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Case Report

Pure posterior chordoid foramen magnum meningioma: A case report and review of literature

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ABSTRACT

Introduction and importance: Meningiomas are extra-axial central nervous system (CNS) tumors that arise from the arachnoid cells of the dura mater. Only 1.8–3.2% of all meningiomas are located at foramen magnum (FM) and pure posterior FM meningioma are very rare. The diagnosis of malignancy in patients with meningiomas has been a controversial issue. Only a histological study can confirm this situation.

Case presentation: We report a case of A 52-year-old female presented with a history of neck pain with progressive spastic quadriparesis.

Clinical discussion: Magnetic resonance imaging MRI T1 and T2 weighted images revealed well-defied pure posterior foramen magnum Lesion. Although the lesion was very sticky to neurovascular components. Simpson grade I was achieved. Histopathology revealed Chordoid meningioma. The patient had a dramatic recovery.

Conclusion: Although choroid meningioma is usually well circumscribed, sticky tumors should be suspected. Recurrence of Chordoid meningioma should be suspected. Total excision should be achieved and routine follow-up should be informed. Reports about chordoid meningioma aren't common, but reports on choroid foramen magnum meningioma are very rare. The opportunity to give the patient a symptom-free and normal life should not be missed in such cases.

1. Introduction

Meningiomas are extra-axial central nervous system (CNS) tumors that arise from the arachnoid cells of the dura mater [1]. Meningiomas represent 14.3–19% of all intracranial tumors, the most common non-glial primary intracranial tumor [1].

Only 1.8–3.2% of all meningiomas are located at foramen magnum (FM). Foramen magnum meningiomas (FMMs) represent a common histological tumor in a rare and eloquent location [1,2]. As these tumors are indolent, there occurs a long interval between onset of symptoms and diagnosis [2].

According to Boulton and Cusimano [3], Most lesions (68%–98%) arising anterolaterally, followed by postolateral, purely posterior and, more rarely, purely anterior [3].

In a review of Kepes et al., a relationship with Castleman syndrome was found, the features of which are delayed somatic and sexual development, hepatosplenomegaly, iron refractory hypochromic microcytic anemia, and bone marrow plasmacytosis with dysgammaglobulinemia [4–6]. This association was not found in the majority of other studies. In our case also, no systemic symptoms were present.

Chordoid meningioma is a rare subtype which is associated with a high likelihood of recurrence [7]. It represents only 0.5%–1% of all meningiomas and is grouped in WHO Grade II tumors [8]. They are mainly distributed in the supra-tentorial region [8. Infra-tentorial chordoid meningioma is rare [7,8].

Our work is a single case report and has been reported in line with the SCARE criteria [21].

2. Case report

A 52-year-old female presented to our department of Neurosurgery (Lattakia, Syria) via community referral with the complaints of neck pain for 9 months in 2016, progressive weakness of all limbs in the form of rotating paralysis starting with weakness in triceps muscle in right upper limb followed by weakness in quadrate muscle of the thigh in right lower limb, then weakness in quadrate muscle of the thigh and biceps muscle in the left lower limb & left upper limb for 4 months with bowel bladder disturbance in the form of urgency & frequency. On

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neurological examination patient was found to be quadri-paretic having muscle power Medical Research Council (MRC) grade 4 on right side & 3 on left side rendering her gait to be hemiplegic and walk with support. She also complained of hyperesthesia in the C2 dermatome (occipital region).

All her deep tendon reflexes were exaggerated with bilateral planter extensor.

She also complained of difficulty in swallowing and respiratory compromise.

There were no subcutaneous nodules, hypopigmented macules, or other stigmata of neurofibromatosis (NF). All other routine investigations were normal.

The patient's review of systems and additional medical history surgical, family, psychosocial and pharmacologic were unremarkable.

Magnetic resonance imaging (MRI) T1 and T2 weighted image revealed well-defied dural-based mass lesion at foramen magnum, located posterior to neuroaxis. Post-contrast T1 weighted images revealed inhomogenously enhancing dural based lesion in the foramen magnum (Fig. 1).

After obtaining the patient's informed consent, surgery was planned.

The procedure was done by a consultant neurosurgeon with 17 years of experience.

Patient underwent surgery by posterior approach. The patient was placed in the park-bench position.

