Carotid sheath meningioma: Case presentation

Rospadilah Idris, Norlisah Ramli, Kartini Rahmat *

Department of Biomedical Imaging, Faculty of Medicine, University Malaya, 50603 Kuala Lumpur, Malaysia

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Abstract

Meningiomas are common intracranial tumours that can extend extracranially. Primary extracranial meningioma, however is a rare condition occurring about less than 1% of all meningiomas. We present a rare case of primary extracranial meningioma occurring in the left carotid sheath that was initially diagnosed as schwannoma.

The case highlights the use of MR spectroscopy in the evaluation of extracranial meningioma.

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1. Introduction

Meningiomas are common intracranial tumours that can extend extracranially. Primary extracranial meningioma however is a rare condition occurring in about less than 1% of all meningiomas. We present a rare case of primary extracranial meningioma occurring in the left carotid sheath that was initially diagnosed as schwannoma.

Radiologic and MR spectroscopic findings of extracranial meningioma is discussed.

2. Case report

A 54-year-old man presented with hoarseness of voice and difficulty in swallowing for 6 months duration. Physical examination revealed deviation of the tongue to the left with mild fasciculation. The gag reflex was intact. There is left vocal cord palsy indicating XI and XII cranial nerve palsy. There was no neck swelling noted.

Spiral CT scan of the neck demonstrated a mass anterior to the bifurcation of the common carotid artery extending to the skull base (Fig. 1). Subsequently a CT Angiogram performed showed a mildly enhancing soft tissue mass seen above the bifurcation of the left common carotid artery extending to the left jugular foramen with widening of the left jugular foramen. It was reported as a possible left glomus jugulare tumour. Magnetic resonance imaging (MRI) revealed a lesion in the left carotid sheath which is encasing the left internal carotid artery from the level of the bifurcation of the common carotid (Fig. 2a and b). It extended into the hypoglossal canal and jugular foramen with an extradural intracranial component. The lesion returned low signal on T1 and high signal on T2 weighted images. It enhanced homogeneously post contrast. The impression at that time was that of a neurofibroma.

Patient underwent a transcervical excision of the tumour and tracheostomy was performed. Intraoperative findings were of a tumour at the skull base which was attached to the left internal carotid artery. The upper end of the tumour extended above the skull base. Cervical lymph nodes at the Levels 1a and 2 were excised.

The excised tumour had a gross appearance of nodular grayish tissue measuring 1.5 cm × 1 cm × 1 cm. Two nerve structures measuring 3 cm in length and 2.5 cm in length was found arising from the centre of the mass. Histology of the mass showed a hyalinised fibrous tissue infiltrated by nests of tumour cells. The tumour cells displayed round vesicular nuclei and abundant cytoplasm (Fig. 3). Numerous psammoma bodies were present. In focal areas large cells showed a mildly enhancing soft tissue mass seen above the bifurcation of the left common carotid artery extending to the left jugular foramen with widening of the left jugular foramen. It was reported as a possible left glomus jugulare tumour. Magnetic resonance imaging (MRI) revealed a lesion in the left carotid sheath which is encasing the left internal carotid artery from the level of the bifurcation of the common carotid (Fig. 2a and b). It extended into the hypoglossal canal and jugular foramen with an extradural intracranial component. The lesion returned low signal on T1 and high signal on T2 weighted images. It enhanced homogeneously post contrast. The impression at that time was that of a neurofibroma.

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with abundant eosinophilic cytoplasm and round nuclei with prominent nucleoli were present. Immunohistochemistry examination showed that the tumour cells were positive for EMA, vimentin and S-100 protein and negative for MNF 116. In conclusion, this was a meningioma.

Follow up MRI done 6 months post surgery revealed residual tumour within the left carotid sheath and left jugular foremen. MR Spectroscopy showed a high alanine peak at 1.5 ppm in keeping with meningioma (Fig. 4).

Currently there is no worsening of the patient’s symptoms. Patient is still complaining of difficulty swallowing and hoarseness of voice. On physical examination, there is still deviation of the tongue to the left as before and evidence of left XI and XII cranial nerve palsy. Patient is planned for gamma knife surgery or open surgery for the removal of the residual tumour.
3. Discussion

Meningiomas account for 15% of all intracranial brain tumours and 12% of intraspinal neoplasm [1]. It is more common in females than in males, with a ratio of 2:1 and peak incidence at the age of 45 years. Extraparenchymal meningiomas can be either primary or secondary. Secondary extraparenchymal meningiomas which are the extracranial extension of the primary intracranial meningiomas are more common compared to the primary type. Up to 20% of intracranial meningiomas may have an extracranial component [2]. It primarily occurs in the orbit, external table of the calvarium, the nasal cavity, paranasal sinuses and the parapharyngeal spaces.

Primary extraparenchymal meningiomas are uncommon. It comprises approximately 5% of all meningiomas. Commonly it occurs in the head and neck region. However, there are reported cases of ectopic meningiomas occurring in the lung, mediastinum, skin, soft tissue and foot [3]. Primary extraparenchymal meningioma also shows a strong female preponderance.

