ISSN: 0975-833X

Available online at http://www.journalcra.com

International Journal of Current Research
Vol. 12, Issue, 01, pp.9659-9662, January, 2020

DOI: https://doi.org/10.24941/ijcr.37614.01.2020

INTERNATIONAL JOURNAL OF CURRENT RESEARCH

RESEARCH ARTICLE

INTRAPARENCHYMAL EXTRAVENTRICULAR SUPRATENTORIAL EPENDYMOMAS- RARE CASE SERIES

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ARTICLE INFO

Article History:
Received 04th October, 2019
Received in revised form
15th November, 2019
Accepted 19th December, 2019
Published online 30th January, 2020

Key Words:
Supratentorial,
Extraventricular,
Intraparenchymal, Ependymoma.

ABSTRACT

Introduction: Ependymomas are glial series tumours that can occur throughout the neural axis, usually in close proximity to the ventricles (especially the fourth ventricle) or central canal., The histological features,which remain identical despite the varied morphology of intraventricular versus intraparenchymal tumours, are also considered. **Case report:** Here we report 4 cases of extraventricular intraparenchymal supratentorial tumors. Headache was the commonest presenting complaint seen followed by vomiting, seizure, raised intracranial pressure etc. Surgery was advised as the first line of management. Two patients recovered post operatively and are asymptomatic under follow up. Two patients were taken for adjuvant radiotherapy, one in view of recurrence and another patient in view of subtotal resection. We present the symptomatology, histology, imaging characteristics and management options in detail. **Conclusion:** As the tumour is amenable to total radical resection, radical surgery alone is an option. The need for postoperative adjuvant therapy has been controversial for supratentorialependymomas. Postoperative radiation therapy must be administered in every case of partially resected ependymomas. Adults have a better five-year survival rate than children. When considered together, age at diagnosis along with extent of the surgical resection was better correlated to outcome.Patients with symptoms lasting less than 1 month before diagnosis have a worse outcome than those with a more protracted course. In regards to tumour location, patients with supratentorialependymomas have generally a better survival rate than patients with posterior fossa ependymomas.

Citation: Dr. Jayakumar Perumal and Prof. Thiruvalluvan, A.. **2019.** "Intraparenchymal extraventricular supratentorial ependymomas- rare case series", *International Journal of Current Research*, 12, (01), 9659-9662.

INTRODUCTION

Ependymomas are glial series tumours that can occur throughout the neural axis, usually in close proximity to the ventricles or central canal. While the fourth ventricle is a common location for ependymoma, we present four rare cases of an entirely intraparenchymals upratentorial ependymoma, remote from the ventricular surface, The histological features, which remain identical despite the varied morphology of intraventricular versus intraparenchymal tumours, are also considered.

CASE REPORT

CASE 1: A 14-year-old girl presented with progressive symptoms of increased intracranial pressure of 1 month duration. CT scan revealed a right frontal extra-ventricular space occupying lesion with calcifications. She underwent gross total resection. Histopathological examination was consistent with ependymoma. Patient is asymptomatic now and under follow up.

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CASE 2: A 11 year old boy presented with headache, vomiting and seizure for 15 days duration. CT scan revealed a left parietal extra-ventricular inhomogeneous space occupying lesion with solid and cystic components with calcifications. He underwent total resection of the solid component. Histopathological examination was consistent with ependymoma-tumor cells characteristically organized in perivascular pseudo rosettes. Eight months later, he presented to us with persistent headache with seizure. MRI brain showed recurrence of the left parietal lesion. The patient underwent surgical revision with total removal of both the solid and cystic components of the tumour and patient was referred to radiation oncology for adjuvant radiotherapy.

CASE 3:6 year old girl presented with headache for 15 days, vomiting – 4 episodes with blurring of vision. Papilledema was seen on fundus examination. MRI revealed an extra ventricular 4.8cm X 6.1cm X 5.1cm left thalamic cystic SOL. She underwent Right ventriculo-peritoneal shunt in private hospital for obstructive hydrocephalus. Left tempero-parietal craniotomy done. Through brain cannula the cyst was tapped at a depth of 2cm and we aspirated about 15ml of cyst fluid. Subtotal excision of the lesion was done under microscopic

guidance. Lateral wall of ventricle with thin rim of brain parenchyma was intact. Post-operative period was uneventful. Patient was referred to radiation oncology for adjuvant radiotherapy and kept under follow up.

