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Case Report

Challenging diagnosis and management of anaplastic meningioma in a pediatric patient: A case report ☆,☆☆,★

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ABSTRACT

This case report highlights the diagnostic and therapeutic journey of a 16-year-old female presenting with chronic headaches, ultimately diagnosed with anaplastic meningioma. Despite its rarity in pediatric patients, anaplastic meningioma necessitates swift recognition and management due to its aggressive nature. Imaging findings, including CT and MRI, initially suggested a provisional diagnosis of hemangiopericytoma, emphasizing the diagnostic challenge posed by this condition. Surgical intervention revealed unexpected histopathological findings, highlighting the importance of thorough evaluation. Treatment involved frontal craniectomy and excision followed by adjuvant radiotherapy. While the patient's postoperative course was uneventful, histopathology confirmed the presence of anaplastic meningioma, leading to the adjustment of her clinical management. This case shows the need for heightened suspicion and comprehensive evaluation in similar presentations to facilitate timely intervention and improve patient outcomes.

Introduction

Anaplastic meningioma, classified as a WHO grade III, is a rare subtype, accounting for 1%-3% of all meningiomas [1]. They are highly aggressive with a worse clinical course, including a higher risk of recurrence ranging from 50% to 94%, and increased mortality, with the median survival typically less than 2 years and the 5-year survival ranging from 35% to 61% [2–4]. Pediatric meningioma, while rare, also presents a worse

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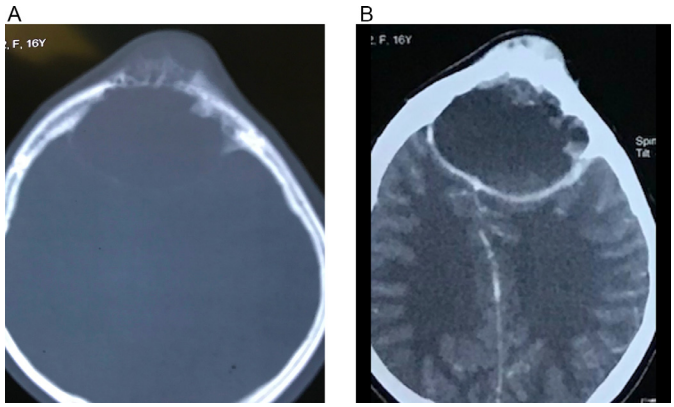

Fig. 1 – (A) Axial CT head bone window shows the extra-axial cysto- solid lesion in left frontal region with erosion of adjacent frontal bone. (B) Axial contrast enhanced CT image showing heterogeneously enhancing extra-axial lesion in left frontal region with erosion of adjacent calvarium and extension to the scalp.

prognosis compared to adults [5]. Anaplastic meningiomas are often found in the cerebral and cerebellar convexities, arising either de novo or from previously atypical or benign growth [6]. Typically, they are diagnosed incidentally or may manifest with nonspecific symptoms such as headache, dizziness, or focal neurological deficits [7]. The primary treatment approach involves surgical resection, often followed by adjuvant radiotherapy to improve outcomes [8]. Herein, we present a case involving a 16-year-old girl diagnosed with anaplastic meningioma in the frontal region during an evaluation for chronic headaches, and subsequently managed through surgical resection and adjuvant radiotherapy.

Case details

A 16-year-old female with no known past medical history presented with a history of headaches for 2 months. The headache was insidious in onset, more on the left occipital aspect, initially mild but recently increasing in intensity over the last 2 months without a change in character. She also mentioned about the swelling in frontal part of scalp, which was first noticed by her friends. The headache had no diurnal variation, no association with the menstrual cycle, and the patient did not experience fever, seizures, photophobia, visual disturbances, or weight loss, or gain. Family history was not significant. The patient did report a slip in the bathroom three months ago but did not lose consciousness or experience vomiting post-fall. She is a nonsmoker and nonalcoholic. Past episodes of headache used to be relieved with NSAIDs, but for the last 2 months, no medication has helped her.

The patient's vitals on presentation were within normal ranges: Blood pressure 120/80 mm Hg, Pulse 72/min, RR 16/min, and afebrile. Systemic examination, including detailed neurological examination, was unremarkable. Swelling and tenderness in the frontal part of scalp is also noted. Her laboratory biochemical and electrolyte investigation shows normal findings. Due to the change in intensity of the headache, the patient was scheduled for an MRI 15 days later due to a long waiting list. Meanwhile, the patient wanted to undergo a CT scan first suspecting her headache to be due to her fall in the bathroom.