There are four mechanisms deducted for the formation of extraparenchymal meningioma as described by Hoye et al. Type A involves direct extension from a primary intracranial tumour through the foramina of the base of the skull. It can also project through the destroyed floor of the cranial fossa. Type B involves extraparenchymal growth from arachnoid cells within the sheaths of cranial nerves. Most of the reported primary extraparenchymal meningiomas in the parapharyngeal space are related to the cranial nerves and those arising far from the base of skull are usually related to spinal or sympathetic nerve roots. Type C involves extraparenchymal growth from embryonic rests of arachnoid without apparent connection to the foramina of the skull base or cranial nerves. Type D involves distant metastases from intracranial meningiomas [1]. This case likely represents a meningioma Type C which originates from the cells of the arachnoid of cranial nerves, most likely the vagus nerve which runs within the carotid sheath. For radiological evaluation purposes, the extraparenchymal meningiomas are divided into two groups. The first group are the tumours which arise in the nasal cavity, oral cavity or paranasal sinuses. This tumour can be detected early and are accessible for biopsy. The second group are tumours which arise in the parapharyngeal space and under the base of skull. The second group as in this case, needs detailed radiological assessment before surgery as they are less accessible clinically.

This is a case of primary extraparenchymal meningioma in the jugulo-carotid space. The main differential diagnosis includes schwannoma and paraganglioma. These lesions may be differentiated by certain features. However, in some cases such as in this case it is difficult to distinguish the lesion as they have common features. There has been no description of primary extraparenchymal meningioma occurring in the carotid sheath at the jugulo-carotid space in the literature.

On CT scan, primary ectopic meningioma appears slightly hyperdense with occasional matrix calcification similar to secondary meningioma. Schwannomas and paragangliomas do not demonstrate matrix calcification.

On MRI, meningiomas are iso to hypointense relative to the brain tissue on T1 weighted sequences and with variable high signal intensity on T2 weighted images depending on histological composition. Schwannomas, on the other hand are usually well-defined, lobulated masses which are hypointense on T1 and hyperintense on T2 weighted images compared to the brain white matter without flow voids or a dural tail. Large schwannomas may have cystic foci (25%). Paragangliomas can be differentiated from both meningiomas and schwannomas on MRI by the presence of multiple flow voids within the lesion which give them the “salt and pepper” appearance. This is best seen on the pre contrast T1 weighted images. Less commonly, hyperintense foci may be seen within the lesion on precontrast T1 weighted image representing foci of haemorrhage. Post gadolinium, paragangliomas enhances intensely but heterogeneously unlike meningiomas and schwannoma which usually show homogeneous enhancement [4].

Regarding the pattern of growth, primary extraparenchymal meningioma at the base of skull tends to show extensive skull base infiltration. It infiltrates the surrounding skull base in a centrifugal pattern. Laterally it extends to the temporal bone until the middle ear cavity and infiltrates other skull base structures such as the jugular tubercle, hypoglossal canal, occipital condyle and the clivus medially. This case demonstrates extension of the tumour into the ipsilateral hypoglossal canal and jugular foramen. Extraparenchymal meningioma can produce hyperostosis of the adjacent bones as well as erosion and expansion of bony margins. The bones may have mixed permeative and sclerotic appearances. It can cause loss of the normal cortex resulting in irregular outline of the adjacent bone. However, the bone density and bone architecture are preserved. In this case, the bony changes described above are not seen. In contrast to meningioma, schwannoma usually causes smooth enlargement and scalloping of the adjacent bone with thin well-defined corticated sclerotic margins without invasion of the marrow space. Paragangliomas tend to behave in a more aggressive manner with permeation and destruction of the adjacent bone. There is also associated loss of bone density of the affected bone unlike in meningiomas [5].

This case also illustrates extension of the lesion intracranially with a cerebrospinal fluid cleft seen in between the mass and the adjacent brain. This superior spread with intracranial extension is not seen with paraganglioma. Schwannomas tend to grow superomedially towards the lateral aspect of the brain stem with variable inferior spread into the nasopharyngeal carotid space of the suprahyoid region of the neck. Medial spread to the hypoglossal canal, jugular tubercle and the clivus are less common. Inferior spread to the nasopharyngeal carotid space is also limited. Primary extraparenchymal meningioma at the base of skull may cause occlusion of the jugular vein and encasement of the carotid artery. In this case, the left carotid artery was encased by the lesion [4].
On angiograms, meningiomas are vascular tumours with prolonged vascular blush without arteriovenous shunting while on the other hand paragangliomas are vascular tumours and on angiogram they demonstrate rapid arterial phase tumour blush and early draining veins.

There has been no reported cases of the use of MR spectroscopy in extracranial meniningiomas. MR spectroscopy measures the biochemical content of living tissues. The metabolic information correlates to the anatomical changes found on conventional MRI images. The spectra reflect on general effects upon brain tissue such as neuronal loss, cell membrane turnover, demyelination, necrosis, ischaemia and gliosis. MR spectroscopy studies of extracts of surgical specimens have generally showed an increase in alanine in meningiomas and an increase in myo-inositol in schwannomas [6]. The most characteristic features of meningiomas are the presence of alanine which was demonstrated in this case. Apart from that, high relative concentrations of choline and low concentration of creatinine containing compounds are also features of meningiomas. N-Acetyl containing compound lipids are also absent or low [7]. However, alanine peak may be masked by lactic acid from anaerobic glycolysis or lipids from cell breakdown and necrosis. Hazany et al. showed that alpha-Glx/glutathione (3.76 ppm) elevation is also a reliable marker for meningiomas on proton MR spectra [8]. In conclusion, we present a case of primary extracranial meningoia occurring in the carotid sheath, the appearances which may mimic other common lesions arising from the carotid sheath region. This case demonstrates the MR spectroscopy findings of an extracranial meningioma.

References