Scholars Academic Journal of Biosciences (SAJB) **ISSN 2321-6883 (Online)**
Sch. Acad. J. Biosci., 2015; 3(2A):170-172 **ISSN 2347-9515 (Print)**

(An International Publisher for Academic and Scientific Resources)
www.saspublishers.com **DOI :** 10.36347/sajb.2015.v03i02.010

Case Report

Anterior Cranial Fossa Based Meningioma in a 5 Year Old Boy: A Case Report

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Abstract: Meningioma is an apparently benign brain tumour originating from the arachnoid cells lining the meninges or the choroid plexuses. Approximately 1% to 4% of primary intracranial tumours in the paediatric group is meningioma and is increasing in incidence with age. Commonly there is variation in site of involvement, symptoms, histological picture and prognosis in relation to adult counterparts. In our report a 5 year old male patient presented with only one episode of generalized tonic clonic seizure (GTCS) followed by unconsciousness. The computed tomography (CT) scan report of brain showed a dural based large mass occupying the anterior cranial fossa. Total removal of the tumour was done carefully. The biopsy report revealed atypical variety of meningioma. The post operative course was uneventful. There is high chance of recurrence for atypical variety of meningioma. Though, no chemotherapy or radiotherapy was given for atypia due to non co operation of the patient. Despite that no recurrence is found in a follow-up examination even after 2 years. We want to highlight this case due to rare incidence, rare location and good prognosis of this tumour despite atypia, possibly due to total excision of the mass carefully.
Keywords: Childhood meningioma, Meningioma, Paediatric neoplasm

INTRODUCTION

Meningioma is the most common benign tumour of meninges covering the brain parenchyma; commonly seen in the fifth decade of life, occupying approximately 15-20% of primary intracranial tumours [1]. Intracranial meningiomas in children and adolescents are rare tumours in relation to adult counterpart. In most large series, the incidence of meningioma before the age of 16 years ranges from 0.4 to 4.6% of all primary brain tumours in this age group [2]. The female preponderance found in adult patients is not seen in children, the reported male to female ratio in children being 1.2:1 [1]. A few known risk factors are responsible for the development of meningioma, as for example neurofibromatosis type two (NF-2), radiation therapy etc [3]. Prognostically the childhood meningiomas have been considered by some to be more aggressive than their adult counterparts [4].

CASE REPORT

This case was diagnosed at Nil Ratan Sir Car Medical College in Kolkata. We report here a case of 5 year old, moderately built and well nourished male, student by occupation presented to medicine outpatient department with complaints of just one episode of GTCS followed by unconsciousness for 5 minutes. She had been delivered normally and was healthy otherwise. There was no history of trauma in head but there was a history of excessive talkativeness and poor school performance. No neurocutaneous marker was found. Vitals were stable. His muscle bulk and tone was normal. Power was 5/5 in all groups. Sensory reflexes, deep tendon reflexes, cerebeller signs and gait was within normal limits but plantar reflexes were down going. All the primary reports such as complete haemogram, biochemical parameters (hepato renal function, sodium and potassium), chest X ray, coagulation profile, urine and stool were unremarkable. Preoperative CT scan of brain revealed a well defined heterogenous lesion of approximately 10cm×7 cm×6 cm in size,dural based likely extra-axial in location at right frontal region which showed significant contrast enhancement with extension to left frontal region with evidence of compression over anterior horn of right lateral ventricle and dilatation of contra lateral ventricle. The midline septum was deviated to left side by 2 cm. Corpus callosum and brain stem was compressed from right side. CT report was suggestive of meningioma with mass effect. Patient underwent bifrontal craniotomy and gross total excision of the mass. We received four grayish brown soft tissue masses together measuring 6cm×5cm ×4 cm.

Histopathology report showed numerous meningothelial cells in whorling pattern, few of them were arranged in patternless sheets along with foci of small cells sowing condensed nuclei. Mitotic activity was 4-5 /10 high power field (HPF) along with early foci of necrosis. Stromal hyalinization and psammomatous calcification also present. The post operative course was uneventful. No chemo radiotherapy was given to the patient for atypia due to non co-operation. There was no recurrence of tumor in a follow-up examination after 2 years.

DISCUSSION

Meningioma is usually attached to the dura and thought to arise from the arachnoid cap or meningothelial cells. It is reported that the frequency is less than 5% of all paediatric brain tumours [3]. Its incidence as reported by Mendiratta *et al* [2] is 1.5% of total meningiomas seen in the population. Children with meningiomas commonly present late in the first decade or early in the second decade of life [5]. In our case the patient is 5 years of age. Meningioma generally differs in various clinical and biological aspects from meningioma in the adult population [6]. Risk factors for meningioma include: radiation treatment, female hormones and inherited nervous system disorders. In contrast to adult meningioma where a female preponderance is seen [7], childhood meningioma shows a distinct male predominance [5]. Our patient is also a male. The greater occurrence of meningioma in males could be related to an absence of the effect of sex hormones on corticosteroid receptors in meningioma cells for low blood concentrations [8].This suggests that different pathogenic factors might account for the occurrence of meningioma in children and adults. Some studies on genetic aberrations in meningioma in children show no differences from meningioma in adults [9]. Signs and symptoms related to raised intracranial pressure following obstruction of the cerebrospinal fluid circulation are most common in childhood meningioma due to the involvement of ventricles (12%) [9] as compared to 0.5-4.5% in adult [10]. Though ventricle was not the site of involvement in our case and the patient presented with only GTCS without any evidence of raised intracranial pressure. No feature of neurofibromatosis was found in this case. In different series in the literature, 0-41% of childhood meningioma is associated with neurofibromatosis [5].