The incision was extended through the deep layers and to the level of the suboccipital bone along the midline avascular fascial planes.

Blunt dissection was done to prevent injury to the vertebral artery.

C1 laminectomy was performed to allow for a wider dural opening and adequate lesional exposure.

The dura was opened in a Y-shaped fashion with a more inferior extension for more lateral mobilization of the dural flaps.

The tumor was encapsulated and well-defined. But it was very sticky to neurovascular components. It was also firmly attached to the dura. Due to the adhesions, a gross total resection was challenging to achieve. The tumor was first devascularized as much as is safely possible along its lateral attachments, and then centrally debulked. The vertebral arteries were dissected free and covered with a small cottonoid patty. Once the tumor was centrally debulked, its capsule was mobilized and all of the pial adhesions was sharply divided and small tumor feeders coagulated. Complete excision was done with coagulation of dural base.

The tumor was resected with preserving the neural and vascular structures. Simpson grade I was achieved.

Histopathology examination showed cords of eosinophilic vacuolated cells in abundant myxoid matrix. The infiltration of lymphocytes or plasma cells was observed at the margin of the tumor.

Cells were immunopositive for epithelial membrane antigen (EMA) and vimentin. However, they were negative for glial fritillary acidic protein (GFAP) and S100. Based on these findings, a diagnosis of chordoid meningioma was rendered.

Post-operative MRI after 3 months shows no residual tumor(Fig. 2).

Patient is on close follow-up for the past 4 years and carrying out her routine life activities. Until now there has been no recurrence.

3. Discussion

The first case of meningioma arising at the foramen magnum was observed by Hallopeau at Lariboisiere Hospital in 1872 [9].

Foramen magnum meningioma is often identified only at an advanced stage due to the unusual and unrecognized pathognomonic symptomatology [10,11].

George et al. defined this area as a zone delimited anteriorly by the lower third of the clivus and upper edge of the body of axis (C2);