Surprisingly, the CT scan showed a large extra-axial heterogeneously enhancing cystic-solid lesion in the left frontal region with multiple calcifications in the solid component and the periphery, with erosion of the adjacent frontal bone (Figs. 1A and B). Protrusion of soft tissue component in frontal part of scalp is also noted. The radiologist made a provisional diagnosis of hemangiopericytoma.

After 15 days, the patient underwent her scheduled MRI brain with contrast which showed a large, well-defined heterogeneously enhancing extra-axial lesion in the bilateral frontal region (predominantly on the left side) with Calvarial extension and signal intensity changes (Figs. 2A and B). The radiologist suggested it to be a malignant lesion with 2 differentials: Hemangiopericytoma or anaplastic meningioma.

As the headache was progressing, the patient was scheduled for surgery within a week and underwent frontal craniectomy and excision with lax duroplasty and cranioplasty. Intraoperative findings suggested a $5 \times 5 \times 5$ cm extradural firm-to-hard mass arising from bone infiltrating the dura, septation, and with blood-filled cavities, honeycombing in the frontal region eroding the bone. The team of neurosurgeons concluded the mass to be likely an aneurysmal bone cyst.

The postoperative hospital stay was uneventful, and the patient was discharged after 1 week with follow-up advice. Postoperative neurology was intact.

The histopathology report arrived 2 weeks later and completely altered the course of the patient's clinical and surgical history showing the typical histological features of an anaplastic meningioma with infiltration into the bone. The patient was advised to an immunohistochemistry panel including the Ki-67 index for further evaluation, but the patient denied the investigation due to unaffordability. The hospital arranged for a PET scan to detect any metastasis, and none were found.

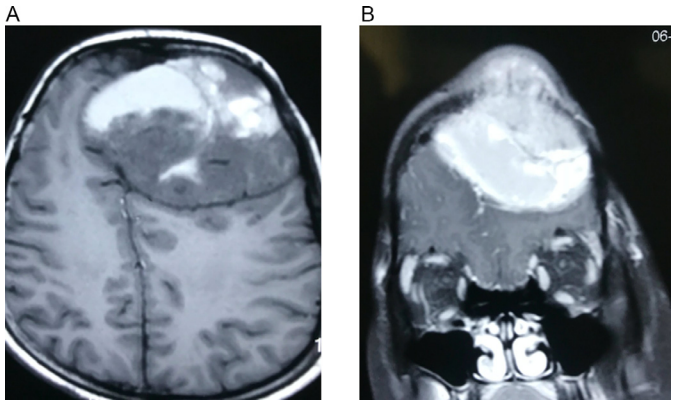

Fig. 2 – (A) T1 weighted axial MRI image showing heterogeneously hyperintense extra-axial lesion in frontal region with mass effect. (B) T1 post contrast coronal image showing heterogeneously enhancing extra-axial mass in frontal region with adjacent bulge.

The patient's headache has resolved, and she is undergoing radiotherapy and is doing well. She has been educated about the prognosis of the disease and is being called for regular follow-up in every 6 month.

Discussion

This is a case of a 16-year-old girl who presented with an insidious headache with progressive change in character. Upon imaging, a mass was found in the frontal region of the brain, subsequently diagnosed through histological analysis as an anaplastic meningioma. Meningioma is predominantly seen in middle-aged or elderly people, with Grade III meningioma typically diagnosed around the median age of 57 years [9]. Childhood meningioma is a rare occurrence, typically detected towards the end of the first or beginning of the second decade of life, as evidenced in our case [10]. Although meningiomas are more prevalent among women (2:1 ratio), anaplastic meningiomas are reported more frequently in men [11–13]. It is by findings from Zhu et al., where among 63 patients diagnosed with anaplastic meningioma, the male-to-female ratio was 1.25:1, with a mean age of 50.4 ± standard deviation (SD) of 14.2 years (range, 20–79 years) [4]. However, we encountered an anaplastic meningioma in a 16-year-old girl.

