

# Primary Extracranial Meningioma As A Part Of Neurofibromatosis-2 In Head And Neck: A Rare Case Report

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## Citation

M Munjal, J Passey, A Sethi, D Singhal, R Bansal. *Primary Extracranial Meningioma As A Part Of Neurofibromatosis-2 In Head And Neck: A Rare Case Report*. The Internet Journal of Otorhinolaryngology. 2004 Volume 3 Number 2.

## Abstract

Extracranial meningiomas, either primary or secondary, are rare. Reported incidences are between 0.9 and 2.0% respectively. The most common form is cutaneous, and, orbital, paranasal, temporal and oral presentations following in descending order. Meningiomas are not usually considered in the differential diagnosis of neck swellings. One such rare clinical case where presentation was a neck mass along with oropharyngeal bulge associated with asymptomatic bilateral acoustic neuromas is presented here along with review of relevant literature. The patient underwent successful excision of cervical mass

## INTRODUCTION

Meningiomas are relatively common neoplasms of the nervous system. They comprise around 18% of all primary intracranial tumors and about 25% of all primary intraspinal tumors<sup>1,2</sup>.

Extra-axial meningiomas are rare in comparison to axial lesions. Reported incidence ranges from 0.9% to 2.0% of all meningiomas<sup>3</sup>. Even more rare, Primary Extracranial Meningiomas (PEM) can be defined as those not associated with an underlying meningioma of the axis. The clinical presentation of PEM can be extremely subtle. Localizing signs or symptoms usually become apparent only when the tumor reaches significant size. Manifestations are related to mass effect at the site and neurological dysfunctions due to intimate involvement of cranial nerves. The rarity of the lesion prompted us to report this case.

## CASE REPORT

A 15-year-old female presented with the complaints of gradually progressive swelling of the neck of 4 years duration along with slurring of speech, dysphagia to solids and liquids and snoring of 2 months duration. Two episodes of excruciating generalized headaches with vomiting and prostration were also reported. The patient also reported decreased hearing from right ear. No history of any hypertensive crisis, vertigo, fever or altered sensorium was reported.

On examination, a firm to hard non-tender swelling measuring 8 by 6 cms. was present in the right posterior cervical region extending posteriorly over the mastoid tip, superiorly over parotid region, anteriorly till the submandibular gland and inferiorly till the level of hyoid bone (FIGURE 1). The swelling was found to have restricted mobility but was not fixed to underlying structures.

## Figure 1

Figure 1: Large neck swelling on right side.



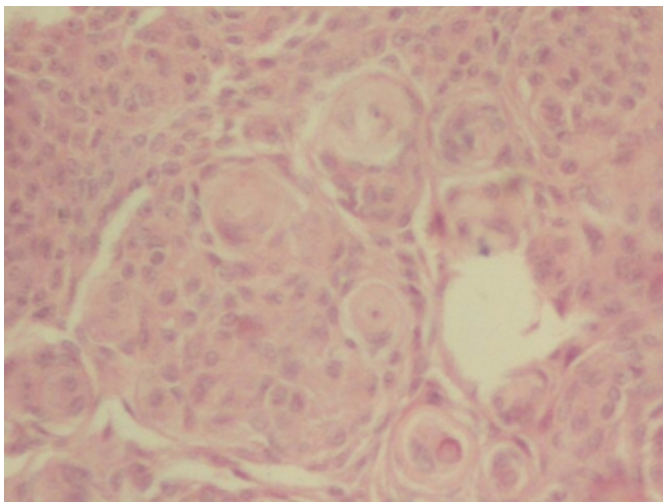
On oral examination, a significant right parapharyngeal bulge pushing the uvula to the left was found. Indirect laryngoscopy revealed restricted mobility of the right vocal cord.

Cranial nerves IX, X and XII on the right side showed features of LMN type palsy. Patient was also found to have Horner's syndrome on examination. There was no other sensory or motor weakness and rest of the systemic examination was normal.

Preliminary investigations performed included a cytopathological evaluation (F.N.A.C.), which showed features of non-calcific psammomatous meningioma (FIGURE 2).

**Figure 2**

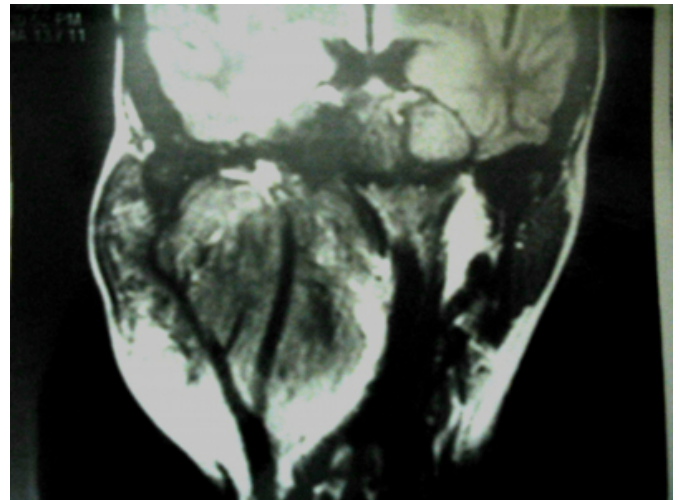
Figure 2: Typical whorled appearance of cells in a psammomatous meningioma. H & E Staining, 400X.



