Primary calcified rhabdoid meningioma of the cranio-cervical junction: A case report and review of literature

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Abstract

Rhabdoid meningioma (RM) is a relatively new, rare, and aggressive subtype of meningioma, classified as Grade III malignancy in 2000, 2007 versions of WHO classification of the central nervous system. We reviewed the data available from all published cases of RMs. To the best of our knowledge, there are more than 100 published cases of RMs; none have documented extensive calcification or origin from the cranio cervical junction. We report the first case of a totally calcified (stony mass), primary RM, at the cranio cervical junction. Also, we highlighted the role of the transcondylar approach to achieve microscopic total removal of such a challenging lesion. A 37 year old female, allergic to erythromycin, presented with 5 years of progressive right upper extremity numbness and weakness, right facial numbness, and occipital pain. Imaging demonstrated a large calcified mass at the right posterior–lateral margin of the cranio cervical junction, encasing the right vertebral artery and right PICA loop. Patient underwent microscopic total resection of the lesion. Pathological diagnosis was confirmed as RM with atypical features. Subsequently, the patient received postoperative intensity modulated radiotherapy (IMRT) on the tumor bed, and close follow up imaging showed no recurrence 2 years after surgery. We report the first case of a primary RM originating from the cranial cervical junction; also, it is the first case to present with extensive calcification in this morphological subtype. We also conclude that RM has now become a feature of newly diagnosed cases and not only a disease of recurrent cases as it was thought in the past. Since RMs are typically considered aggressive, total surgical resection with close follow up and postoperative adjuvant radiation should be considered. However, the adjuvant therapy of each separate case of RM should be tailored according to its particular histopathologic profile.

Keywords: Calcified, cranio cervical junction, primary, rhabdoid meningioma

INTRODUCTION

Rhabdoid meningioma (RM) is a relatively new subtype of meningioma, first described in 1998 by Kepes et al.[1] Then, Perry et al.[2] in the same year had published a bigger case series of 15 patients under the
frank name of (rhabdoid meningioma). RMs are quite rare, accounting for only 0.004% of all meningiomas.[2–5]

RM is an aggressive variant of meningiomas, classified as Grade III malignancy in 2000, 2007 versions of WHO classification of the tumors of the central nervous system.[6,7] There are more than 100 published cases of intracranial RMs; none have documented extensive calcification or origin from the cranio-cervical junction. We report the first case of a totally calcified, primary RM at the cranio-cervical junction. Also, we put the spot on the role of the transcondylar approach to achieve microscopic total removal of such challenging lesion to increase the recurrence-free survival periods.

CASE REPORT

History and examination
A 37-year-old female, allergic to erythromycin and who had family history for heart disease, hypercholesterolemia, hypertension, and diabetes, presented to neurosurgery clinic with 5 years of progressive subjective right upper extremity numbness and weakness. She also had some right occipital pain and facial numbness. She denied bowel or bladder changes. Her physical exam showed that she had subjective decreased sensation to pinprick in the right V1 and V2 distributions that became more marked in the V3 distribution. She had full strength in her left upper and left lower extremity. She had right-sided weakness of grade 4+ in most of the muscle groups on MRC scale. She had diminished sensation to light touch in a glove distribution in her right upper extremity. Sensation to light touch was otherwise normal in her left upper extremity and bilateral lower extremities. She had diminished proprioceptive sense in her right upper extremity as well as her bilateral lower extremities, right side worse than left side. Reflexes were 2+ and symmetrical. She had negative Hoffman's and Babinski's signs, positive Romberg sign, and difficulty with tandem gait. Finger-to-nose testing demonstrated no dysmetria.

Imaging
With and without contrast, head and cranio-cervical junction computed tomography demonstrated a calcified mass, measuring 4.3 × 2.5 cm in the axial cuts [Figure 1a], and its height was 3.5 cm in sagittal reconstruction [Figure 1b]. The mass was occupying the postero-lateral portion of the right compartment of the cranio-cervical junction, displacing the medulla oblongata and upper spinal cord anteriorly to the left side. CTA demonstrated that the anterior portion of the mass engulfing the right vertebral artery [Figure 2], hence transcondylar approach was planned for surgery.

