Case Report

Parasagittal Ependymoma Mimicking Falcine Meningioma

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Abstract

This is a case of a parasagittal ependymoma, radiographic resemblance to falcine meningioma.This is the first case operated at our hospital with radiographic appearance of falcine meningioma but histopathological examination revealed features of an ependymoma.

A five year old female child presented with two months history of seizures. Clinical examination and neuroimaging studies revealed a contrast enhancing calcified lesion in the right parietal lobe. Complete resection of the lesion was performed without complication and neurological deficit. The final immunohistochemical findings were consistent with those of an ependymoma.

Extraaxial ependymomas rarely occur in the supratentorial parasagittal region and may pose a difficult diagnostic dilemma because of their radiographic and gross appearance, as seen in our present case. Because of this difficulty, histopathological examination as well as immunohistochemical confirmation are required to make a definitive dignosis. As these lesions have a propensity to recur, they have to be managed by a complete surgical resection followed by post-operative radiation therapy for residual tumour. Although its use has not been validated by controlled studies, radiation therapy generally is used for children older than 3 years.

Key words: Brain neoplasm, Ependymoma, Falcine meningioma.

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Introduction

Ependymomas are infrequently seen brain neoplasms that have been defined as neoplasms arising from ependymal cells lining the ventricles and the central canal of the spinal cord. Cerebral ependymomas are uncommon tumours with a peak incidence in early childhood. The four most prevalent locations are supra and infratentorial, spinal and conus-cauda-filum. Ependymomas represent 1.2% to 7.8% of brain tumours. Ependymomas presumably arise from ependymal cells or rests, so they should occur only in relation to the ventricular surface. In fact, although most do arise adjacent to a ventricle, they may occur in the supratentorial parenchyma, in the filum terminale or within the substance of the spinal cord. Ependymomas may develop outside the nervous system in the ovaries, soft tissue and mediastinum.These extraneural ependymomas may arise through various mechanisms, including metastases or direct extensions following surgical excision; direct extensions to the soft tissues from a primary ependymoma of the lower spinal cord,cauda equine or filum terminale.

Despite their relatively benign histological appearance, they recur relentlessly and are among the most difficult neoplasms to cure. If the tumour is accessible, it is managed by gross total resection, followed by magnetic resonance imaging to monitor residual tumour.

Primary extraaxial ependymomas, however, very rarely occur. Search of literature revealed five such cases. We present a case of extraaxial parasagittal falcine based ependymoma without parenchymal involvement which was radiologically identical to a falcine meningioma.

Case report

The patient, a five year- old girl daughter of a serving soldier presented with a two months history of partial seizures involving left upper and left lower extremities. She was referred to our hospital after initial management with anticonvulsant drug and investigation with CT scan head and EEG in another institution. At presentation the patient denied gait abnormality, headache, vomiting, dizziness or diplopia. Before the onset of seizures the patient seemed healthy and her medical history was not significant, with no prior surgeries and no known drug allergy.

On physical examination, the patient was alert and oriented, in no distress and without papilloedema.Cranial nerves and motor and sensory examination did not show focal deficit, with the exception of mild hyperreflexia of the left upper and left lower extremities compared with the right side. The patient's plantar reflexes were downgoing, and there were no other long-tract signs.

A Computerized Tomography scan head revealed a 3.5 cm X 3 cm. hyperdense, calcified mass in the right parasagittal region of the parietal lobe adjacent to the precentral gyrus .Gadolinium contrast enhanced MRI was requested for defining the detailed anatomy of the motor cortex. MRI revealed a 3.5cm X 3cm X 3cm.right parafalcine mass adjacent to the precentral gyrus.The lesion appeared hypointense to isointense with grey matter on T1-weighted images, hyperintense on T2-weighted images with contrast enhancement. The preoperative diagnosis of falcine meningioma was made and planned for surgery.

Fig. 1, Preoperative CT scan head.

