



Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.casereports.com

A rare case of extracranial meningioma in parapharyngeal space presented as a neck mass



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ARTICLE INFO

Article history:

Received 3 January 2015

Received in revised form 19 March 2015

Accepted 3 April 2015

Available online 13 April 2015

Keywords:

Meningioma
Parapharyngeal space
Neck mass

ABSTRACT

BACKGROUND: Meningiomas are the most common intracranial tumor, but rarely, they can develop extracranially, usually in the neck. There are very few cases of parapharyngeal meningioma reported in literature and little is known about their biological behavior and operative management. We present a patient with a primary parapharyngeal meningioma that presented as an anterior neck mass.

CASE PRESENTATION: The patient is a 55-year-old female who presented with neck mass. A CT scan and MRI revealed a large, well defined, mildly enhancing soft tissue mass located in the right carotid sheath extended from the level of the thyroid gland into the skull base jugular foramen superiorly. Cervical exploration with partial excision of the mass was performed. Histological examination revealed meningiothelial cells with intranuclear inclusions, arranged in a syncytial pattern. Multiple psammoma bodies these findings are consistent with the diagnosis of meningioma.

CONCLUSION: Extracranial meningiomas are quite rare. The diagnosis of these types of tumors is challenging due to the non specific nature of the symptoms. The anatomic complexity of the region of parapharyngeal space also makes their detection difficult. Imaging modalities can aid in the diagnosis, but pathological examinations are essential in confirming a definite diagnosis.

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1. Introduction

Meningiomas are the most common non- glial tumors of the brain and spine which constitute about 15% of all intracranial and 25% of all spinal tumors [1]. There are two forms of meningiomas, a more common intracranial and the rare extracranial tumor. Women make up the greater percentage of patients with intracranial meningiomas, whereas male patients are more likely to have extracranial meningiomas [2]. These two forms of meningiomas are histologically quite similar, and distinguishing them based on immunohistochemistry is not possible.

Meningiomas originate from pia-arachnoid cells, particularly those of the arachnoid villi, and are classically attached to the dura. Meningiomas exhibit diverse clinical manifestations and histological features such as psammoma bodies, abundant cytoplasm and vesiculated nuclei [3]. Meningiomas have been classified by the

WHO into three different malignancy grades: benign (grade I), atypical (grade II) and anaplastic (grade III). Regardless of the grade the recommended treatment is complete surgical excision if possible, followed by oncological support if necessary.

Due to the rare nature of extracranial meningiomas and the lack of localized and specific symptoms the diagnosis of these tumors can prove to be challenging. The presence in the neck as a parapharyngeal space mass is quite rare [4]. We present a rare case of parapharyngeal meningioma in a 55 year old female patient who presented with a painless right neck mass.

2. Case report

A 55 year old woman presented with a five years history of painless neck mass. The patient noticed a gradual increase in its size over the past year. She denied any shortness of breath, dysphagia, hoarseness or hearing problems at any time. The patient did not have any neurological or constitutional symptoms.

On clinical examination the mass (Fig. 1) was found to be firm, non tender, fixed and non pulsatile, without being attached to the overlying skin. The patient did not have any skin changes, bruits, palpable lymph nodes or associated neck masses. Upon intra-oral

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Fig. 1. Right side neck mass measuring about 15 cm in diameter.

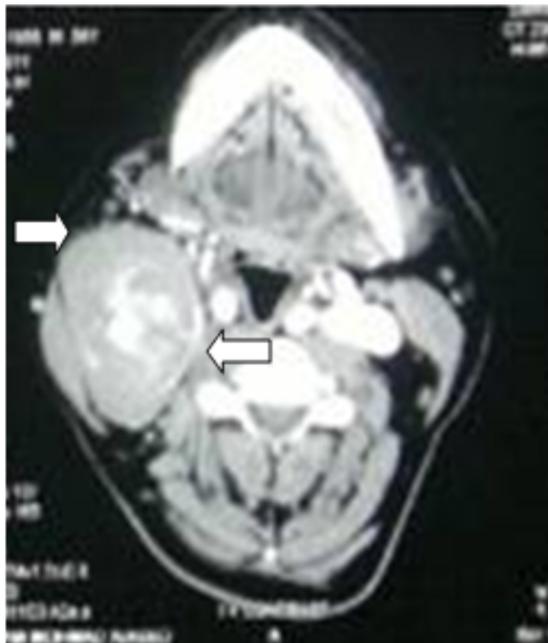


Fig. 2. Axial CT imaging demonstrating large well defined mildly enhancing soft tissue mass.

examination the tonsils were found to be regular sized and centered, however the lateral pharyngeal wall was pushed medially.