Operation
An elective resection of the tumor was done using the transcondylar approach. Drilling of the posterior half of the occipital condyle along with the lateral mass of atlas bone was done to get the anterior pole of the tumor and perform careful dissection of the vertebral artery without any traction on the artery or the lower cranial nerves. The transcondylar approach is one of the skull base approaches that respect the paramount principles of skull base surgery by removing the basal bone to shorten the distance, improve the angle of work, and avoid any traction on the neural or vascular tissues. Intraoperatively, the tumor was a rock-like mass with scanty soft tissue at its periphery; it was engulfing the vertebral artery, posterior inferior cerebellar artery, and rootlets of lower cranial nerves in its substance [Figure 3]. The tumor was dissected from all these structures, and microscopic total resection was achieved. Frozen section suggested the diagnosis of meningioma.

Pathological findings
Grossly the specimen consisted of small fragments, most of which were sufficiently bony requiring
decalcification and a minority was soft tissue. The soft tissue consisted of plump epithelioid cells with round to oval nuclei with prominent, usually single nucleolus. The striking feature was the nuclei frequently pushed to one side by an eosinophilic inclusion. The growth pattern was of nests and vague large whorls, with the foci becoming sheet-like. Necrosis was not present and mitoses were infrequent (2 or less/10 hpf).

The bony fragments had large, semi-confluent lamellated mineralized nodules, which were larger and somewhat different from psammoma bodies. The appearance was more suggestive of the lobular areas of tumor that had become acellular and calcified. Nevertheless, in the small amounts of viable tumor that could be found nestled around and between these mineralized concretions, the cells again had the eosinophilic rhabdoid morphology described. A vimentin immunostain was strongly positive and highlighted the cytoplasmic inclusions [Figure 4]. An MIB-1 proliferation index was quantitated at 12.9% (32/247) [Figure 5]. Although the amount of viable tumor was small, it exhibited WHO atypical features of large vesicular nuclei with prominent nucleoli, and focally sheet-like areas, and a significantly elevated MIB-1 index. The predominant cellular morphology was unequivocally and predominantly “rhabdoid,” although the tumor did not have a high mitotic rate or other anaplastic features.

**Postoperative course**

The patient recovered very well. She noticed subjective improvement of the facial numbness and right-sided weakness few days after surgery. Definite histopathologic diagnosis of RM was made, and she received adjuvant intensity-modulated radiotherapy (IMRT) to the resection cavity (5220 Gy in 30 fractions). Her follow-up images after 14 months of surgery showed no recurrence in the tumor bed [Figure 6]. And the patient is alive normal with no disease till the date of writing this article (2 years after surgery).

**DISCUSSION**

We have reviewed all available published data regarding the RM. To the best of our knowledge, the number of published cases diagnosed as RMs reached more than 102 patients (2 cases of spinal RM, and 100 cases of intracranial RM). Most of them are in the English literature and a few cases have been published in non-English language too.[2–5,8–12] However, the number of tumor specimens is much higher because of high incidence of recurrence in these cases.

In the first series of four cases published by Kepes et al.,[1] they suggested that RMs are highly aggressive tumors and the rhabdoid phenotype represents a marker of malignant transformation in meningiomas that indicates a poor prognosis.[1] The WHO criteria for classification of RM are based solely on the presence of the distinctive rhabdoid morphology in the majority of the specimen. Additional features such as high mitotic index, or anaplastic (malignant) features are supportive, but not necessary.[7,13] One can only speculate whether with a larger mass of viable tumor, the amount of “rhabdoid tumor” might have been less; but this cannot form the basis for a diagnosis. Anecdotally the presence of such abundant calcification, the relatively modest proliferative markers, and absence of frank anaplastic features may indicate that this lesion will behave more favorably than the WHO Grade III that is a canonic requirement for RM.

Not all RMs behave with the same aggressiveness; there are benign, atypical, and malignant meningiomas with rhabdoid morphology. However, most of the cases behave aggressively.[4,14–17] Benign RM is the presence of rhabdoid features, either diffuse or focal, lacking the histological features of malignancy.[4,18]
This lack of malignant features significantly inhibits the clinically aggressive behavior of RMs.[2,4,18] In Perry et al.’s series, 4 of 15 tumors were clinically classified as benign tumors and the clinical course was less aggressive than in tumors histologically classified as atypical or malignant ones. In our review, we found that 11% of diagnosed RMs was classified as benign tumors. So, the term benign RM became more acceptable and many neurosurgeons now believe that presence of rhabdoid features alone without the malignant features or elevated MIB-1 LI may be associated with benign course of the disease as well as other conventional meningiomas.[4,18] In the present case, there were no malignant features, but cytological atypia was found. According to our review, Atypical RMs constitute 28% of all published cases of RMs.