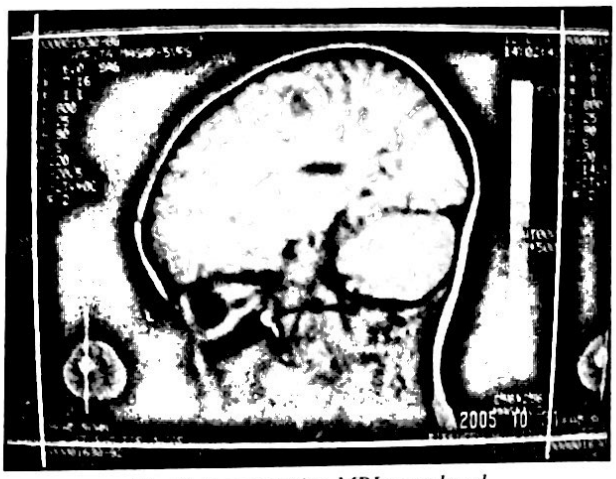

Fig. 2, Preoperative MRI scan head

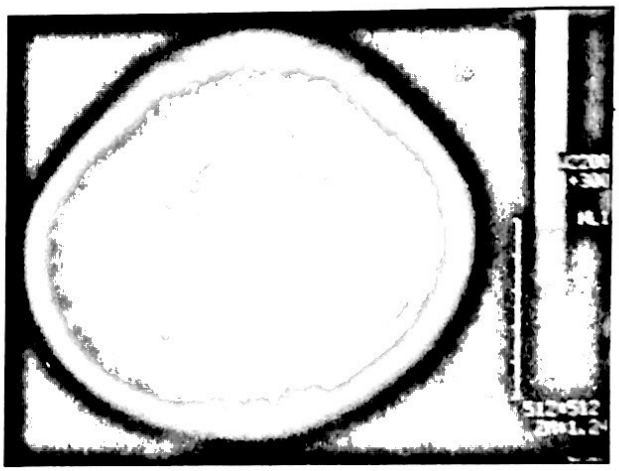

Fig. 3, Tumour calcification seen in bone window.

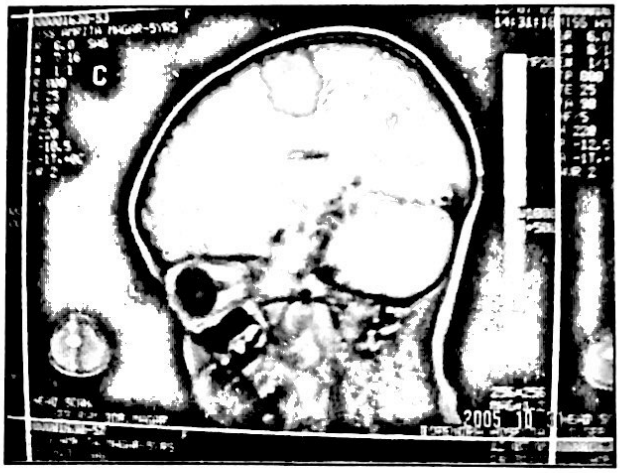

Fig. 4, Preoperative contrast enhanced MRI brain.

Operation

The patient was admitted to the hospital one week prior to surgery and was put on steroid therapy and anticonvulsant therapy. Under general anesthesia a right-sided, parietal craniotomy was performed via a horseshoe-shaped incision crossing the midline.

tumour was removed piecemeal using rongeurs. Exenteration of the tumour was done and the attachment to the falx was coagulated, divided and the tumour excised completely. No parenchymal invasion was noted. The haemostasis was secured by bipolar diathermy and dura was closed with 4/0 vicryl. The bone flap was placed back and the wound was closed in layers with subgaleal suction drain.

Fig. 5, Operation photograph. Arrow indicates tumour.

Fig. 6, A part of resected tumour.

The postoperative period was uneventful and no neurological deficit was noted. Steroid therapy was gradually tapered in two weeks and patient was discharged from the hospital without histopathological report of the resected tumour. The pathological examination result was obtained only after one and half months because a part of the specimen was sent to India for immunohistochemistry.

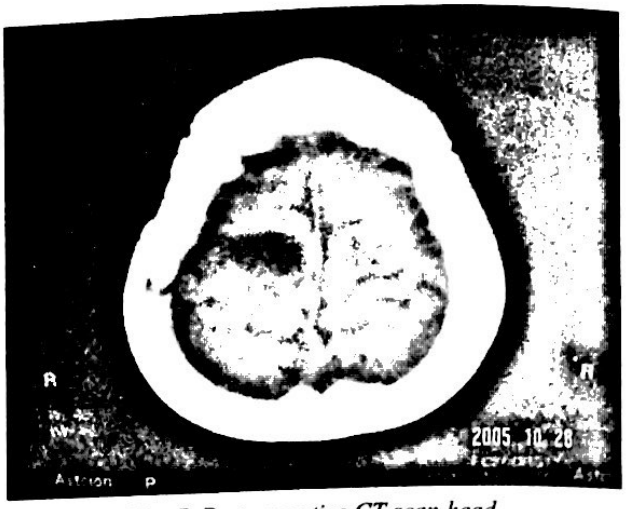

Fig. 7, Post operative CT scan head.

