

Case report

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Left Carotid Sheath Meningioma

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Abstract

Although meningiomas are common tumors of the central nervous system, primary extracranial meningiomas are extremely rare. These tumors constitute approximately 1% of all meningiomas. This study has reported a very rare case of meningioma on the left carotid sheath that presented as a left neck mass. We performed a core biopsy of the mass. Subsequently, the tumor was excised and histopathologic examination confirmed the diagnosis of primary extra cranial meningioma.

Keywords: Meningioma, Carotid sheath, Extracranial, Neck

Introduction

Meningiomas are common tumors of the central nervous system that originate from the meningeal coverings of the spinal cord and brain. They account for approximately 13%-26% of all

primary brain tumors.^{1,2} Primary extracranial meningioma, however, is a rare condition that occurs in less than 1% of all meningiomas.³ There are few reported cases in the literature with limited information on carotid sheath meningioma. We present a

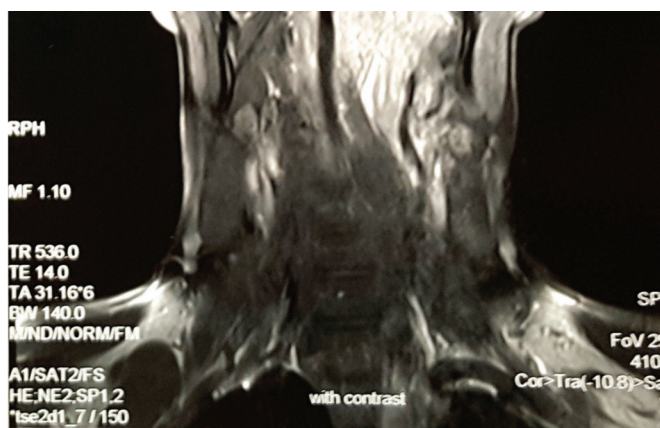


Figure 1. Magnetic resonance image (MRI) demonstrating an intense enhancing lesion in the left carotid sheath with pressure effect on adjacent structures.

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rare case of extracranial meningioma that occurred in the left carotid sheath. Magnetic resonance imaging (MRI) revealed an intense enhancing lesion in the left carotid sheath with the impression of a schwannoma or neurofibroma.

Case history

A 26-year-old man presented with hoarseness and difficulty in swallowing, and fullness at the mandibular angle from 5 years ago that gradually worsened. Physical examination revealed a firm, non-tender, fixed neck mass. Computed tomography (CT) scan of the neck demonstrated a soft tissue mass with foci of calcification that measured approximately 60×40 mm in the left carotid sheath with extension from the carotid bifurcation to the skull base with bony erosion of the apex of the petrous bone, and extension into the left parapharyngeal space. MRI revealed an intense enhancing lesion in the left carotid sheath from the level of the bifurcation of the carotid that extended into the parapharyngeal space, and associated with a pressure effect on the right aspect of the nasopharynx, with an impression of a schwannoma or neurofibroma (Figure 1).

Subsequently, we performed a core biopsy of the mass. The results showed neoplastic

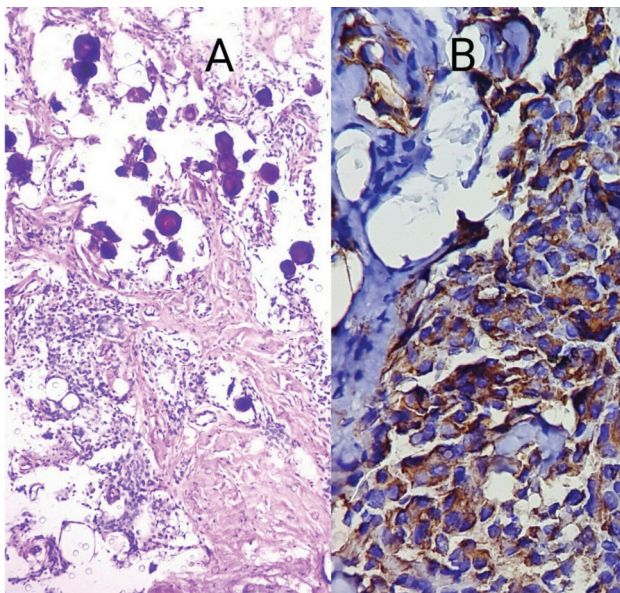


Figure 2. Histologic features of the core biopsy. A) The tumor is composed of epithelioid cells that formed syncytial and sheet patterns with abundant psammoma bodies. (H&E, 10×); B) Tumor cells reactive for epithelial membrane antigen (EMA) (IHC, 40×).

proliferation of epithelioid cells that formed a syncytial and sheet pattern with abundant psammoma bodies (Figure 2A). Immunohistochemistry study showed that the tumor cells were positive for epithelial membrane antigen (EMA; Figure 2B) and negative for CK and TTF1.

The patient underwent transcervical excision of the tumor and four cervical lymph nodes. Histology of the mass showed fibrovascular and striated muscle fibers with small foci of tumor cells that grew in a syncytial pattern. The tumor cells displayed an oval shaped with psammoma bodies. Mitotic activity was less than 4/10 High power field (HPF) (Figure 3). The lymph nodes were remarkable for sinus histiocytosis.

