Meningiomas are frequent in adults but rare in children, though they are not uncommon. They are known to occur in the pediatric population in all age groups. In children, meningiomas are usually known to be large, cystic, and even aggressive. Among them, skull base meningiomas constitute a distinct entity. Meningiomas arising from the skull base in those younger than the age of two years are rarely reported in literature. We report one such skull base meningioma, involving the middle and posterior cranial fossa, in a child of one year and eight months. The challenges associated in its diagnosis and management are presented.

**Keywords:** Children younger than 2 years, skull base meningioma, surgical strategy

A one year and eight-month-old girl child was brought to our service with a history of imbalance in walking since two months. Till then, the child had apparently normal physical and mental milestones. The mother had noticed a slight difference in the child’s walking, which progressively became noticeable in a month’s time. The child also started having a preference of the left hand over the right. The altered gait became more obvious by the time they sought our consultation. The child never had any evidence of headache or vomiting or other related symptoms. The child was pleasant, cooperative, spoke simple sentences, and followed commands. On careful examination, the child had mild weakness of the right upper limb associated with gait imbalance. There were no neurocutaneous markers.

Imaging performed prior to our consult revealed a large, well-circumscribed, and lobulated hyperdense mass involving the middle fossa extending to the para sellar, petroclival and to the posterior fossa. The tumor had a wide attachment to the basal dura and to the tentorial edge. The petrous bone had shown hyperostosis with truncation of the apex. The tumor was extending to the posterior fossa through the tentorial hiatus up to the level of the internal acoustic meatus, compressing the brainstem and the cerebellum. The imaging characteristics were suggestive of a meningioma [Figure 1]. However, schwannoma of the fifth cranial nerve, sarcoma, and Eving’s tumor were also considered as differential diagnosis. Biopsy and the role of angiography were discussed in the plan of management. However, a well-circumscribed lesion, with no bony erosion or evidence of parenchymal infiltration suggested a meningioma. Angiogram and biopsy were deferred due to the requirement of additional anesthesia and the associated risks. A decision was made to go ahead with the definitive surgery. However, it was planned to stage the procedure to minimize the surgery, anesthesia time, and blood loss. Accordingly, a large hockey stick like incision to access both middle and posterior fossa was planned. First, the middle fossa tumor was approached: The tumor was firm, moderately vascular and could be decompressed with an ultrasonic aspirator. During surgery, it typically looked like a meningioma with an extensive attachment to the base, a tentorial edge, and the petrous dura. The middle fossa tumor was completely removed up to the point of the petrous apex. Then, we decided to stop and to come back after a week for the rest of the tumor. A week later, the posterior fossa was exposed through a classical CP angle approach. The tumor was detached from the petroclival dura and the tentorium;
the tumor was debulked till we accessed the previous surgical cavity in the mid fossa through the tentorial hiatus. Gross total resection of the tumor was achieved. Staging had helped us to minimize the anesthesia time and blood loss.

The histological examination of the tumor proved it to be a meningothelial meningioma. There was no evidence of mitosis. The proliferative index was less than 1%, hence it was classified as WHO Grade 1. Postoperatively, the child recovered completely. The hemiparesis improved, and the child was discharged.

Postoperative imaging confirmed gross total resection [Figure 2]. The child was clinically normal without any neurological deficits. An MRI repeated two years later showed no evidence of any residual tumor. The child continued to be normal in her clinical status as well as during the milestones of development.

**Discussion**

Meningiomas constitute 20–30% of brain tumors in adults.[1,2] However, in the pediatric population, they are uncommon, contributing to less than 5% of all pediatric brain tumors and less than 2% of meningiomas.[3–5] An Indian publication described 0.4% to 4.6% of incidence of meningiomas among all brain tumors in people younger than the age of 16. This accounts for 0.9 to 3.1% of intracranial meningiomas.[6] They are known to be atypical, malignant, and large in size and they are more likely to be associated with genetic disorders such as neurofibromatosis type 2, Gorlin syndrome, or Rubinstein-Taybi syndrome.[7,8] There are very few reviews published for meningiomas in children. In a review of 22 studies, including 582 meningiomas in children, the skull base was involved in only 126, constituting 21.6%. The distribution in the skull base showed sphenoid in 5.9%, frontal and mid fossa in 5.1%, and foramen magnum in 2%.[9] Similarly, another meta-analysis reported a 29% incidence of skull base meningiomas.[10] The other common locations are convexity, parasagittal, intraventricular, and posterior fossa. Brockmeyer emphasized the fact of the rarity of these tumors in children, constituting 8.3% only. A recently published meta-analysis with characteristics of 700 pediatric meningiomas from 35 studies reported a higher incidence of non-skull base (73%) meningiomas compared with that of skull base ones (27%).[10,11] The incidence of meningiomas in children increases with age and are more common during the second decade than the first.[9,12]