A CT scan (Fig. 2) and an MRI (Fig 3.) were performed, which demonstrated the presence of a large, well defined, mildly enhancing soft tissue mass measuring about $8.5 \times 6 \times 5$ cm located in the right carotid sheath. The mass extended from the level of the thyroid gland into the skull base jugular foramen superiorly, and displayed dense peripheral and central calcification. The surrounding structures were compressed by the mass, and a few small adjacent lymph nodes were seen.

Fine needle aspiration (FNA) of the mass showed sheets of cells with frequent microfollicle formation. The surrounding colloid material displayed grooving as well as nuclear pseudo-inclusions and scattered psammoma bodies. The pathologist could not make a clear diagnosis on FNA only, and asked for a tissue diagnosis.

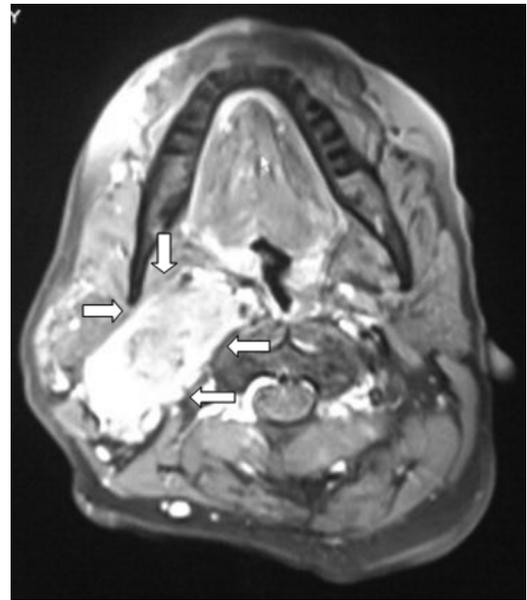


Fig. 3. MR imaging demonstrating a soft tissue mass compressing adjacent structures.

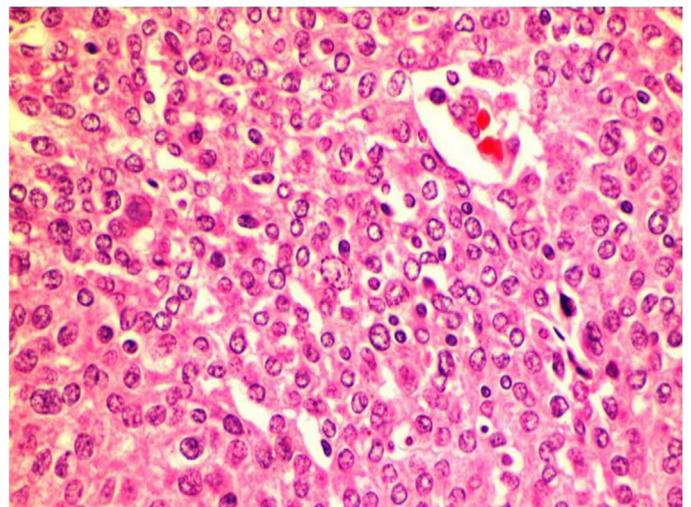


Fig. 4. Histological features of extracranial meningioma demonstrating meningeothelial cells with intranuclear inclusion and multiple psammoma bodies.

Although the patient had no symptoms, the mass was obvious on her right upper neck, and was disfiguring, the patient underwent right cervical exploration for excisional biopsy and because of the encasement of the right internal jugular vein she underwent partial excision of the mass. Histological examination (Fig 4) revealed a tumor composed of bland looking meningeothelial cells with intranuclear inclusions, arranged in a syncytial pattern. Multiple psammoma bodies were also observed, the mass presented with a mitotic index less than 3/10HPF.