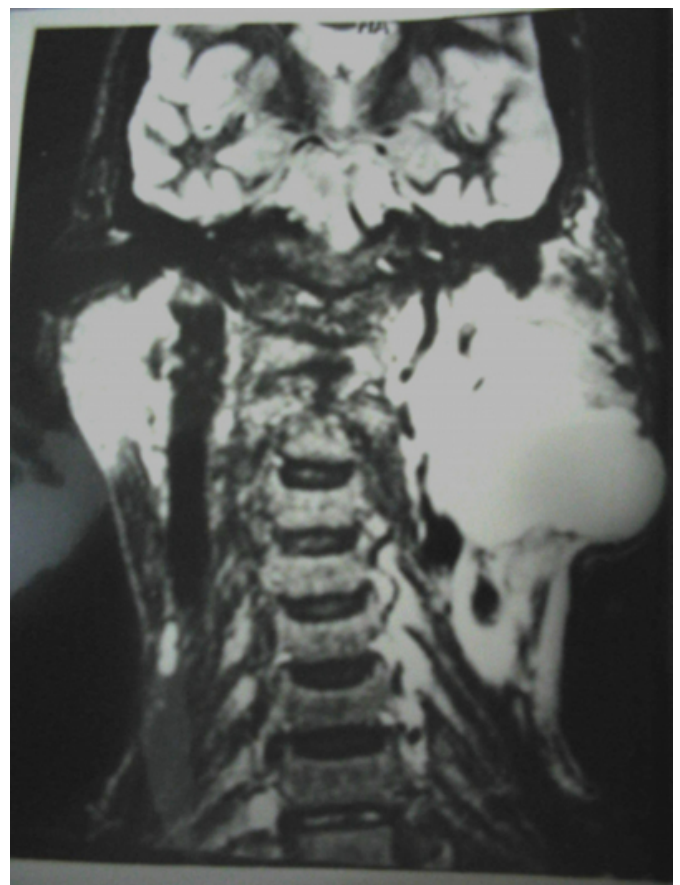
MRI Head and neck showed the presence of a large iso-intense (T1 & T2 gadolinium) lobulated mass extending from the skull base to hyoid bone (FIGURES 3 and 4). The mass caused attenuation of pharyngeal airway, anterior displacement of pterygoid muscles, and, posterior displacement and splaying of sternomastoid. The mass also showed engulfment of internal and external carotid arteries and internal jugular vein, starting bifurcation upwards. MR-Angiography showed narrowing of the lumen of both carotids, but blood flow was normal. Scans of the brain revealed small round ovoid extra axial masses in B/L cerebellopontine angles with extension into internal auditory canals. No contiguous spread between the intracranial and extracranial parts was noted.

**Figure 3**

Figures 3 & 4: The extent of lesion on an MRI-Scan.



**Figure 4**



All other investigations including hematocrit, urine for metanephrines, thyroid tests and chest X-ray were unremarkable.

## TREATMENT

For surgical excision, a transcervical-transmandibular approach was employed to expose the tumor from base skull down. Initially using sharp dissection, the tumor mass was separated from the carotids, but due to the dense adherent nature of the tumor, the internal jugular vein, lower tributaries of the external carotid and the vagus nerve had to be sacrificed.

A triradiate osteotomy at the ascending ramus of the mandible was then performed, and, using blunt finger dissection, tumor was removed from the jugular foramen area, the infratemporal and parapharyngeal regions. The mandible was then repaired using mini plates. Recovery was uneventful except for mild aspiration and also voice change, which is presently compensating well. The patient is currently under neurosurgical observation.

## DISCUSSION

An extracranial meningioma is not likely to be considered in the differential diagnosis of a cervical neck mass<sub>4</sub>. Fewer than hundred cases have been reported in literature till date. The classification system of Hoyer<sub>5</sub> delineates the major etiologies of the tumor and its development:

- A. Extracranial extension of an intracranial meningioma (secondary).
- B. Extracranial extensions of a meningioma arising in a neural foramen (primary).
- C. Ectopic, without any connection either to a foramen of a cranial nerve or to intracranial structures (primary).
- D. Extracranial metastasis of an intracranial meningioma (secondary).

Histologically, four microscopic patterns dominate<sub>2</sub>:

1. Syncytial, characterized by uniform sheets of polygonal cells,
2. Transitional or psammomatous, with a whorled pattern of polygonal or spindle cells with a number of interspersed psammoma bodies,
3. Fibrous, manifesting as an increase in collagenous tissue and a corresponding decrease of cellularity,

and

4. Angioblastic.

With the advent of newer techniques like MRI, the detection rate has increased, origin can be ascertained and associated intracranial extension can also be ruled out. These diagnostic modalities are essential in preoperative surgical planning which stays the mainline treatment of choice. In the age group less than 30 years, it is particularly important to rule out associated neurofibromatosis-2, which further may require genetic counseling and lifestyle changes.

Although seen associated with bilateral acoustic neuromas and intracranial meningiomas, most of the times the surgery involves resection of the extracranial meningioma separately using appropriate base skull approach and observing the intracranial lesions only if there are no symptoms, and later, plan a second stage if need arises. Our patient also had bilateral acoustic neuromas and was managed on the same lines.

## CONCLUSIONS

We would like to conclude that, although rare, an extracranial meningioma should be kept in the differential diagnosis of neck masses, especially if associated with rare conditions like neurofibromatosis-2. We would also like to emphasize that isolated removal of the extracranial meningioma is the standard procedure that is followed in cases with associated intracranial asymptomatic masses like in neurofibromatosis-2.

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