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Extracranial presentation of a common intracranial tumor: A case report

[1]Dr. Abin Thomas, Junior Resident, Department of Radiodiagnosis, Pushpagiri Institute of Medical Science and Research Centre, Thiruvalla.

[2]Dr. Geena Benjamin, Professor, Department of Radiodiagnosis, Pushpagiri Institute of Medical Science and Research Centre, Thiruvalla.

[3]Dr. Mangalanandan S, Additional Professor, Department of Radiodiagnosis, Pushpagiri Institute of Medical Science and Research Centre, Thiruvalla.

[4]Dr. Mathew George, Associate Professor, Department of Radiodiagnosis, Pushpagiri Institute of Medical Science and Research Centre, Thiruvalla.

[5]Dr. Safvana P P, Junior Resident, Department of Radiodiagnosis, Pushpagiri Institute of Medical Science and Research Centre, Thiruvalla

Corresponding Author: Dr. Abin Thomas, Junior Resident, Department of Radiodiagnosis, Pushpagiri Institute of Medical Science and Research Centre, Thiruvalla.

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Abstract

Meningiomas are mostly benign slow-growing tumors with certain histological subtypes having higher proliferative potential. Extracranial meningiomas constitute about 2% of all meningiomas. In this case, the patient is a 26-year-old female who presented with swelling over the right side of the neck for the past 4 years. The swelling was diffuse, and firm with a size of 7 x 7 cm located over right side of neck behind the angle of mandible, anterior end being 10 cm away from symphysis mention and posterior end extending beyond mastoid process. MRI showed soft tissue mass distorting right carotid and posterior cervical spaces. CT angiography revealed a well circumscribed lobulated heterogeneous soft tissue density mass with multiple foci of calcification. The patient underwent surgery. There was a firm to hard lobulated tumor located in the carotid triangle. Bulk of the tumor was located behind the ICA and posterolateral to the carotid vessels, deep to the right sternocleidomastoid. Cranial nerves 9-12 were embedded in the tumor and densely adhered. Histopathologic examination of the surgically removed

specimen confirmed the diagnosis of Grade I Meningioma. Immunohistochemistry evaluation showed positivity to EMA, S100, VIMENTIN and P63. Even though it is rare, extra cranial meningiomas should be considered as a differential diagnosis while evaluating neck swellings.

Keywords: Meningioma, Extracranial meningioma, posterior cervical space, Immunohistochemistry

Introduction

Meningiomas are mostly benign tumours that develop from arachnoid cap cells, which are non-neuroepithelial progenitor cells. Most meningiomas are slow-growing, benign tumours, but certain histological subtypes have a higher proliferative potential.[1] Extracranial meningiomas are a rare type of head and neck tumours. They constitute only about 2% of all meningiomas.[2] Data from the Central Brain Tumour Registry of the United States (CBTRUS) show that females have more than two-fold higher incidence of meningioma than males.[3] Extracranial meningiomas are most commonly caused by extension from an intracranial primary tumour, and meningocytic cells found with arachnoid clusters or modified mesenchymal cells are less common. Usually, primary extracranial meningiomas are found in the skin, scalp, middle ear, and nasal cavity.[4] Magnetic resonance imaging (MRI) is the preferred modality for assessing meningiomas, but computed tomography (CT) can be used in the absence of MRI or in conjunction with MRI to better depict dystrophic calcifications and bone changes. In recent years, radiomics applications have demonstrated the potential to provide additional information from medical images of meningioma patients.[5] This case report describes a rare case of extra cranial meningioma presented as swelling over the right side of the neck.

Case Report

A 26-year-old female presented with a painless swelling over the right side of the neck for 4 years and difficulty in swallowing for 5 months. Gradually the swelling increased in size and she developed pain over the swelling. On examination, the swelling was diffuse, firm with a size of 7 x 7 cm located over right side of neck below behind the angle of mandible, anterior end being 10 cm away from symphysis menti and posterior end extending beyond mastoid process. Transmitted carotid pulsation was felt over the swelling.

Magnetic Resonance Imaging (MRI) study of head and neck (Fig 1-3) showed moderate to avidly enhancing T1W isointense and T2W heterogeneous mildly hyperintense signal intensity soft tissue mass occupying right carotid and posterior cervical spaces and protruding into the adjacent spaces with the encased carotid vessels pushed anteriorly and the compressed internal jugular vein being displaced posterolateral, and away from the carotids. There is encasement of the carotid bifurcation as well as proximal aspects of internal and external carotid arteries. There is no “salt and pepper” sign on T1W sequence.