In younger patients, meningiomas are particularly associated with genetic syndromes such as neurofibromatosis type 2 (NF2) and past exposure to brain radiation [7,13,14]. Other factors that increase the risk of developing these tumors include exposure to certain chemicals like pesticides or herbicides, brain trauma, aging, obesity, and certain medications like cyproterone acetate [2,6,9,11]. Nevertheless, the majority of meningiomas occur spontaneously and their exact cause remains unknown, as seen in our case [6].

Patients diagnosed with anaplastic meningioma usually experience nonspecific symptoms such as headache and vomiting [5]. Additionally, symptoms may include seizures, focal neurological impairments, visual, and auditory disturbances, memory loss, urinary incontinence, confusion, and difficulty swallowing, depending on the tumor's location and its impact on adjacent structures [6]. Furthermore, some individuals are incidentally discovered to have anaplastic meningioma during brain imaging for unrelated symptoms [9].

CT and MRI findings indicative of anaplastic meningioma include heterogeneous contrast enhancement with or without cystic changes, irregular, or nodular tumor contour suggesting infiltration into the brain, significant surrounding edema, necrosis, osteolysis, bone invasion, "mushrooming" of the tumor, and the presence of deep draining veins [4,10,15]. According to a recent review by Servo et al., statistically significant indicators of malignancy include cyst formation, nodular contour, and absence of calcification [16]. Conversely, our case exhibits both cystic and solid components, along with calcification in the solid portions. Ahmeti et al. documented a case where an anaplastic meningioma extensively damaged the calvarial bone, aligning with our findings [17]. Anaplastic meningiomas are more commonly found on the cerebral convexities rather than at the skull base. In our case, the tumor is located in the frontal convexities [7,13].

Anaplastic meningioma can be mistaken for hemangiopericytoma (HPC) due to similar imaging features such as irregular margins, heterogeneous enhancement, significant edema, and skull erosion, as observed in our case. However, HPC tends to have a higher incidence of cystic components and hemorrhage compared to meningiomas, while calcification is rare in HPC. HPCs are highly vascular tumors known for their aggressive nature, characterized by a high rate of local recurrence and a tendency for both neuraxis and extracranial metastases. Similar to anaplastic meningioma, HPC is more prevalent in males. Pediatric cases of HPC account for approximately 10% of reported cases, whereas meningiomas make up around 1.1%-2.9% of intracranial pediatric tumors [18].

Anaplastic meningioma is confirmed by histology when the tissue demonstrates either excessive mitotic activity (>20 mitoses per 10 high-power fields), focal or diffuse loss of meningothelial differentiation at the light microscopic level resulting in sarcoma, carcinoma, or melanoma-like appearance or a predominant papillary or rhabdoid morphology [7]. Histopathological analysis often reveals frequent atypia and

necrosis, loss of architecture, increased cellularity, nuclear pleomorphism, and mitotic activity [4,6].

The standard treatments for anaplastic meningioma are surgical resection and adjuvant radiotherapy [1,3,6,8]. However, the use of radiotherapy in infants and young children remains a topic of debate due to the increased vulnerability of their central nervous system to potential long-term radiation-related complications [14]. In our case, we opted for the standard treatment approach, performing surgical removal of the tumor followed by adjuvant radiotherapy. The use of chemotherapy in grade 3 meningiomas is currently considered experimental and is usually reserved for cases of recurrent grade 3 meningiomas where further surgery or radiotherapy is not feasible [2].

In general, pediatric meningiomas have a poorer prognosis compared to those in adults, with higher recurrence rates and reduced overall survival [14]. Risk factors associated with recurrence include histologic grade, incomplete tumor removal, young age, specific subtypes of meningiomas, brain infiltration, and a high proliferative rate [8]. Despite their aggressive nature, anaplastic meningiomas typically exhibit less metastatic potential [14]. In our case also, no metastasis was detected on the PET scan.

Conclusion

Anaplastic meningioma in pediatric patients poses diagnostic challenges due to its rarity and nonspecific symptoms. Prompt recognition, thorough imaging, and histological confirmation are crucial. Treatment with surgical resection and adjuvant radiotherapy can improve outcomes, necessitating long-term follow-up for recurrence monitoring.