CASE 4:18 year old male came with complaints of headache for past one month, 2 episodes of seizures and right hemiparesis for 15days.Left fronto-parietal craniotomy was done. Corticotomy was done and we exposed the solid component which was reddish-gray, glistening, highly vascular and friable. Gross total resection of the tumor was done..Microscopic examination of the resected specimen showed a clear cell tumor with cells predominantly arranged in sheets and nests, without any unequivocal areas of perivascular pseudorossettes or ependymal rosettes. Individual tumor cells were round to oval with clear cytoplasm, having round hyperchromatic nuclei and inconspicuous nucleoli. Focally, cells showed mild anisonucleosis, nuclear atypia and increased mitosis (4-6/hpf). Areas of reactive gliosis, calcification and endothelial proliferation were also present. IHC showed immunoreactivity for GFAP, strong positivity for vimentin and dot positivity for EMA. This confirmed the diagnosis of intraparenchymal clear cell ependymoma.

DISCUSSION

In 1863, Virchow defined ependymomas as tumors with ependymal cells forming ependymal rosettes and perivascular pseudorosettes (Ho, 2001). Ependymomas are glial derived tumours that can occur throughout the neural axis, usually in close proximity to the ventricles (especially the 4th ventricle) or central canal. Extra ventricular ependymoma arise from trapping of embryonic rests of ependymal tissue in the developing cerebral parenchyma. They account for ~5% of all neuroepithelial neoplasms, ~10% of all pediatric brain tumors and up to 33% of brain tumors occurring in those less than 3 years of age. This distribution correlates with molecularly distinct tumors which in turn have different epidemiology and prognosis : posterior fossa: 60%,supratentorial ependymoma: 30%, spinal cord/canal: 10%.

Pathophysiology: As its name implies, extra-ventricular ependymomas arise outside the walls of the ventricular system. One of the etiological theories is based on the postulate that, remnant nests of ependymal cells are present in the frontal lobe, anterior and inferior to the normal extent of the frontal horns of the lateral ventricles; these remnants come from an extension toward the frontal lobe bases of the frontal horns, so-called “olfactory ventricle”, that collapses and regresses during normal development. However, random distribution of ependymomas around the periventricular region is also seen, rather than restriction to the angles of the ventricles (Furie, 1995).

Macroscopic appearance: Macroscopically, ependymomas tend to be well defined, lobulated, grey or tan-colored soft and frond-like tumors which are moderately cellular They may have focal areas of calcification.

Microscopic appearance: Microscopically, these tumors are characterized by well-differentiated cells. Characteristic features include ependymal rosettes, which are uncommon but pathognomonic and perivascular pseudorosettes which are far more common and seen in most of ependymomas (Wippold, 2006).

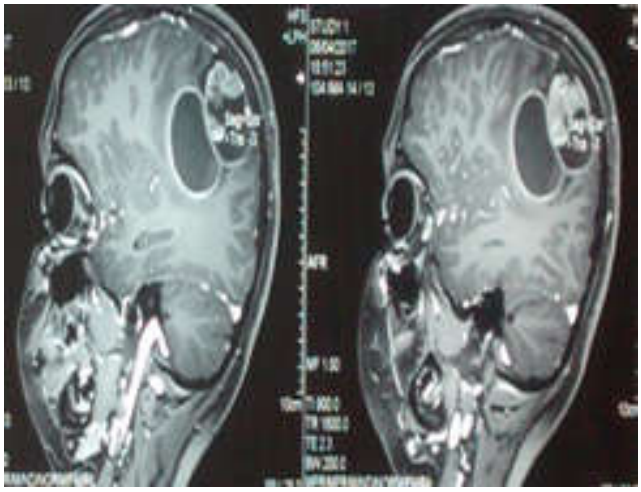

Figure 1. T1 weighted MRI with contrast showing a heterointense lesion in left frontoparietal region with hemorrhagic/necrotic component with irregular peripheral enhancement