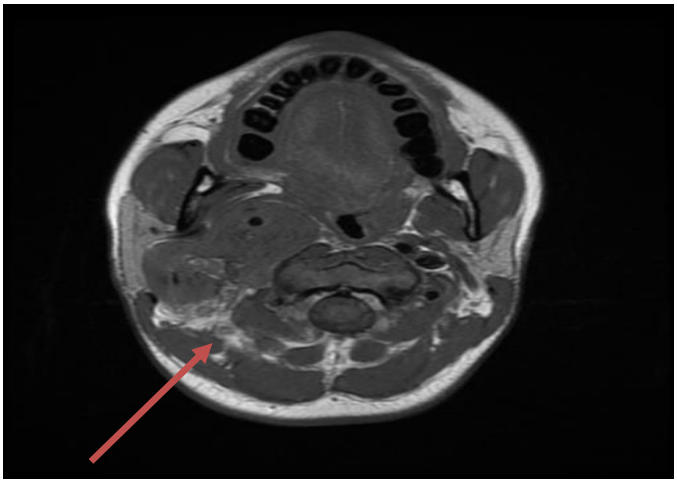

Fig. 1: T1W image showing a soft tissue lesion (arrow) distorting right carotid and posterior cervical spaces

which is isointense to surrounding muscle. There is no "salt and pepper" sign.

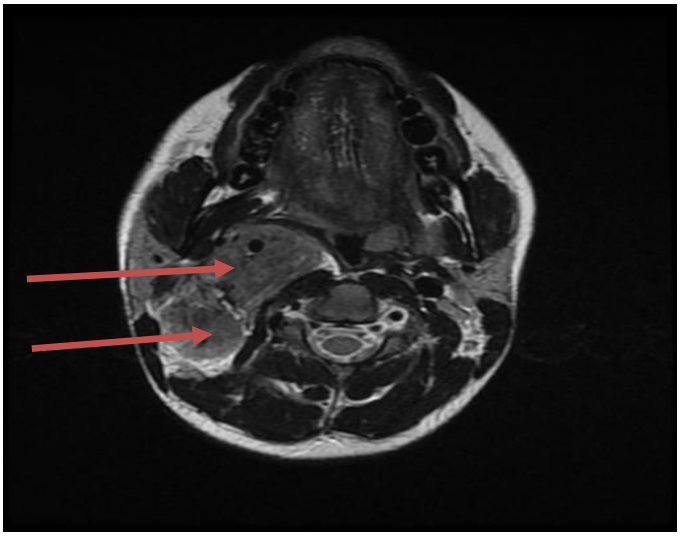

Fig. 2: T2W image showing heterogeneous mildly hyperintense signal intensity lesion encasing the right carotid vessels (arrows), displacing the same anteriorly and the compressed IJV pushed poster laterally.