Author contributions

Shailendra Katwal: Conceptualization, as mentor and reviewer for this case report and for data interpretation. **Srijana Katwal:** Contributed in performing literature review and editing. **Sushmita Bhandari:** Contributed in writing the paper and reviewer for this case. **Alisha Adhikari:** Contributed in writing the paper. All authors have read and approved the manuscript.

Registration of research studies

Not applicable.

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Patient 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.

REFERENCES

[1] Cao H, Jiang B, Zhao Y, Fan C. A rare subtype of meningioma: case series of anaplastic meningioma and review of the literature. Medicine (Baltimore) 2018;97(23):e11019.

[2] AlAnsari GA, Bukhari N, Abdulkader MM, Alotain I, Taha MS. Malignant anaplastic meningioma in neurofibromatosis type 1 patient: a rare case report. AME Case Rep 2022;6:36.

[3] Ajler P, Davila EZG, Plou P, Casto F, Christiansen S, Boccalatte LA, et al. Multidisciplinary approach to anaplastic and metastatic meningioma: a case report and review of the literature. Surg Neurol Int 2023;14:230.

[4] Zhu H, Xie Q, Zhou Y, Chen H, Mao Y, Zhong P, et al. Analysis of prognostic factors and treatment of anaplastic meningioma in China. J Clin Neurosci 2015;22(4):690–5.

[5] Intraparenchymal anaplastic meningioma in a child: a rare entity - PMC [Internet]. [cited 2024 May 4]. Available from: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4421947/

[6] Najjar MW, Abdul Halim NH, Sukhon FR, Youssef B, Assi HI. Anaplastic meningioma presenting as a left parietal mass: a case report. Am J Case Rep 2017;18:1166–70.

[7] Buttrick S, Shah AH, Komotar RJ, Ivan ME. Management of atypical and anaplastic meningiomas. Neurosurg Clin N Am 2016;27(2):239–47.

[8] Kessler RA, Garzon-Muvdi T, Yang W, Weingart J, Olivi A, Huang J, et al. Metastatic atypical and anaplastic meningioma: a case series and review of the literature. World Neurosurgery 2017;101:47–56.

[9] Apra C, Peyre M, Kalamarides M. Current treatment options for meningioma. Expert Rev Neurother 2018;18(3):241–9.

[10] Ray C, Nijensohn E, Aduana V, Mcdonald LCT. MRI, and angiographic findings of a highly aggressive malignant meningioma a case report. Clin Imaging 1993;17(1):59–63.

[11] Joselow A, Raugi G, Knezevich SR. Aggressive meningioma presents as innocuous forehead lesion: a case report. Dermatol Online J 2014;20(11):1–5 13030/qt7tm8t9p0.

[12] Yang SY, Park CK, Park SH, Kim DG, Chung YS, Jung HW. Atypical and anaplastic meningiomas: prognostic implications of clinicopathological features. J Neurol, Neurosurg Psychiat 2008;79(5):574–80.

[13] Wilson TA, Huang L, Ramanathan D, Lopez-Gonzalez M, Pillai P, De Los Reyes K, et al. Review of Atypical and Anaplastic Meningiomas: Classification, Molecular Biology, and Management. Front Oncol. 2020;10:565582.

[14] Lucchesi M, Buccoliero AM, Scoccianti S, Guidi M, Farina S, Fonte C, et al. A successful case of an anaplastic meningioma treated with chemotherapy for soft tissue sarcomas. CNS Oncol 2016;5(3):131–6.

[15] Falavigna A, Santos JA, Chimelli L, Ferraz FA, Bonatelli Ad A, de P. Anaplastic meningioma: case report. Arq Neuropsiquiatr 2001;59(4):939–43.
[16] Servo A, Porras M, Jääskeläinen J, Paetau A, Haltia M. Computed tomography and angiography do not reliably discriminate malignant meningiomas from benign ones. Neuroradiology 1990;32(2):94–7.
[17] Ahmeti H, Maslehaty H, Petridis AK, Doukas A, Mahvash M, Barth H, et al. Extensive growth of an anaplastic meningioma. Case Rep Neurolog Med 2013;2013:1–4.
[18] Mama N, Ben Abdallah A, Hasni I, Kadri K, Arifa N, Ladib M, et al. MR imaging of intracranial hemangiopericytomas. J Neuroradiol 2014;41(5):296–306.