Most of the RMs are mixed with otherwise conventional types of meningioma.[19] However, many reported cases have diffuse or prominent rhabdoid morphology.[1,2,4,8,9,18–24] Rhabdoid features are likely to be associated with malignant features and elevated MIB-1 LI. The most aggressive RMs are those mixed with papillary morphology.[13,15,17,18,23,25–29] The question is how to manage cases of RMs without malignant features. Perry et al. suggested close follow-up for cases displaying only focal rhabdoid cytology within classic or benign meningioma.[2] A recent study suggested aggressive management and close clinical follow-up for cases with atypical histology and lacking malignant features, like the one reported here. The authors postulated that RMs with atypical histology generally behave aggressively.[16] However, the jury is still out as to whether conventional meningiomas with focal rhabdoid areas or rhabdoid meningiomas without cytological atypia are as aggressive as their atypical cousins.[6]

In the first published two series and the following early reports, an interesting feature found was that rhabdoid morphology was evident in recurrent cases of meningiomas and it was thought to be a transformation of conventional types of meningioma.[1,2,12,13,26,30–32] Almost all reported cases of recurrent RMs agreed that the rhabdoid components became more prominent with subsequent resections. [2,13,25,26,33,34]

Few years ago, the published cases of primary diagnosed RM had increased. Recently, with increased awareness of this histological subtype, the number of primary diagnosed cases have increased. Now most of the reported cases show diagnosis of RM in the first specimen. Furthermore, in one recently published series of 13 patients, rhabdoid features were identified at the first resection in all patients. In this study, all tumors had conventional meningioma features mixed with the rhabdoid areas.[17] The authors of this study very recently republished a new series of another six cases of rhabdoid papillary meningioma with the rhabdoid features appearing also in the first presentation.[16] In our analysis of the available RM cases, we found that 79% of RM diagnosis was made in the first specimen. Now one can say that rhabdoid morphology is more common in primary diagnosed cases and consistently increases with subsequent resections.

Calcification is not a dominant feature of RM.[14,35] Vassilouthis and Ambrose have pointed out that calcifications are absent or scant in malignant meningiomas, and they considered cystic component to be a CT criterion to evaluate the aggressiveness of meningiomas.[36] CT remains the study of choice in evaluating bone changes in meningiomas. Bone involvement occurs in approximately 20-25% of all types of meningiomas, Bone osteolysis has been mentioned as a factor in helping to predict the malignancy of meningiomas.[37] Kim et al. published a series of MR findings in 15 patients of RM; in this series, 8 patients had CT for the brain and little calcification was seen only in one case. The authors suggested that primordial edema and cystic component may be strongly associated with RM as these features were relatively common in their cases.[14] Data available from most of the reported cases were insufficient with regard to the CT findings, as most authors neglect mentioning the CT findings in their reports, but scant
calcification was reported only in other two cases.[28,38] Apart from these three cases, we did not find any published case that presented as a diffusely hard calcified rock-like mass, as our case. This diffuse calcification may represent a diffuse metaplastic transformation in otherwise conventional type of meningioma was mixed with the rhabdoid feature. Another case of RM was presented with hyperostotic bony involvement of the frontal and sphenoid bones; however, most of the bony specimen showed no tumor infiltration.[5]

The site of presentation in our case is extremely rare too. This case may be the first published case of RM that presented as a primary frank foramen magnum RM involving the cranio-cervical junction down to the level of posterior arch of C1. One reported case showed spine metastatic multifocal and diffuse leptomeningeal enhancement; in this case, there was metastatic tumor at the cervico-medullary junction. [25] In our primary case, the meningioma originated from the cranio-cervical junction, exactly the posterolateral part of foramen magnum. Another case involved the foramen magnum as an extensive extension after multiple resections of tentorial meningioma.[26]

Gross total resection remains the mainstay of treatment of all types of meningiomas, whether benign or malignant. In many published cases of RM, the cause of rapid recurrence apart from the aggressive nature of the tumor was the subtotal resection (STR) of the lesion.[2,13,25,26,33,34] Neurosurgeons should make every effort and pay all attention to achieve GTR as much as they can, minimizing patient's morbidity, mortality, and suffering with big residuals and rapid recurrence after that. Because of this, it is very important to achieve the lowest Simpson grade even when dealing with such aggressive meningiomas. Most of these aggressive meningiomas in the current practice receiving postoperative adjuvant radiation therapy which make the next surgery for residual and recurrence more challenging due to arachnoid scarring and loss if anatomical plans. So, the GTR in the first surgery is vital, especially in such aggressive meningiomas.