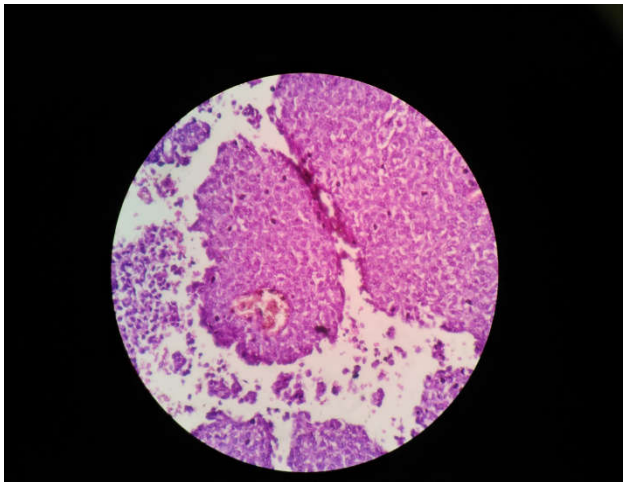

Figure 2. HPE showing perivascular pseudorosette

Diagnostic imaging: Ependymomas are typically heterogeneous masses with areas of necrosis, calcification, cystic change and hemorrhage frequently seen. This results in a heterogeneous appearance on all modalities. Intraparenchymal lesions (usually supratentorial) are generally large and variable in appearance, ranging from completely solid, enhancing masses to cysts with a mural nodule, or more heterogeneous masses (Smith, 2013). They are believed to arise from trapping of embryonic rests of ependymal tissue in the developing cerebral parenchyma. In CT, coarse calcification is common (50%) , followed by cystic areas (50%) with solid component iso- to hypointense with heterogeneous enhancement and variable haemorrhage. In MRI, T1 shows solid portions of ependymoma typically asisointense to hypointense relative to white matter (Smith, 2013). T2 shows hyperintense region relative to white matter which is more reliable in differentiating tumor margins than non-contrast T1-weighted images (but less reliable than contrast enhanced T1)T2* (e.g. SWI) shows foci of blooming from hemorrhageor calcification. T1 Contrast enhanced (Gd)- shows heterogeneous enhancement, useful in differentiating tumor from adjacent vasogenicedema and normal brain parenchyma. DWI/ADC shows restricted diffusion of solid components, especially in anaplastic tumor. Diffusion should be interpreted with caution in masses with significant hemorrhage or calcification.

MRS shows Choline peak elevation according to the cellularity of tumor, NAA peak reduction, elevated Cho/Cr ratio and lipid and lactate rise when degeneration occurs. Although CSF seedingis uncommon when compared to tumors like medulloblastomas, careful examination of the entire neuraxis is required. There is no specific MR image for intra parenchymal ependymomas. They may include calcifications in different patterns, or have heterogeneous enhancement. Differential diagnoses include oligodendroglioma, astrocytoma, germ cell tumors, desmoplastic infantile ganglioglioma, and primitive neuroectodermaltumor. The supratentorialependymomas tend to be larger in size than those infratentorial. Armington *et al*. (1995) found that 94% of supratentorialtumors manifest with a size larger than 4 cm, while most infratentorialependymomas are significantly smaller at presentation. Their shape is irregular in the posterior fossa accommodating to shape of ventricle or cisterns; however they are spherical in the cerebral hemisphere. Supratentorialependymomas often contain a cystic component, while infratentorialependymomas are more often solid tumors (Furie, 1995; Armington, 1985). Signal from ependymal cysts is higher than that from CSF. FLAIR sequence better visualizes the border between tumor and ventricular wall. Calcifications, ranging from small punctuate foci to large masses, are very common in both infra and supratentorialependymomas (40-80% of cases) (Armington, 1985; Morrison, 1984). Our patient presented with a tumor larger than 4 cm in size, mainly cystic with calcifications imbedded in the wall and in the solid part.