Discussion

According to the World Health Organization, meningioma is a tumor that arises from arachnoidal cells and, in the majority of cases, its behavior is benign. This tumor accounts for 24%-30% of all intracranial tumors,^{4,5} and is more common in females than males with a ratio of 2:1 and peak incidence at the age of 45 years.⁶ Primary extracranial meningioma, however, is a rare condition that occurs in approximately less than 1% of all meningiomas.³ Individuals who present

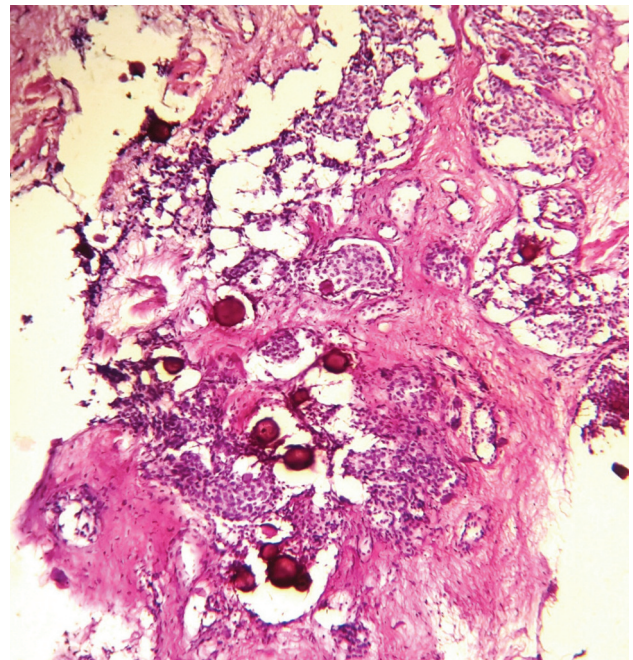


Figure 3. Histopathology of the excised mass that demonstrated meningeothelial cells with multiple psammoma bodies (H&E, 10×).

with extradural lesions need to have a metastatic process excluded, including hematopoietic disease (i.e., lymphoma). In this type of patient, with a negative metastatic evaluation and from a younger age group, a wider differential diagnosis must be considered. Additional possibilities include schwannoma, neurofibromas, infectious processes, and meningioma.⁷ The general histologic features and immunohistochemical findings can usually distinguish these tumors.⁸⁻¹⁰ Schwannoma demonstrate different degrees of cellularity, with areas of myxoid change, perivascular hyalinization, wavy nuclei, and strong, diffuse S-100 protein immune-reactivity. Metastatic carcinomas or carcinomas in general tend to show more pleomorphism, a much higher mitotic rate, and immune-reactivity to a variety of keratins. Whereas psammoma bodies can be seen in papillary carcinomas (thyroid, lung, ovary), the growth pattern of meningioma tends not to be papillary in these extracranial sites. Neuroblastomas usually maintain a lobular growth pattern of small-to-intermediate cells with scant cytoplasm, a fibrillary background, rosette and/or pseudorosette formations, and characteristic immunohistochemical features (chromogranin, synaptophysin, neuron specific enolase, CD56 tumor cell staining, and S100 protein sustentacular staining) that are easy to distinguish from meningiomas. There are two forms of meningiomas, the more common intracranial and an extracranial form. Up to 20% of intracranial meningiomas may have an extracranial component.¹¹ They are found most often in male patients. Due to their unusual symptoms and lack of prevalence, these tumors are often misdiagnosed. Extracranial meningiomas are categorized as primary or secondary tumors. Primary tumors are isolated extracranially and not associated with an intracranial mass. Secondary tumors, however, arise as extensions of intracranial masses.¹² The most frequent extracranial sites are the scalp skin, ears, temporal bone, and sinonasal tract.^{13,14}

The etiopathogenesis of extracranial meningiomas involves the migration of arachnoid

cells derived from the neural crest. However, additional mechanisms are proposed. For example, extracranial meningiomas may potentially originate from i) arachnoid cells of nerve sheaths that emerge from the skull foramina; ii) pacchionian bodies displaced or entrapped in an extracranial location during embryological development; iii) arachnoid islets displaced due to trauma or cerebral hypertension; and iv) undifferentiated mesenchymal cells.¹³

The immunohistochemistry profile of an extracranial meningioma is indistinguishable from an intracranial lesion. In a study of 146 cases, all tumors that expressed vimentin (100%), epithelial membrane antigen (76.3%), cytokeratin (24%), and S100 (19.2%) were identified.¹⁴ If complete resection is possible, there may be no difference in patient prognosis.¹⁵

Conclusion

Extracranial meningiomas are rare but should be included in the differential diagnosis of extradural masses. We suggest that core needle biopsy maybe a suitable technique to diagnose extracranial meningiomas. Diagnosis may be suggested on CT and MRI, but the core needle biopsy and pathologic examination are essential for diagnosis confirmation.

Conflict of interest

No conflict of interest is declared.

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