Transcondylar approach is the best approach to remove the anterior, and anterolateral Foramen magnum (FM) meningiomas, many neurosurgeons would not do transcondylar approach for posterolateral FM meningiomas such as the presented case.[39] However, it's crucial to plan how one will dissect the vertebral artery, PICA, and the lower cranial nerves, in this case these vital structures were located just medial to the atlanto-occipital joint [Figure 1c] necessitating its drilling to give us space for dissection instead of applying dangerous and unsecured traction on the vital structures from behind. This case also is an excellent example of the ability of the transcondylar approach to shorten the distance and provide a better angle of work on the anterior pole of this tumor. Excessive removal of bone through drilling the condyle, lateral mass, and the arch of C1, transpositioning of the extradural vertebral artery (mainly V3 segment) after opening the C1 cervical foramen, and drilling the lateral mass of C1 and posterior part of the occipital condyle. All these enable the surgeon to work around a bigger surface area of the tumor and successfully dissect vital structures such as vertebral artery, PICA, and rootlets of lower cranial nerves from such a rocky mass without causing any harm to the patient. In this case, MTR was achieved and patient received postoperative adjuvant IMRT on the empty tumor bed to gain the maximal Recurrence free survival (RFS), as she was relatively young and the tumor showed atypical features.

**CONCLUSION**

We report the first case of a primary RM originating from the cranial-cervical junction; also, it is the first case to present with extensive calcification in this morphological subtype. Since RMs are typically considered aggressive, total surgical resection with close follow-up and postoperative adjuvant radiation should be considered. However, the treatment plan of each separate case of RM should be tailored
according to its particular histopathologic profile. The transcondylar approach is of great importance to help in achieving microscopic total resection. It led to safe dissection of the anterolateral structures of the foramen magnum, decreasing the morbidity and chances of mortality in this case. Finally, after reviewing all available cases of RMs, we can conclude that the number of RMs diagnosed in the first specimen now, after 14 years of adoption of this variant, is much exceeding the reported recurrent cases.

Footnotes

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REFERENCES


**Figures and Tables**

**Figure 1**

CT of cranio-cervical junction, (a) Axial cut shows the mass attached to right posterior rim of atlas, (b) Sagittal
reformation shows the relation of the mass to the bony structures of the cranio-cervical junction, (c) Axial bone window shows the relation of the mass to the atlanto-occipital joint and the vertebral artery

**Figure 2**

CTA of cranio-cervical junction. Right and middle compartment in this figure is a coronal reformation of the vertebral artery pathway in the cranio-cervical junction. The right and middle arrows point to the vertebral artery passing inside the calcified mass. Left arrow points to the vertebral artery in axial insight encased within the anterior pole of the tumor with the PICA running inside the mass

**Figure 3**
Intraoperative microscopic picture showing the free dissection of vertebral artery, PICA loop, and the spinal accessory nerve at the anterior pole of the rock-like tumor after drilling of the occipital condyle.

Figure 4
Histopathologic slides of the specimen, (a) Low magnification of the growth pattern of the non calcified portion of the lesion. Note the sheet-like growth interrupted by scattered nodules or nests (H and E, ×20), (b) High magnification of the cytology of the meningothelial tumor cells; note the eosionophilic inclusions in the cytoplasm of the tumor cells (H and E, ×60), (c) Calcified portions of the specimen showing the lamellated vaguely psammomatous concretions. Note that in the meningioma between calcified areas, the morphology (black arrow) remains rhabdoid (H and E, ×20), (d) Vimentin immunohistochemistry showing the strong cytoplasmic labeling characteristic of rhabdoid meningioma (×60)

**Figure 5**

Immunohistochemical staining for Ki-67 antigen, (a) ×60, MIB-1 proliferation index was quantitated at 12.9%, (b) Shows
Post-contrast axial view MRI follow-up done after 14 months shows no recurrence in the tumor bed.