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Case Report

Ear,Nose and Throat Section

Extracalvarial Meningioma in the Parapharyngeal Space: Presentation, Diagnosis and Management

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ABSTRACT

Extracalvarial meningioma in the cervical region presenting as a parapharyngeal mass lesion is seldom encountered in clinical practice and poses great challenge in its diagnosis and surgical management. In this report, we present a case of extracranial meningioma in a middle-aged person who presented with a large, gradually progressing cervical swelling with multiple cranial nerve pareses. The difficulties in diagnosis and surgical management of this unusual neoplasm in the setting of partial encasement and thrombosis of the internal jugular vein have been discussed, along with the computed tomography and magnetic resonance imaging providing details of its extent and character. The report emphasizes the need to consider extracalvarial meningioma as a less common but important differential diagnosis of parapharyngeal space neoplasms.

Keywords: Extracranial, Internal jugular vein, Paraganglioma, Parapharyngeal tumour

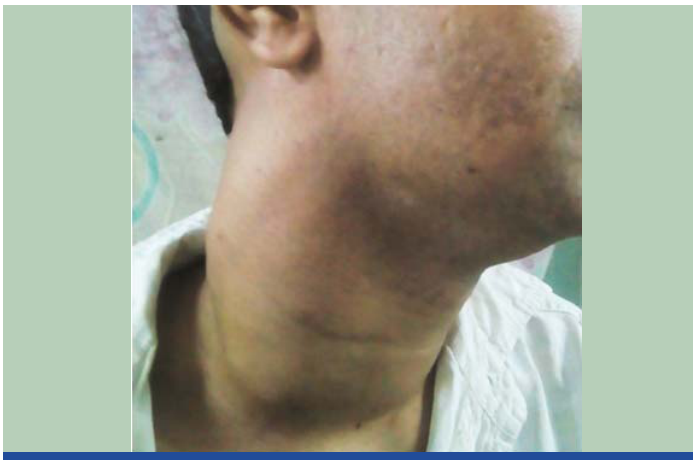
CASE REPORT

A 35-year-old man presented in the otolaryngology outpatients' department with a swelling in the right side of his neck and recent-onset hoarseness of voice. The cervical mass was noticed 18 months back and since then it progressed gradually. It was about 10 cm x 6 cm in dimension, extended from the right infra-auricular region to just above the clavicle, was firm in consistency and appeared bosselated [Table/Fig-1]. The lesion was fixed to the underlying structures; however, the overlying skin was free. Because of the mass effect, the trachea was shifted to the left and the right-sided sternocleidomastoid could not be appreciated. During oral cavity examination, the uvula was seen pushed to the opposite side. The gag reflex was diminished and there was deviation of the tongue to the ipsilateral side on attempted protrusion. Fiber-optic laryngoscopy revealed immobile right vocal cord. Examination of other cranial nerves, including that of the spinal accessory, was unremarkable.

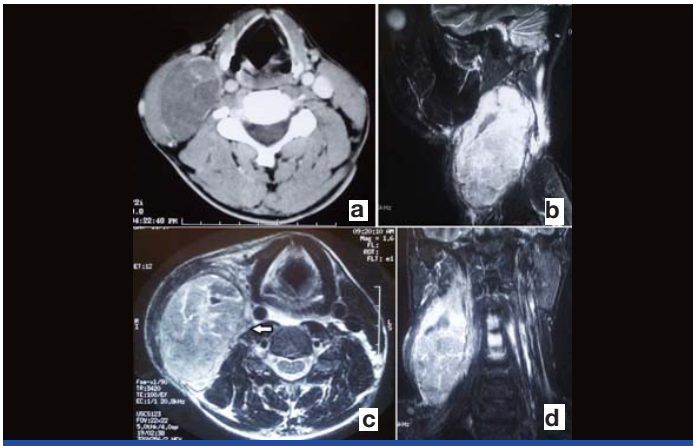
The patient sought medical advice one year back for the enlarging cervical mass. Contrast-enhanced Computed Tomography (CT)-scan done at that time showed a lobulated mass at the right upper neck involving the parapharyngeal space [Table/Fig-2a] and jugular foramen. The Fine Needle Aspiration Cytology (FNAC) report was also made available to us which suggested the lesion to be a paraganglioma. However, the patient refused surgical intervention and did not turn up for the next one year. It was the recent-onset hoarseness and further increase in the size of the neck mass that prompted him to change his mind.