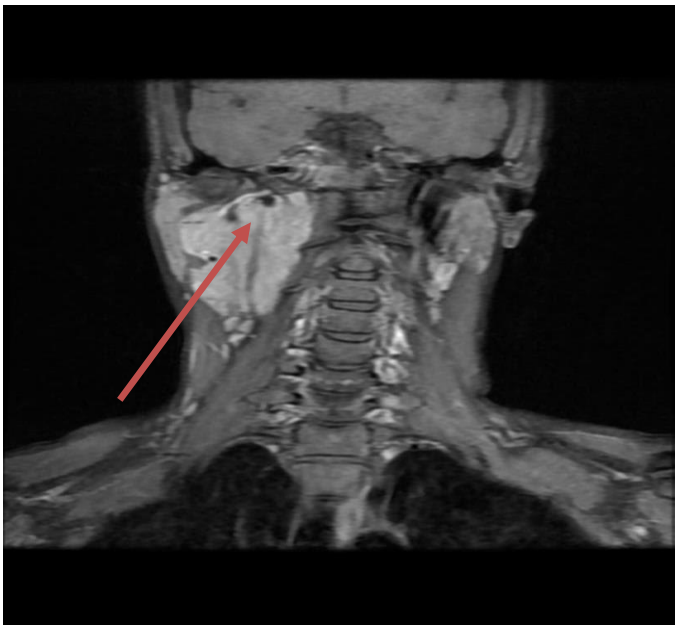

Fig. 3

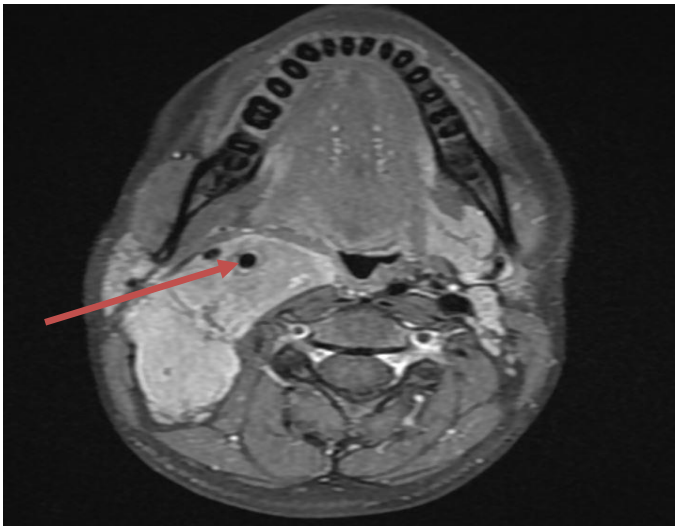

Fig. 4

Figure 3 and 4 - Post contrast T1W sequence demonstrates the lesion showing moderate to avid near homogeneous enhancement. Note the encasement of ICA and ECA (arrows) with no appreciable splaying.

Contrast enhanced CT of neck demonstrated a well circumscribed lobulated heterogeneous soft tissue density mass with multiple foci of calcification, occupying right carotid and posterior cervical spaces protruding into the parapharyngeal, parotid and paravertebral spaces. The swelling extended from jugular fossa region cranially till C4 vertebral level caudally (immediately distal to carotid bifurcation). The lesion was displacing parapharyngeal fat space anteriorly, parotid space laterally, right pharyngeal wall anteromedially (no significant airway luminal narrowing). The lesion is encasing right cervical ICA just distal to its origin till carotid foramen, displacing it anteriorly. No splaying of the carotid bifurcation is seen. The compressed IJV is displaced posterolaterally. The lesion was iso to slightly hyperdense in non-contrast CT with moderate post contrast enhancement in the arterial phase and progressive enhancement in the venous and delayed phases. CT also revealed prominent hyperostosis involving squamous occipital bone with

marked luminal narrowing in the region of right jugular fossa. The right sigmoid sinus, jugular bulb and IJV till the level of carotid bifurcation are not opacified. There are multiple foci of punctate, linear and amorphous calcifications noted within the cranial aspect of the lesion in the retro-styloid portion. No hemorrhagic or cystic areas seen within. Based on the clinical and investigation findings, the diagnosis of extra-cranial meningioma was made.

The differential diagnosis to be considered in this case is carotid body tumor. However, there is no splaying of the ICA and ECA and the lesion is not completely filling up the crotch. On contrast enhanced sequences, the lesion is showing progressive enhancement with no obvious intense enhancement seen in the arterial phase sequence.

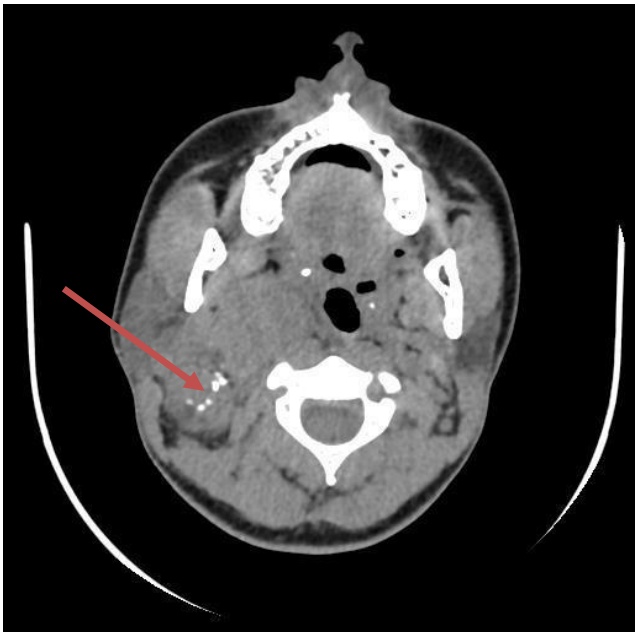

Fig. 5: Non enhanced CT image revealed a heterogeneous iso to slightly hyperdense soft tissue density mass with multiple foci of calcification(arrow),