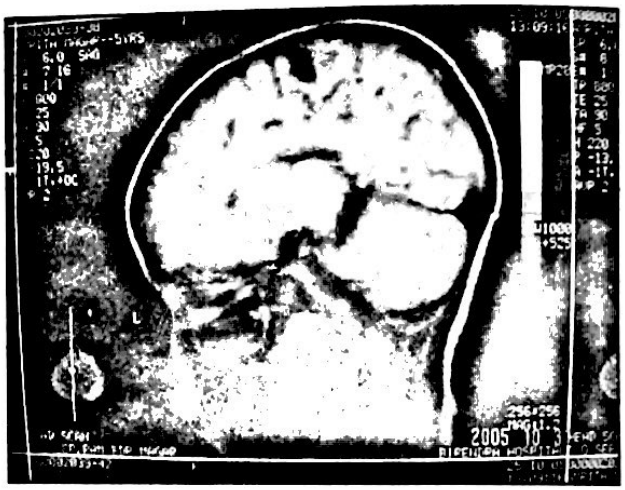

Fig. 8, Post operative MRI scan.

Pathological examination

Microsections show tumour fragments composed of uniform ovoid to elongated cells in a diffuse fibrillary background with perivascular pseudorosettes and extensive calcification. No mitosis, necrosis, or atypia seen. Impression: Ependymoma (WHO grade II).

Immunohistochemistry: Epithelial Membrane Antigen (EMA): Negative.

Glial Fibrillary Acidic Protein (GFAP): Strong diffuse staining in tumour cells and fibrillary processes.

Fig. 9, Histopathological section. (H&E)

Discussion

Cerebral ependymomas are uncommon tumours with a peak incidence in early childhood. They were first classified as a distinct entity in 1926 by Bailey and Cushing, and in the past, they were considered favourable tumours.Depite their relatively benign histological appearance, they recur relentlessly and are among the most difficult neoplasms to cure. Ependymomas can be considered rare because they represent about 6% of all gliomas.More than 90% of childhood ependymomas arise within the cranium; two thirds occur below the tentorium, and one third above it. In contrast, in adults, more than 75% occur within the spinal canal, and majority of intracranial ependymomas arise within the supratentorial compartment.

Ependymomas presumably arise from ependymal cells or rests, so they should occur only in relation to the ventricular surface. In fact, although most do arise adjacent to a ventricle, they may occur in the supratentorial parenchyma, in the filum terminale or within the substance of the spinal cord. Ependymomas may develop outside the nervous system in the ovaries, soft tissues and medistinum.

Extraaxial location of supratentorial ependymoma is extremely rare. In literature review only five cases of extraaxial ependymomas were found. We had an opportunity to operate on an extraaxial, supratentorial ependymoma without parenchymal extension.

No definitive mechanism for the development of these extraaxial ependymomas has been postulated. One hypothesis involves the extension of subcortical, subependymal rests extraaxially with the subsequent development of tumour. Necrosis and calcification of the originating subependymal rests would then follow, leaving a predominately extraaxial ependymoma. Heterotopic placement of ependymal cell rests during fetal development with subsequent growth of tumour was also offered as a possible mechanism. In our case the lack of parenchymal involvement and falcine base of the tumour suggests a heterotopic placement of ependymal cell rests in the falx as the origin of tumour.

The cause of ependymomas is unknown. No environmental factor has been implicated and recent work has concentrated on cytogenetic abnormalities. There have been several reports of loss of material from chromosome 22 in the tumour tissue. Mutations in p53 tumour suppressor gene, which are found in many human cancers, do not appear to play a role in ependymomas.

Presentation is determined by the location of the tumour within the central nervous system. In children, most originate within the posterior fossa and typically present with the "midline posterior fossa syndrome" of headache, vomiting and lethargy. Because all these tumours may arise within the fourth ventricle, obstructive hydrocephalus occurs early and is usually the cause of early symptoms.

Atypical presentations are not unusual. Supratentorial ependymomas may present with signs and symptoms of generalized intracranial hypertension, seizures or neurological symptoms referable to the area of brain involvement. If tumour progression is permitted to continue, pressure waves, opisthotonos, bradycardia, apnea and death ensue. In our case the presenting symptoms was seizures.

