibromas;

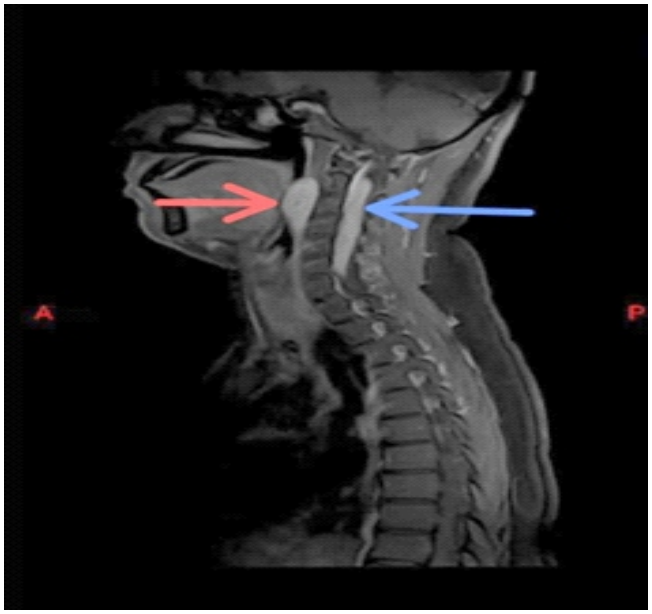

Figure 1: Sagittal view of the pre-op T1 weighted MRI; Blue arrow denotes the intra spinal schwannoma, Red arrow denotes the pre vertebral schwannoma.

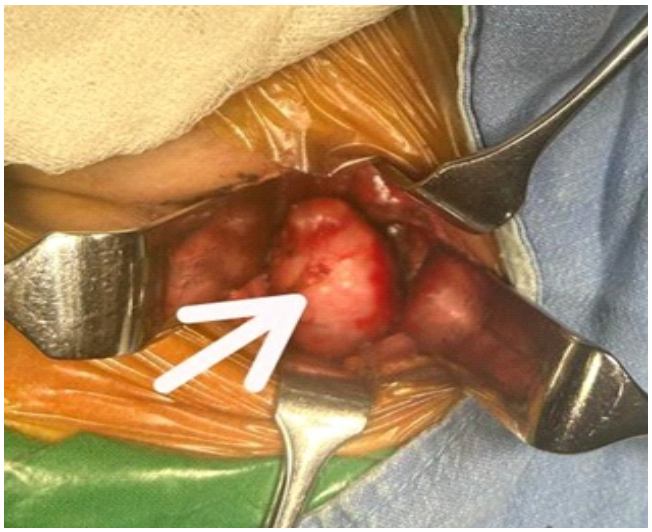

Figure 2: Surgical resection of the retropharyngeal neurofibroma. White arrow depicts the pre-vertebral mass

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the largest was seen within the cervical spinal canal anterior to the cord, measuring 7.3x1.1x1.9cm, compressing the cervical spinal cord. In addition, a similar lesion measuring 4x2x1.6cm was seen at the C2–C4 level shifting the anterior cervical soft tissues to the right of the oropharynx, down to the epiglottis. It compressed and distorted the oropharynx (Figure:1).

Following pre-operative assessment, the patient underwent C2–C6 laminectomy and cervical neurofibroma excision under general anaesthesia. A tumour was identified at the C5 ventral root, and complete macroscopic resection was performed. The patient had an uneventful recovery. The bilateral upper limb and lower limb power were improved to 4/5 post operatively. The histology report revealed Schwannoma CNS WHO grade 1, containing Antoni A and B tissue architecture. The patient was discharged with the plan of elective surgery for the excision of the right-sided retropharyngeal schwannoma after 2 months.

Complete excision of the retropharyngeal schwannoma was performed under general anaesthesia using the Smith-Robinson approach, and a nasogastric tube was inserted for immediate post-op feeding. A right-sided transverse cervical incision was made, and subplatysmal myocutaneous flaps were raised. The blunt and sharp dissection was carried out medial to the sternocleidomastoid muscle. Once the cervical oesophagus was retracted medially, the schwannoma was identified anterior to the pre-vertebral plane (Figure 02).