We performed a gadolinium-enhanced Magnetic Resonance Imaging (MRI) of neck which showed a lobulated, multiloculated, heterogeneous soft tissue lesion in the right parapharyngeal space, about 64 mm x 41 mm x 94 mm in size, with areas of necrosis and with an intact capsule [Table/Fig-2b-d]. A concomitant imaging of the brain excluded any intracranial extension. In both the present MRI and earlier CT images, the lesion could be seen abutting the carotid sheath and incorporating the internal jugular vein within its capsule, with obliteration of the venous lumen. A repeat FNAC corroborated the earlier findings, that showed round-to-oval cells in discrete and clustered pattern, with stippled chromatin and fine granular cytoplasm in a haemorrhagic background. A provisional diagnosis of paraganglioma confined to the neck with multiple cranial nerve pareses (IX, X, XII) was made, and the patient was prepared for excision of the tumour. Informed consent in written was obtained from him and prognosis regarding the variable outcome of the affected cranial nerves was duly explained.

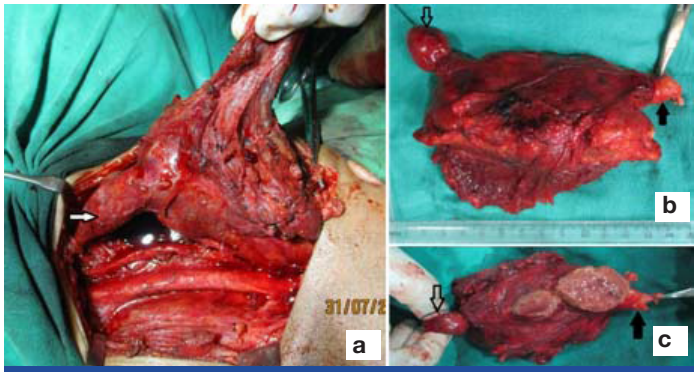
Intra-operatively, the tumour was found densely adhered with a segment of the internal jugular vein which was thrombosed and could



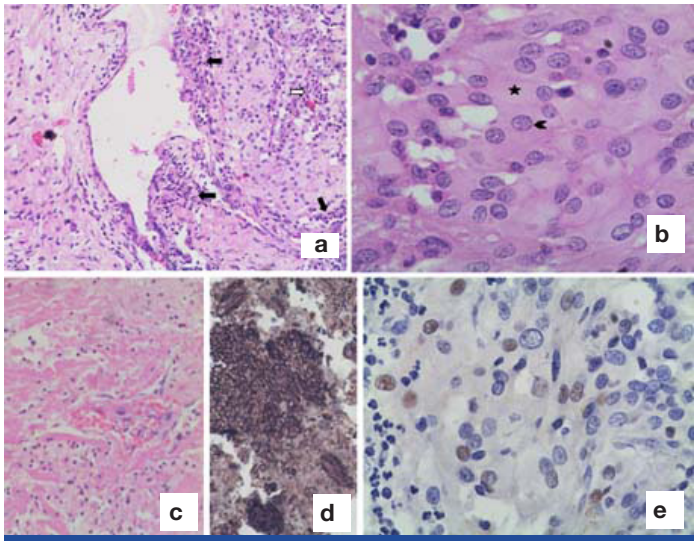
[Table/Fig-1]: The cervical mass in this patient extended from the right infra-auricular region up to the supraclavicular area and was firm and bosselated in appearance.



[Table/Fig-2]: The CT-scan done one year back showed an encapsulated mass in the right parapharyngeal space that took up patches of contrast (a). Subsequent contrast-enhanced MRI showed the mass had increased in size, and was heterogeneous (b). In the axial cuts of both CT and MRI (a,c), the internal jugular vein in the right side could not be visualised as it was incorporated within the tumour capsule with thrombosed lumen (c; arrow)



[Table/Fig-3]: Per-operative photographs showed dense adhesion of the tumour mass with the internal jugular vein in such a way that the vein had been incorporated within the tumour capsule. A considerable section of the internal jugular vein was thrombosed (a; white arrow). The excised specimen showed the tumour with sectioned internal jugular vein (b,c); the transparent arrow indicates the caudal end of the vein, and the black arrow the cranial end.