Histopathology: Contrary to the brain infiltrating fibrillaryastrocytomas, most extraventricularependymomas are well circumscribed and well demarcated from the normal cerebral parenchyma thus amenable to total surgical excision. At histological analysis, ependymomas are moderately cellular tumors with rare mitotic figures. The tumor cells are characteristically organized in perivascular pseudorosettes and, less commonly, ependymal rosettes. Shuangshoti *et al*. (Shuangshoti, 2005) found that there was no significant relation between histopathology, Ki-67 proliferation index, p53 immunolabeling, tumorploidy, and biological behavior.

Treatment: As the tumor is amenable to total radical resection, radical surgery alone is a reasonable option as the initial treatment for solid extraventricula rtumors located far from clinically eloquent brain areas. A postoperative MRI with contrast has also been recommended for further evaluation of the extent of resection (Vinchon, 2001; Palma, 2000). Early second-look surgery may be proposed to achieve total excision in selected patients with accessible residual tumor detected on postoperative MRI (Vinchon, 2001). The need for postoperative adjuvant therapy has been controversial for supratentorialependymomas. Postoperative radiation therapy must be administered in every case of partially resected ependymomas due to proximity to eloquent areas. So, in general, it is considered safe to observe the patient when postoperative CT or MR shows gross total excision, particularly when the tumor is of low grade. Besides, supratentorialependymomas tend to recur in regions amenable to surgery. Therefore, reoperation to attempt complete tumor resection before initiation of radiotherapy should be considered (Vinchon, 2001; Palma, 2000). Some authors recommend adjuvant radiotherapy if the tumor is cystic, even after apparently total resection. Adjuvant radiotherapy has also been suggested to be given to patients with anaplastic ependymomas (Vinchon, 2001; Palma, 2000). In our case we considered complementary radiotherapy as the cyst wall was incompletely removed after the first surgical procedure. Of the different variants of ependymomas, cellular and papillary are the most common, while clear cell and tanycyticependymomas are uncommon and constitute rare case reports. Unlike most ependymomas, clear cell ependymomas (CCE) are associated with an aggressive behavior and early recurrence despite gross total resection, and needs more vigorous management. Prophylactic craniospinal irradiation is no longer advocated unless cerebrospinal seeding is evident on imaging or cerebrospinal fluid studies (Palma, 2000). The benefit of chemotherapeutic agents is still questionable. They took place particularly in children for whom radiotherapy is contraindicated or in previously irradiated subjects with an inoperable tumor (Vinchon, 2001).

Prognosis: Throughout the literature, three major factors were identified determining the outcome. The most important prognostic variable was the presence of radiologic residual disease, seen at postoperative MR imaging or CT. The 5-year-tumor-free survival rate was 75% and 15% for patients with no radiologic evidence of residual tumor and those with residual disease in which progression cannot be stopped, respectively (Healey, 1991). Poor prognostic factors include a 4th ventricular location, anaplastic variant and incomplete resection. As such, children have a worse prognosis (both 4th ventricular location and anaplastic variant are more common in children). Overall, the 5-year survival rate in children ranges from 50 to 75% [7]. Once recurrence has occurred, the prognosis is very poor, with a mortality rate of 90% [7]. When considered together, age at diagnosis along with extent of the surgical resection was better correlated to outcome (Spoto *et al*., 1990). The last prognostic variable is the duration of symptoms preceding diagnosis. Patients with symptoms lasting less than 1 month before diagnosis have a worse outcome than those with a more protracted course (Pollack, 1995). In regards to tumor location, patients with supratentorialependymomas have generally a better survival rate than patients with posterior fossa ependymomas (Kudo, 1990).

Conclusion

As the tumour is amenable to total radical resection, radical surgery alone is an option. The need for postoperative adjuvant therapy has been controversial for supratentorialependymomas. Postoperative radiation therapy must be administered in every case of partially resected ependymomas. Adults have a better five-year survival rate than children. When considered together, age at diagnosis along with extent of the surgical resection was better correlated to outcome.Patients with symptoms lasting less than 1 month before diagnosis have a worse outcome than those with a more protracted course. In regards to tumour location, patients with supratentorialependymomas have generally a better survival rate than patients with posterior fossa ependymomas.

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