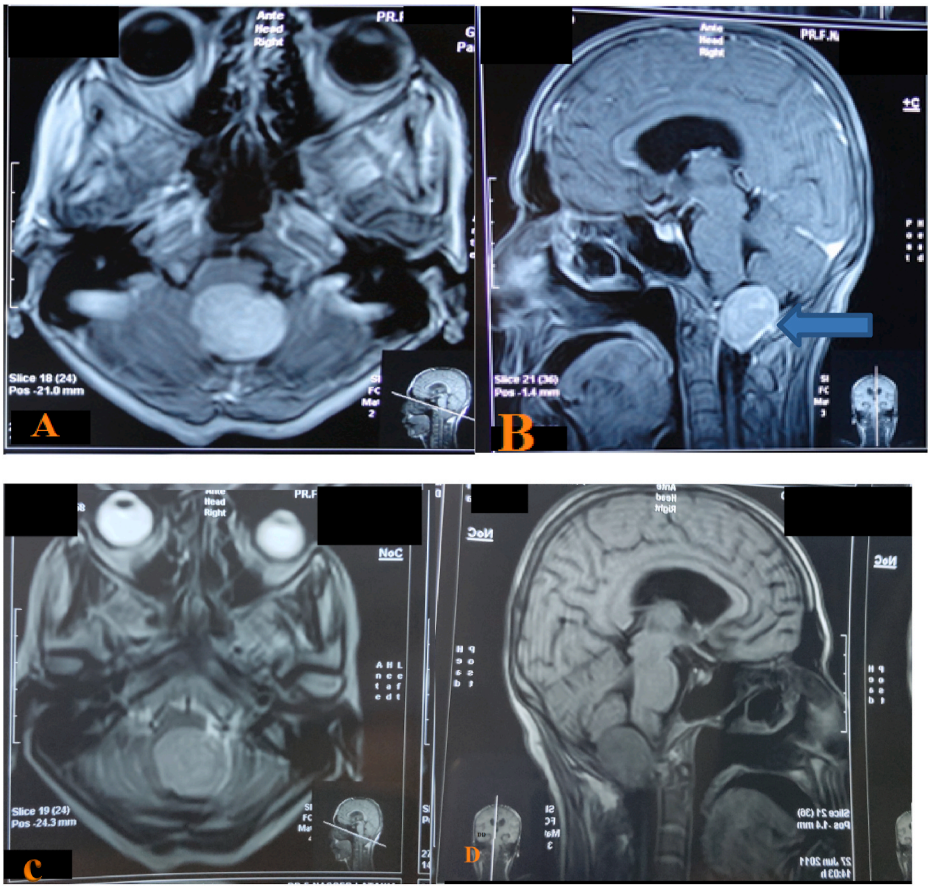

Fig. 1. A: Post-contrast T1 weighted axial image, B: Post-contrast T1 weighted sagittal image: magnetic resonance imaging (MRI) showing pure posterior placed, inhomogenously enhancing dural based lesion measures 2 *2.5 CM in the foramen magnum (Blue arrow). C: T2 weighted axial image shows isotense lesion in the foramen magnum
T1weighted sagittal image shows hypo-tense lesion in the foramen magnum. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

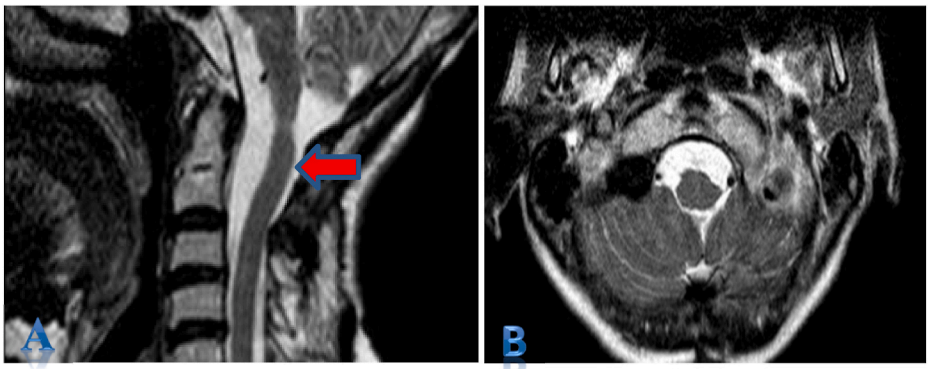

Fig. 2. Post-operative T2 -MRI (A: sagittal, B Axial) after 3 months shows no residual tumor. Red arrow indicates Normal spinal cord surrounding by CSF. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

laterally by the jugular tubercles and upper aspect of C2 laminas; and posteriorly by the anterior edge of the squamous occipital bone and C2 spinous process [9,12].

The term chordoid meningioma was given by Kepes et al. in 1988 to describe a meningeal neoplasm that has chordoma-like appearance on histopathology [13].

Marhx-Bracho et al., reported a case of 3-year-6-month-old boy with chordoid meningioma in the foramen magnum complained from quadriparesis, cervical pain, and neck stiffness. (MRI) demonstrated an intradural–extramedullary well-circumscribed, homogeneous enhancing mass, in the anterior part of the foramen magnum with cervical extension. The operative technique was the extreme-lateral craniocervical retrocondylar approach with total removal and dural coagulation. Postoperative radiotherapy was not given as there was complete resection of the tumor [14].

Kumar et al. reported a case of 22-year-old female with chordoid meningioma in the foramen magnum who admitted with progressive spastic quadriparesis with bowel and bladder involvement. The tumor was decompressed postoperatively, patient showed dramatic improvement. Patient was planned for postoperative radiotherapy because of residual tumor on post-operative MRI [15].

The rarity of our case comes from the position of the tumor which is a pure posterior foramen magnum meningioma and from the histopathology which was confirmed as WHO II grade, Chordoid meningioma. Postoperative radiotherapy was not given as there was complete resection of the tumor.

From the first case of foramen magnum meningioma reported up to now, there were many cases reported; however, the mention about the chordoid foramen magnum meningioma is very rare. Besides the cases in the reports of Marhx-Bracho et al. [14] and Kumar et al. [15], ours is the third case.

In a study of 42 cases of chordoid meningioma by Couce et al., the majority (88%) were large and supratentorial. No manifestation of systemic disease was there. None of the patients had a lesion at FM [7].

Tena-Suck et al. in their report of ten cases of chordoid meningioma found that 80% of the tumors were in supratentorial region. The age range was from 30 to 67 years old (mean, 34.2 years). The duration of symptoms varied from 3.5 months to 5 years (mean, 14.1 months). No systemic symptoms were noted and none of the patient had lesion at FM [16].