In many studies, the average age of diagnosis is reported as 13.7 years.[1,2,4,9,13] Our patient was only one year eight months at the time of diagnosis. Meningiomas in childhood can occur anywhere in the
cranial, but they are exceptionally common in the infratentorial, intraventricular regions and at the skull base.[4,14] Among them, tumors involving the skull base are quite rare. We have not come across a meningioma involving the middle and posterior fossa skull base, such as a dumbbell involving two compartments in the literature published. The female preponderance observed in adults is not seen in children. The size of the meningiomas in children can be quite large in comparison to their counterparts in adults.[15] In our case also, the tumor was significantly large considering the age of the child. The ability of the pediatric skull and brain to compensate and to accommodate in children may be responsible for such mammoth sizes of tumors at the time of presentation.[13,16,17] The symptoms can be nonspecific and often may not correlate to the location involved. Once the tumor reaches a sufficient size, further on the symptoms can become rapid. This can explain the short duration of symptoms often noted among the published series as well as in our case. These tumors are known to arise from the dura, as well as purely from the leptomeninges without a significant dural attachment. In our case, the tumor had a clear attachment to the dura as well as to the tentorium. Enlargement of the skull, cranial nerve deficits, and local compressive symptoms are the other known presentations among the younger age groups. The other most common association reported with meningiomas is neurofibromatosis. However, our child did not have any such association or family history.

In general, meningiomas in infancy are exceptionally rare.[7,11,18-23] An extensive search of literature has revealed only three such cases, one each reported by Kotecha et al.[11] and Rushing et al.[7] In a data analysis reported by Kotecha et al.[11], there was a collection of 31 meningiomas, including all varieties, in the population younger than three years of age. In their analysis, the prognosis depended on achieving the gross total resection, histological grading, and associated conditions such as neurofibromatosis or other syndromes. The same authors also mentioned poorer outcomes of these tumors in the population younger than the age of three in comparison to those who were 3–12 years. The reasons attributed were the possibility of the tumors being congenital in nature, with an aggressive biological behavior and associated surgical morbidity.[24] In all the earlier mentioned series, the anterior and middle cranial skull base is reported independently but the combination of middle and posterior fossa is not mentioned. Gross total resection is the best option to offer cure and to avoid recurrence. The aggressive nature of pediatric meningiomas is well known in comparison to their adult counterparts, as largely described in literature.[22,23] The WHO classified meningiomas as grade 1 (benign), grade 2 (atypical), and grade 3 (anaplastic/malignant).[2,25] The histopathological features in our child were suggestive of WHO grade 1, which is benign in nature. Such tumors are known to have better outcomes in comparison to the higher grade tumors.[24] However, all meningiomas in children warrant a long-term follow-up and surveillance to evaluate their progression-free interval as well as recurrence. No definite protocol for imaging is forthcoming in the literature. The consensus seems to be to perform imaging every year till five years and then perhaps at longer intervals. The higher grade tumors possibly need more frequent imaging. In benign tumors, if there is residual tumor in the postoperative imaging it is worthwhile to reexplore to get the gross total resection.

Surgery in children younger than two years has its own challenges, such as prolonged anesthesia, hypothermia, blood loss, fluid and electrolyte disturbances, and problems related to blood transfusion. Hence, we preferred elective staging of the surgery to enhance the safety and tolerance. Accordingly, the mid fossa tumor first and later the posterior fossa one gave us satisfactory results. Our case is unique in many ways. Though the

*Figure 3: Postop MRI T2 sequence with gross total resection*
diagnosis was made at one year eight months, we feel the tumor must have started much earlier and possibly in infancy. The presentation, location, involvement of the middle and posterior fossa with the truncated petrous apex, and being surrounded by critical structures pose challenges in the diagnosis and surgical strategy. We could achieve gross total resection without any morbidity, which should contribute to the duration of a progression-free interval. Two-year follow-up had shown normal physical and mental development, with no evidence of residue or recurrence on the MR imaging [Figure 3]. However, a long-term follow-up and serial imaging is necessary to determine the long-term outcome.

Although pediatric meningiomas are quite rare, they constitute a definite entity among children and should be considered in the differential diagnosis wherever the imaging characteristics are suggestive. The surgical strategy should be planned with a goal to achieve gross total resection, including attachments, wherever possible.

**CONCLUSION**

Pediatric skull base meningiomas form a small subset of pediatric intracranial tumors. Double compartmental meningioma involving the middle and posterior cranial fossa in the age group younger than two years of age is exceedingly rare. We report one such child with benign skull base meningioma successfully removed by staging the surgery. Gross total resection and histological pattern will decide the long-term outcomes of meningiomas in children. Long-term follow-up is mandatory despite the gross total resection.

**Financial support and sponsorship**

Nil.

**Conflicts of interest**

There are no conflicts of interest.

**REFERENCES**

1. Peng J, Liang ZG, Li KC. Intracranial malignant meningioma with cerebrospinal fluid dissemination: a case report. Chin Med J (Engl) 2011;124:1597-9.

2. Jaiswal S, Vij M, Mehrotra A, Jaiswal AK, Srivastava AK, Behari S. A clinicopathological and neuroradiological study of paediatric meningioma from a single centre. J Clin Neurosci 2011;18:1084-9.