Radiographycally, extraaxial ependymomas can be difficult to differentiate from other dura-based lesions. On computed tomography scans, ependymomas are isodense to cerebral cortex. Calcification and cystic components are frequent.Haemorrhage is reported in up to 10% of cases. Low density necrotic areas are also seen. MRI allows multiplanar imaging as well as better tissue differentiation compared with computed tomography. The solid portion appears hypointense to isointense with gray matter on T1-weighted images, hyperintense on proton-density weighting, and isointense to hyperintense on T2-weighted images. There is usually nonhomogenous enhancement with intravenous contrast material.

Grossly, most ependymomas are greyish or red, lobulated, gritty and firm. They are relatively well circumscribed.Supratentorial tumours are often periventricular and may have a large lobulated portion within the ventricle with a subcortical extension.

Microscopically, the degree of cellularity is variable even within a single tumour.The cells are typically polygonal and form a uniform background. A diagnostic feature is the presence of rosettes, representing the tumours' attempt to recapitulate an ependyma-lined central canal. Most only contain perivascular pseudorosettes,in which a blood vessel is surrounded by an eosinophilic halo composed of radiating processes of the cells. Common to ependymomas are vascular hyalinization and microcalcifications and some may even exhibit metaplastic bone or cartilage.

Specific immunohistochemical stains for intermediate filament proteins are typically positive for glial fibrillary acidic protein and vimentin and may be positive for neuron-specific enolase and S-100. Ependymomas are typically negative for alpha-fetoprotein, carcinoembryonic antigen and desmin. Extraaxial ependymomas are so rare that they may be overlooked during pathological differential dignosis.They often exhibit unusual structural features. For these reasons, immunohistochemical evaluation is very useful in their identification.

Glial fibrillary acidic protein is a critical marker. Positivity eliminates major alternative possibility of meningioma and highlights glial processes in perivascular rosettes.

There is considerable variation in the way ependymomas are graded. Several systems have been used in the past, but the current system is that of the World Health Organization which includes the designation "ependymomas" and "anaplastic ependymomas." Ependymoma (WHO grade II) and anaplastic ependymoma (WHO grade III) are neoplasms of children and young adults that originate from the ependymal lining of the cerebral ventricles or the spinal canal. Myxopapillary ependymoma (WHO grade I) is a distinct low grade variant of ependymoma that arises almost exclusively from the caudal portion of the spinal cord of adults in the conus medullaris-filum terminale region. The management of intracranial ependymomas nearly always begins with surgery with the goal being complete resection. Radical surgery alone may be sufficient for infants and adults with low grade tumours. Unfortunately, aggressive efforts at local control, and surgery in eloquent areas, can lead to significant morbidity.

Radiotherapy has been used routinely in adults and older children to eliminate residual tumour, although its use has not been validated by controlled studies. Children younger than three years do not receive radiation and generally have a more adverse outcome than do older children who may receive radiation. For adults, total resection followed by craniospinal radiotherapy has been described as the gold standard.

There is no convincing evidence that adjuvant chemotherapy improves survival when added to standard surgery and radiation therapy for newly diagnosed ependymomas.

Various prognostic factors have been presented in the literature, including age, tumour location, histology and the extent of resection. Most studies show increased survival with gross total resection. Relapse-free survival is the best indicator of long-term success in the treatment of ependymomas, because patients who develop recurrent disease after surgery and radiotherapy are rarely cured. The advent of radiation therapy and improved surgical techniques resulted in improved survival, and contemporary series consistently show five year progression-free survival rates of about 40%.

The current management of childhood ependymoma begins with an attempt at maximal surgical resection, followed by staging MRI to assess residual tumour. Field radiation of involved area is offered in most instances, except in case of infants who receive postoperative chemotherapy. Chemotherapy is considered if there is residual disease. Supratentorial ependymomas in children under the age of 10 years have a recurrence rate of 58%; new tumour growth can be delayed by post operative irradiation.

Conclusion

Ependymomas are tumours of the central nervous system that derive from the ependymal cells lining the cerebral ventricles and the central canal of the spinal cord and from ependymal rests in cortical white matter.

The exact origin of supratentorial extraaxial ependymomas is unclear, although it is believed that these tumours derive from embryonal cell rests. There is a possibility of overlooking these extraaxial supratentorial ependymomas only on grounds of clinical presentation, radioimaging findings and

histological examination if immunohistochemical evaluation is not performed. Immunohistochemistry plays a crucial role in establishing the diagnosis of these tumours. The gold standard of treatment is complete surgical resection followed by craniospinal radiotherapy in adults and older children but infants receive postoperative chemotherapy. Routine surveillance MRI head at regular intervals are needed for detection of asymptomatic recurrences.

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