Yang et al. in their recent series of sixty cases of chordoidmeningioma have reported 33 tumors in infratentorial region (31 skull base and 2 intraventricular) and 2 in cervical region. As the exact breakup of these infratentorial lesions has not been mentioned and none of the lesions are mentioned in FM location, we assume that no lesion was present in this region [17].

Differential diagnosis of tumor with chordoid/myxoid features includes chordoma, chordoid glioma, myxoid chondrosarcoma, myxopapillary ependymoma, and mucinous metastatic carcinoma. A panel of immunohistochemical markers is required to differentiate between these tumors. Chordomas are EMA/cytokeratin (CK)/S-100 positive. Chordoid gliomas are GFAP positive; chondrosarcomas stain positively with S-100 while chordoid meningioma is EMA positive. Myxopapillary ependymomas are exclusively located in conus medullaris, cauda equina, and fium terminale and are GFAP positive; metastatic carcinoma shows reactivity with CK [18].

Histologically, these tumors consisted of spindle or epithelial cells in a myxoid matrix. Two thirds of adult tumors showed mild-to-moderate inflammatory cells infiltration. It was infiltrated by many reactive lymphoplasmacytes with formation of follicles and germinal centers [7]. The inflation of lymphocytes could be seen in our case, especially in the margin of the tumor, but without forming germinal centers. Immunochemical staining could be helpful for differential diagnosis. Chordoid meningioma is often immunoreactive by EMA, vimentin, PR, and D2-40, and occasionally positive by S-100 and cytokeratin, but consistently negative by GFAP [19]. Thus, a panel of antibodies comprising D2-40, EMA GFAP, and S-100 can distinguish CM from its histologic mimics as depicted in (Table 1).

In a study by Violaris et al., the majority (55.2%) of recurrences for Grade II meningioma was observed within 2 years from surgery and 94.7% within 5 years from surgery [20].

Fortunately, our patient shows no recurrence after 4 years of operation.

Surgical management of craniocervical junction meningiomas continues to present as a challenge to neurosurgeons especially in cases involving neighboring neurovascular structures.

The diagnosis of malignancy in patients with meningiomas has been a controversial issue. Only a histological study can confirm this situation. The association of pathological examination and imnunohistochemical study can make a diagnosis of chordoid meningioma.

Table 1
Immunohistochemical profile in the differential diagnosis of tumors exhibiting chordoid morphology.

Tumor	D2-40	EMA	CK	GFAP	S-100
Chordoid meningioma	+	+	±	–	–
Chordoid glioma	+	+	–	+	+
Chordoma	–	+	+	–	+
Extraskeletal myxoid chondrosarcoma	–	±	±	–	±
Skeletal myxoid chondrosarcoma	+	–	–	–	+
Low-grade chondrosarcoma	+	–	–	–	+
Myxopapillary ependymoma	–	±	–	+	+
Metastatic carcinoma	–	+	+	–	–

EMA = Epithelial membrane antigen, CK= Cytokeratin, GFAP = Glial fibrillary acidic protein.

4. Conclusion

Although choroid meningioma is usually well circumscribed, sticky tumors should be suspected.

Complete resection without neurovascular injuries is the key for managing such lesions.

Recurrence of Chordoid meningioma should be suspected. Total excision should be achieved and routine follow-up should be informed.

Ethical approval

This study was not applicable for ethical approval.

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Author's contribution

Dr. Ali Hammed (corresponding author): Contribution to the paper: first author, data collection, data analysis and interpretation, writing the paper.

Prof. Dr. Moufid Mahfoud: Contribution to the paper:
Main Surgeon. Treatment and examination of the patient.
Writing Case Presentation.

Dr. Salah Hammed: Contribution to the paper: Writing the paper.

Dr. Alaa Sulaiman: Contribution to the paper: Writing the paper.

Dr.Adnan Najm: Contribution to the paper: Writing the paper.

Registration of research studies

The case report at hand is not a first-in-man case report of a novel technology or surgical technique, therefore a registration of these case reports according to Declaration of Helsinki 2013 is not required.

Guarantor

Ali Hammed.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Declaration of competing interest

All authors declared no conflict of interest.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.amsu.2021.102325.

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