3. Auguste Ki, Rutka JT. Meningiomas in children. In: DeMonte F, McDermott MW, Al-Mefty O, editors. Al-Mefty's Meningiomas. 2nd ed. New York, NY: Thieme; 2011:69-74.

4. Greene S, Nair N, Ojemann JG, Ellenbogen RG, Avellino AM. Meningiomas in children. Pediatr Neurosurg 2008;44:9-13.

5. Tufan K, Dogulu F, Kurt G, Emmez H, Ceviker N, Baykaner MK. Intracranial meningiomas of childhood and adolescence. Pediatr Neurosurg 2005;41:1-7.

6. Sanat Bhagwati, Nirav Mehta, Geeta Parulekar. Meningiomas in children: a study of 18 cases. J Pediatr Neurosurg 2009;4:61-5.

7. Rushing EI, Olsen C, Mena H, Rueda ME, Lee YS, Keating RF, et al. Central nervous system meningiomas in the first two decades of life: a clinicopathological analysis of 87 patients. J Neurosurg 2005;103:489-95.

8. Morrison AL, Rushing E. Pathology of meningiomas. In: DeMonte F, McDermott MW, Al-Mefty O, editors. Al-Mefty's Meningiomas. 2nd ed. New York, NY: Thieme; 2011. pp. 40-8.

9. Menon G, Nair S, Sudhir J, Rao BR, Mathew A, Bahuleyan B. Childhood and adolescent meningiomas: a report of 38 cases and review of literature. Acta Neurochir (Wien) 2009;151:239-44; discussion 244.

10. Jan – Karl Burkhardt, Marian C. Neidert, Michael A. Grotzer, Niklaus Krayenbuhl, Oliver Bozinov. Surgical resection of pediatric skull base meningiomas. Childs Nerv Syst 2013;29:83-7.

11. Kotecha RS, Pascoe EM, Rushing EJ, Rorke-Adams LB, Zwerdling T, Gao X, et al. Meningiomas in children and adolescents: a meta-analysis of individual patient data. Lancet Oncol 2011;12:1299-39.

12. Li Z, Li H, Wang S, Zhao J, Cao Y. Pediatric skull base meningiomas: clinical features and surgical outcomes. J Child Neurol 2016;31:1523-7.

13. Im SH, Wang KC, Kim SK, Oh CW, Kim DG, Hong SK, et al. Childhood meningioma: unusual location, atypical radiological findings, and favorable treatment outcome. Childs Nerv Syst 2001;17:656-62.

14. Martínez-Lage JF, Poza M, Alcaraz J, Molina E. Giant meningioma of the III ventricle in a child: case report and review of the literature. Childs Nerv Syst 1993;9:306-8.

15. Turgut M, Ozcan OE, Bertan V. Meningiomas in childhood and adolescence: a report of 13 cases and review of the literature. Br J Neurosurg 1997;11:501-7.

16. Liu Y, Li F, Zhu S, Liu M, Wu C. Clinical features and treatment of meningiomas in children: report of 12 cases and literature review. Pediatr Neurosurg 2008;44:112-7.

17. Arivazhagan A, Devi BI, Kolluri SV, Abraham RG, Sampath S, Chandramouli BA. Pediatric intracranial meningiomas—do they differ from their counterparts in adults? Pediatr Neurosurg 2008;44:43-8.

18. Grossbach AJ, Mahaney KB, Menezes AH. Pediatric meningiomas: 65-year experience at a single institution - J Neurosurg Pediatr 2017;20:42-50.

19. Caroli E, Russillo M, Ferrante L. Intracranial meningiomas in children: report of 27 new cases and critical analysis of 440 cases reported in the literature. J Child Neurol 2006;21:31-6.

20. Ferrante L, Acqui M, Artico M, Mastronardi L, Rocchi G, Fortuna A. Cerebral meningiomas in children. Childs Nerv Syst 1989;5:83-6.

21. Kolluri VR, Reddy DR, Reddy PK, Naidu MR, Rao SB, Sumathi C. Meningiomas in childhood. Childs Nerv Syst 1989;5:307-31.

22. Kotecha RS, Juncerstorff RL, Lee S, Cole CH, Gottardo NG. Pediatric meningioma: current approaches and future direction. J Neurooncol 2011;104:1-10.

23. Thuijs NB, Uitdehaag BMJ, Van Owerkerk WJR, van der Valk P, Vandertop WP, Peerdeman SM. Pediatric meningio- mas in the Netherlands 1974–2010: a descriptive epidemiological case study. Childs Nerv Syst 2012;28:1009-15.

24. Rochat P, Johannesen HH, Gjerris F. Long-term follow-up of children with meningiomas in Denmark: 1933 to 1984. J Neurosurg 2004;100:179-82.

25. Maranhão-Filho P, Campos JC, Lima MA. Intracranial meningiomas in children: ten-year experience. Pediatr Neurol 2008;39:415-7.