Complete resection was carried out. The patient resumed oral feeding on postoperative day 2 and was discharged from the hospital the next day following an uneventful recovery. The histology report confirmed a schwannoma. The patient was asymptomatic during the follow-up.

Discussion

The hallmark of NF2 is a mutation in the NF2 gene responsible for the production of a protein called merlin, which acts as a tumour suppressor gene located on chromosome 22. This genetic mutation results in abnormal cell proliferation and loss of apoptosis, leading to an increased risk of developing various benign and malignant tumours, particularly within the CNS [3]. Patients with NF2 often develop spinal schwannomas, which grow from the dorsal roots of spinal nerves, can be intradural or extradural. Extradural schwannomas, comprising 60% to 70% of cases, usually appear as painless masses that compress and displace the spinal cord, nerve roots, and nearby structures such as the vertebral artery.

Tumors of the parapharyngeal space (PPS) are rare, accounting for less than 1% of all head and neck neoplasms. Approximately 50% of PPS schwannomas originate from the vagus nerve, while the cervical sympathetic chain is the next most common source. The clinical presentation of schwannomas varies depending on the anatomical area involved but often includes pressure-related symptoms such as dysphagia and hoarseness of voice. In the present case, patient initially presented with spastic paraplegia and non-progressive dysphagia. Hence, an MRI was carried out, and the lesion was noted in the right parapharyngeal area distorting the oropharynx.

MRI is particularly helpful in identifying the originating nerve, with some studies reporting a perfect success rate. Although closely associated with the nerve of origin, neurological function is maintained in 86% of patients after resection. A biopsy is used to diagnose schwannomas, which typically exhibit both Antoni A and B tissue patterns. Antoni A areas are characterized by densely packed nuclei with central thickening and tapered ends, while Antoni B areas display a more relaxed arrangement of slender, spindle-shaped cells. [4]. Similar findings are seen in our patient's histology report. Hence, the biopsy was not obtained.

Transoral biopsy and removal of schwannomas in the parapharyngeal space can result in complications, including incomplete excision, bleeding, infection, and damage to cranial nerves. The preferred surgical methods primarily involve a transcervical approach, but other options include transparotid, transzygomatic, transmandibular, and a combination of transparotid and transcervical techniques.. The transcervical approach is typically adequate for most tumours in the post-styloid compartment. However, total resection is the gold standard for treating patients with sensory or motor deficits due to extradural intradural spinal tumours.

Surgical excision is the preferred treatment, with recurrence being infrequent if the tumour is completely removed. However, incomplete removal can lead to recurrence, necessitating repeat excision.

In conclusion, retropharyngeal schwannomas can be asymptomatic and cause challenging in diagnosis. The Smith Robinson approach would give adequate exposure. Complete resection will cure the symptoms and minimal chance of recurrence.

References

1.Tamura R. Current understanding of neurofibromatosis type 1, 2, and schwannomatosis. International journal of molecular sciences. 2021;22(11):5850.

2.Plotkin SR, Messiaen L, Legius E, Pancza P, Avery RA, Blakeley JO, et al. Updated diagnostic criteria and nomenclature for neurofibromatosis type 2 and schwannomatosis: An international consensus recommendation. Genetics in Medicine. 2022;24(9):1967-77.

3.Bachir S, Shah S, Shapiro S, Koehler A, Mahammedi A, Samy RN, et al. Neurofibromatosis type 2 (NF2) and the implications for vestibular schwannoma and meningioma pathogenesis. International journal of molecular sciences. 2021;22(2):690.

4.Carroll C, Jagatiya M, Kamel D, Siddiqi J. A parapharyngeal space schwannoma arising from the vagus nerve: A case report. International Journal of Surgery Case Reports. 2017;41:22-5.

Learning Points:

- In rare cases, retropharyngeal schwannoma can present with local obstruction symptoms
- They can pose challenges in both diagnosis and surgical approach.
- Complete surgical resection is the gold standard to preserve or improve neurological function.