Meningioma shows characteristic imaging features that include broad-based dural attachment, signal changes in the skull due to tumor infiltration, sharp demarcation between the tumor and the brain, mass effect on adjacent brain tissue and homogeneous enhancement of a contrast agent [11]. The site of origin provides a clear diagnosis in most cases, which often facilitate their diagnosis without the need of invasive diagnostic procedures. We faced no microscopical diagnostic difficulties; rather we were very much concerned about the site, size as well as prognosis of that tumour.

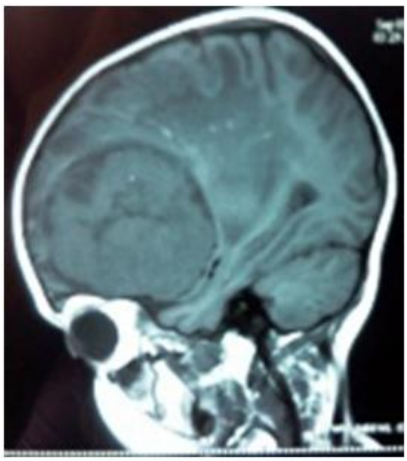

Fig. 1: MRI of brain without contrast: sagital view showing a large well circumscribed dural based meningioma occupying the anterior cranial fossa

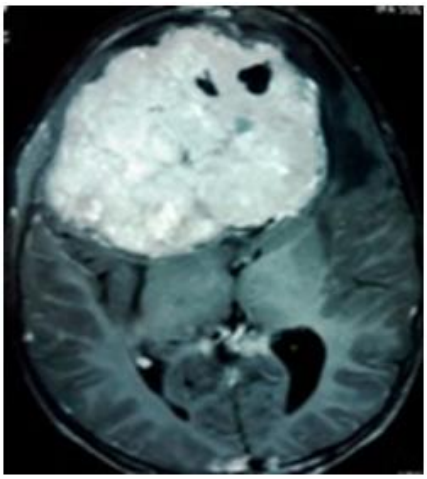

Fig. 2: MRI of brain with contrast: coronal view showing intensely enhancing meningioma

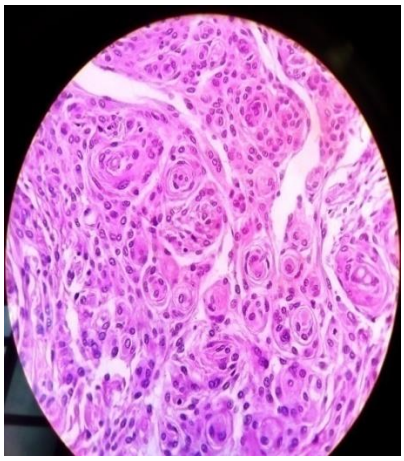

Fig. 3: Microscopy: Sheets of meningothelial cells in whorling pattern, low power (Haematoxylin and Eosin stain, H & E, 10 X)

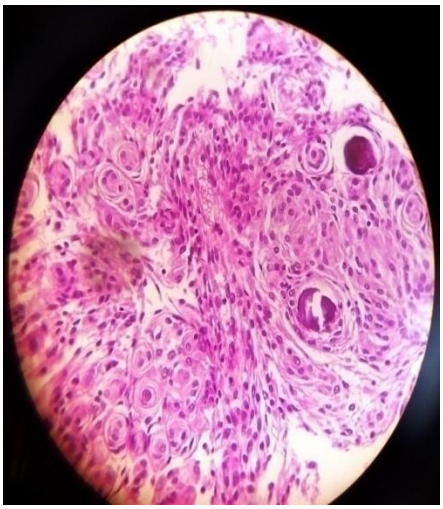

Fig. 4: Microscopy: Sheets of meningothelial cells in whorling pattern with focal areas of psammomatous calcification, low power (H & E stain, 10 X)

The incidence of cystic changes is commonly seen in meningioma of children [12]. In our case no cystic component was found.

Immunohistochemically this tumour is glial fibrillary acidic protein (GFAP) negative and epithelial membrane antigen (EMA) and vimentin positive [13]. Surgical excision has been the treatment of choice for this tumour. Surgical management poses a formidable challenge considering their peculiar location, larger size at presentation and relatively less blood volume in children. We got atypical variety. Overall, most series have shown a high incidence of atypical and anaplastic meningiomas in children as compared to the adult population [6]. In the literature, adjuvant therapy has been advised in these patients in the form of radiation therapy [6]. Though, no chemotherapy or radiotherapy was given to patient despite the atypia due to non cooperation of the patient. Literature has shown that in different series recurrence rate of approximately 13% [9]. Recurrence seems to be strictly related to incomplete resection and/or histological subtype of the meningioma. Atypical, aggressive and meningiomas with cortical invasion show a higher rate of recurrence. This patient was followed up closely and had not shown any recurrence even after 2 years, might be due to total removal of the mass by the surgeons carefully.

CONCLUSION

Through this report we want to highlight the rarity of the childhood meningioma and some characteristic differences when compared with their adult counterparts. These include slight preponderance in male subjects, higher incidence of intraventricular and skull base location and frequent cystic changes, though these last three were not found in our case. Total surgical excision should be performed wherever feasible due to higher incidence of atypical and aggressive histological subtypes in the paediatric population. Children with complete resection of meningioma have a good prognosis.

ACKNOWLEDGEMENT

The author is grateful to Mr. Animesh Hazra, Assistant Professor, Department of Computer Science & Engineering, Jalpaiguri Government Engineering College, Jalpaiguri, West Bengal, India, for the provision of valuable technical and mental support.

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