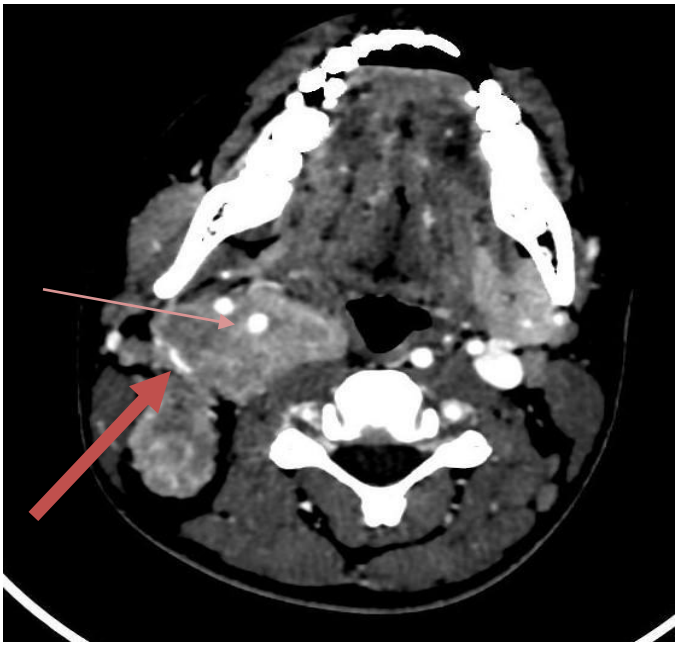

Fig. 6: Contrast CT image shows the lesion exhibiting moderate post contrast enhancement with encasement of the carotid bifurcation (thin arrow) and displacement of the carotid arteries anteriorly as well as compressed IJV (thick arrow) posterolaterally.