[Table/Fig-4]: Histopathology revealed palisading histiocytes (white arrow), foamy macrophages and inflammatory cells (black arrows) (a; Haematoxylin-Eosin; x100) along with characteristic round, oval or polygonal tumour cells with vesicular nuclei (arrowhead) and ample eosinophillic cytoplasm (asterisk) (b; Haematoxylin-Eosin; x400), associated with extensive necrosis (c; Haematoxylin-Eosin; x100). Subsequent immunohistochemistry showed positive staining with epithelial membrane antigen (d) and progesterone receptor (e).

not be separated from the mass [Table/Fig-3a]. The tumour was removed in toto along with the thrombosed and engulfed portion of the internal jugular vein. The excised tumour mass measured 14 cm x 9 cm [Table/Fig-3b,c]. Histopathology revealed extensive areas of necrosis with palisading histiocytes, foamy macrophages and inflammatory cells along with some round, oval or polygonal cells with vesicular nuclei and ample eosinophilic cytoplasm [Table/Fig-4a-c]. An infarcted tumour embolus was seen inside internal jugular vein. Subsequent immunohistochemistry showed positive staining with epithelial membrane antigen and progesterone receptor [Table/Fig-4d,e] and was negative for cytokeratin, synaptophysin, chromogranin A, S-100 protein, p63, CD21, CD23 and CD35. Based on the histopathology and immunohistochemistry findings, the final impression of the lesion needed to be revised to extracalvarial meningioma.

The patient recuperated well following surgery, although he developed transient weakness of the right spinal accessory nerve. He was followed up for one year without any recurrence; however, there has been no appreciable improvement of the functions of the IXth, Xth and XIIth cranial nerves till date.

DISCUSSION

Meningiomas are the most common benign intracranial tumours, comprising of 24%-30% of the primary intracranial neoplasms, which originate from the “cap” cells in the arachnoid villi in close proximity to the meninges (parasagittal, falx cerebri, sphenoid ridges) [1,2]. Only 1-2% of the meningiomas are extracranial/extracalvarial [3], mostly involving the orbit, skin and paranasal sinuses [4]. Primary extracalvarial meningiomas in the parapharyngeal space are rarely encountered, with only few published reports in the world literature [5]. A parapharyngeal space meningioma might be secondary to direct extension of intracranial meningioma through the skull base foramina, can arise de novo from the pluripotent mesenchymal cells, or can arise from the meningocytic cell-rests of the cranial nerve-sheaths [6]. Engulfment of the great vessels of neck is also unusual for extracranial meningiomas [6].

As we have observed in our patient, the hallmark of the presenting features of extracranial cervical meningioma is a gradually progressive, painless swelling which might be associated with multiple cranial nerve pareses (especially IX-XII) [5]. The pattern of cranial nerve involvement might indicate whether the tumour involves the region at or near the jugular foramen. Although in our patient there was no radiological or operative evidence of intracranial extension, it might so happen that the tumour had originated near the jugular foramen and subsequently enlarged only in the caudal direction, thereby explaining both multiple cranial nerve pareses due to pressure effect and also the absence of intracranial component.

Several authors are of the opinion that FNAC may not be able to diagnose an extracalvarial meningioma. This experience was shared by us too when twice the cervical lesion was erroneously reported to be paraganglioma on FNAC, which could be partly because paragangliomas along with schwannomas form the bulk of the neurogenic lesions occupying the parapharyngeal space. CT and MRI form the mainstay of radiological evaluation, which provide a precise idea regarding the extent of an extracalvarial cervical meningioma. Intracranial component of this tumour when detected is either in continuity with the primary lesion through the jugular foramen, or cause destruction of the base skull [7]. In the present patient however there was no radiological evidence of intracranial involvement. The multiple cranial nerve pareses could be explained by the fact that the tumour encroached the skull base and partly by the mass effect it caused due to its enormous size. One of the unique features here was the involvement of internal jugular vein, a considerable length of which was found densely adhered with the tumour mass and needed to be sacrificed. Per-operatively the segment of the internal jugular vein from jugular bulb to lower neck was found to be totally thrombosed without any blood flow, and was difficult to identify initially. There have been two instances in record where extracalvarial meningioma in the neck encased the carotid arteries [8], but complete engulfment of the internal jugular vein is unusual and truly unprecedented.

CONCLUSION

Extracalvarial meningioma in the neck constitutes a rare form of parapharyngeal tumour. It poses a diagnostic challenge for the otolaryngologists because cytology reports are often inconclusive and misleading. Pre-operative CT-scan and MRI evaluate the tumour’s extent and exclude intracranial involvement. Histopathology along with immunohistochemistry confirms the diagnosis; however, a high index of suspicion from clinicians and pathologists and the knowledge of its existence in the given anatomical subsite are essential to build up a proper management algorithm for the neoplasm. The tumour in the neck might produce remarkable features like the enormous size at presentation and adhesion with great vessels that might result in difficulty in surgical removal, along with multiple cranial nerve pareses and intracranial extension. The present report illustrates the clinical, imaging and histological details of an extracalvarial cervical meningioma, and suggests it to be one of the important differential diagnoses of parapharyngeal space neoplasms.

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