No Radiotherapy has been given to the patient, and after 8 months of follow up, the patient had no new complaints and the mass was almost the same size.

3. Discussion

Meningiomas can exist as intracranial or extracranial brain tumors. Despite their similarities, these two forms of tumors have very different characteristics and presentations. Intracranial brain tumors are more common, accounting for 30% of all brain tumors.

These neoplasms arise from meningocytes of neuroectodermal origin and are much more likely to present in female patients [2]. Extracranial tumors 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 tumors are often misdiagnosed [4], fortunately, 80% of extracranial tumors are benign [5].

Extracranial meningiomas are categorized as primary or secondary tumors. Primary tumors are isolated extracranially, and are not associated with an intracranial mass. Secondary tumors, however, arise as extensions of intracranial masses [4]. Surgical defects, foramina in the cribriform plate and the skull base and preformed bony pathways are possible pathways for the extension of an intracranial mass into the neck. 20% of all intracranial tumors present with extracranial components [6].

The parapharyngeal space is enclosed by the skull base superiorly, the vertebral column posteriorly, the mandible and the parotid gland anteriorly and the submandibular gland inferiorly. This space contains many organs and tissues. Tumors in the parapharyngeal space constitute less than 0.5% of head and neck neoplasms [7]. Often these tumors are not readily diagnosed due to their similarities between lymph node tumors, paragangliomas, or neurogenic tumors [8], of the cranial nerves, the 7–12th are most likely to be related to the extracranial tumors [9,10].

Extracranial and intracranial meningiomas are difficult to be distinguished histologically, as they both present with solid nests of meningothelial cells arranged in sheets or whorls. A fibroadipose background is often present in both, and psammoma bodies can also be seen. Meningothelial cells often display abundant cytoplasm with nuclei that are vesicular and occasionally vacuolated [3].

Immunohistochemistry is an essential tool in confirming the diagnosis of an extracranial meningioma [11,12]. These types of tumors have a strong tendency to be positive towards vimentin and epithelial membrane antigen (EMA) and are focally positive for S-100, keratin and CEA [13,14]. This was true in the immunohistochemistry of our patient as well.

Meningiomas in the parapharyngeal space present with nonspecific symptoms. The tumors are often not detected for long periods of time due to the fact that localized signs do not exist until the tumor has reached a significant size. This was the case with our patient who had a neck mass for five years without any pain or symptoms of a meningioma other than the neck mass which has been neglected by the patient. 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 [15]. The tumor may present as a neck mass, a bulge in the oral cavity, or a preauricular mass [15].

The varied presentations of parapharyngeal meningiomas create a diagnostic challenge for surgeons. However, a calcification seen on a CT scan and the presence of intracranial extensions and dural components can aid in the identification of an extracranial meningioma. Advanced imaging such as CTs and MRIs are essential diagnostic tools for parapharyngeal masses. These imaging modalities are also extremely useful in pre-operative surgical planning [6].

The long term prognosis of parapharyngeal space meningiomas is unclear, partly due to the rarity of these tumors. However due to the fact that they are histologically indistinguishable from other meningiomas the essentials of the treatment should remain the same [3]. Surgical excision of the mass should be performed if possible [16] 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 [17–19].

4. Conclusion

Primary extracranial meningiomas of the parapharyngeal space are quite rare, and often pose a diagnostic challenge. Imaging modalities can aid in the diagnosis, but pathological examinations are essential in confirming a definite diagnosis. Although the majority of these tumors are benign, surgical excision is the primary method of treatment, and complete post-operative care often requires a multidisciplinary approach.

Conflict of interest

None.

Funding

None.

Consent

Consent form has been filled and signed by the patient.

Author contribution

Nader Albsoul: study design, writing, supervision.

Badi Rawashdeh: manuscript editing.

Ahmad Albsoul: writing.

Simin Golestani: manuscript editing.

Mohammad Abdullah: data collection.

Aasem Rawshdeh: data collection.

Mona Mohammad: data collection.

Mohammad Alzoubi: supervision.

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