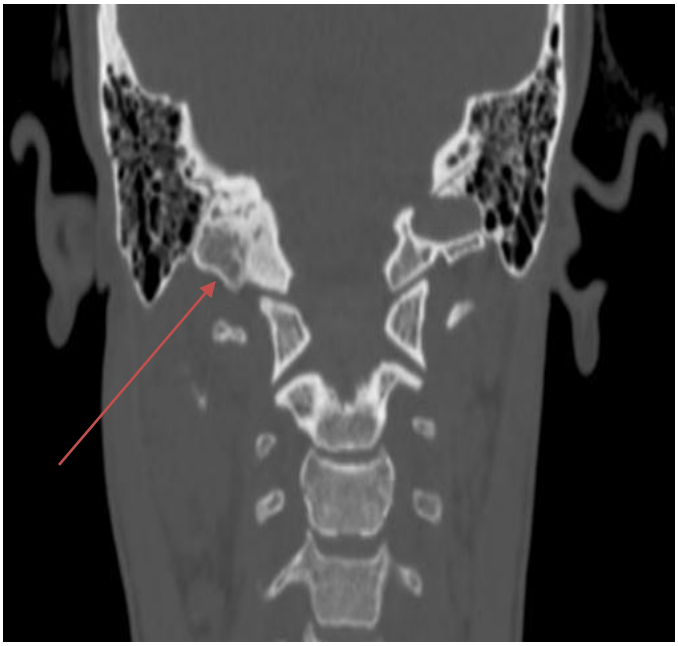

Fig. 7

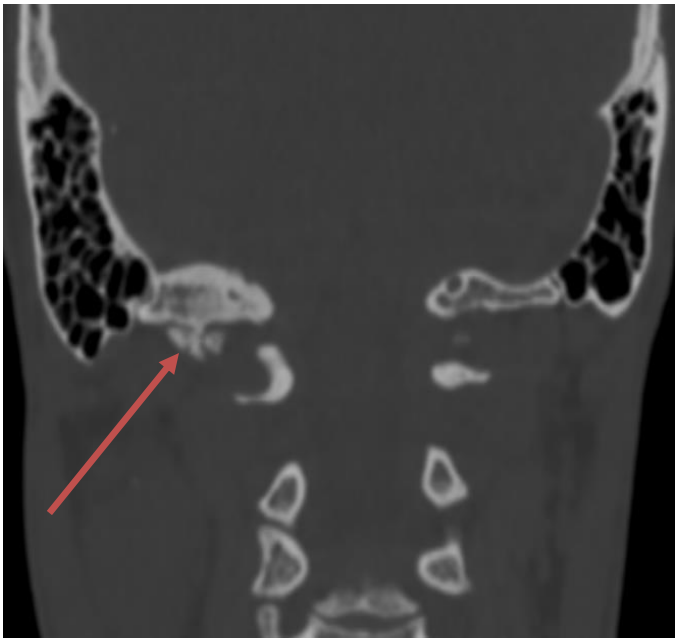

Fig. 8

Figure 7 and 8 - CT (bone window) revealed prominent hyperostosis (arrows) involving the adjacent squamous occipital bone on right side when compared to the left side.

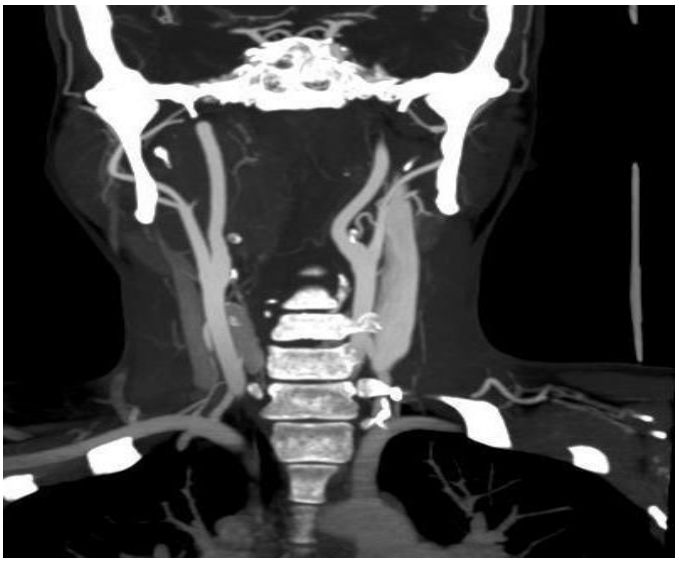

Fig 9: MIP (maximum intensity projection) image– showing the lack of splaying of right ICA and ECA.

Surgical findings

The patient underwent surgery and the following findings were noted. There was a firm to hard lobulated tumour located in the carotid triangle. Bulk of the tumor was located behind the ICA and lateral to the carotid vessels, behind the sternocleidomastoid. Cranial nerves 9-12 were embedded in the tumour and densely adherent. Cranial nerves were hypertrophied with proximal bulbar dilatation, ending in the tumour. ICA had posterolateral encasement. ECA was relatively spared by the tumour.

Histopathologic examination of the surgically removed specimen confirmed the diagnosis of Grade I Meningioma. IHC evaluation showed positivity to EMA, S100, VIMENTIN and P63.

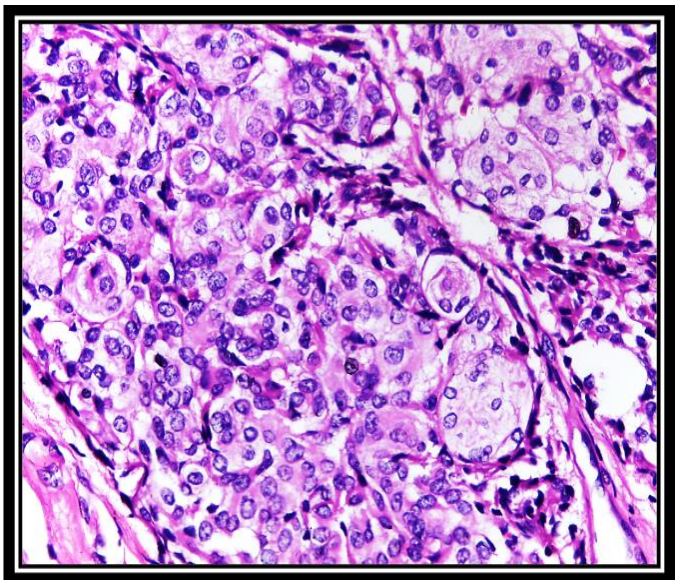

Fig. 10: Cells arranged as nests with few thin-walled blood vessels seen with the fibrous stroma. Individual cells have moderate amount of eosinophilic cytoplasm.

Discussion

Meningiomas make about 13% to 26% of all CNS tumors which are mostly benign, slow-growing tumors originating from non-neuroepithelial progenitor cells, the cells of the arachnoid cap. They often manifest between the fourth and the sixth decade of life, are uncommon in children, and are more prevalent in women. These can exist as intracranial or extracranial brain tumors. Despite their similarities, these two forms of tumours have very

different characteristics and presentations. Extracranial meningioma could be classified into two forms, which are primary and secondary tumors. Primary tumors are completely extracranial, and not associated with leptomeninges or intracranial mass, while secondary extradural meningiomas refer to intradural meningiomas with extradural extension and/or metastasis. [7] Extracranial tumours are very rare, comprising only 2% of all meningiomas and found most often in male patients. Due to their unusual symptoms and lack of prevalence, these tumours are often misdiagnosed, fortunately, 80% of extracranial tumours are benign. [7]

Meningiomas in neck spaces present with vague symptoms. The tumours are often not detected for long periods of time as localized signs do not exist until the tumor has reached a significant size. . When symptoms do present, they can be quite varied based on the site involved. Neurological dysfunction and cranial nerve deficits are the most common signs, and sinusitis, proptosis, epistaxis can also occur depending on the site of the lesion. [8]

Surgical excision of the mass should be performed if possible and external beam radiation therapy post operatively has been shown to be effective in targeting the microscopic remainder of the disease. Radiation therapy can also be used as part of a palliative approach in elderly patients or if the mass is inoperable. [7]

The differential diagnosis to be considered in this case is carotid body tumour. However, there is no splaying of the ICA and ECA and the lesion is not completely filling up the crotch. On contrast enhanced sequences, the lesion is showing progressive enhancement with no obvious intense enhancement seen in the arterial phase sequence. There is no “salt and pepper” sign on T1W sequence.

In our patient the extra cranial meningioma presented as a neck swelling. A case reported by Deshmukh SD et al described a primary extra cranial meningioma in the sub-mandibular region.[10] Mendoza YM et al reported a case of extra cranial meningioma presented as infratemporal fossa mass in a patient presenting with a history of traumatic brain injury.[11] In our case the symptoms included swelling for 4 years with recently developed difficulty in swallowing and pain over the swelling. A study among 146 cases of extra cranial meningioma by Rushing EJ et al observed that the symptoms reported were generally non-specific and varied according to the anatomic site of involvement.[11]

In our case, MRI showed a soft tissue mass occupying right carotid and posterior cervical spaces and protruding into the adjacent spaces. CT angiography revealed a well circumscribed lobulated heterogeneous soft tissue density mass with multiple foci of calcification along with prominent skull base hyperostosis. A similar case of posterior pharyngeal space meningioma reported by Taori K et al showed that a dominant left parapharyngeal is dense mass lesion with moderate heterogeneous contrast enhancement and central amorphous and dense calcifications on CT scan. There were two to three continuous linear vertical hypodensities with surrounding calcifications, giving the lesion a "tram track appearance," possibly indicating cranial nerve encasement. On MRI evaluation, a heterogeneously enhancing altered signal intensity mass lesion in the neck was discovered, along with multiple signal void foci, indicating the presence of calcifications.[12]

In our study, the swelling was surgically removed and histopathologic examination of the surgically removed specimen confirmed the diagnosis of psammoma Tous variant of Grade I Meningioma. IHC evaluation showed

positivity to EMA, S100, VIMENTIN and P63. The study by Rushing EJ et al reported that, among the 146 cases in their study, the majority of tumours (77.4%) were meningothelial, followed by atypical (7.5%), psammoma Tous (4.1%), and anaplastic (2.7%) on histological examination. The tumor cells were immunohistochemically labelled for EMA (76%), S-100 protein (19%), CK 7 (22%), and while ki-67 was labelled in 27%, only 3% of cells were positive.

Conclusion

This case gives an insight that even though it is rare, extra cranial meningiomas can present as neck swelling, and it should be considered as a differential diagnosis while evaluating neck swellings. Prominent hyperostosis involving the adjacent bone should be considered as a lead to the diagnosis of meningioma as in our case. This will help to avoid delay in diagnosis.

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