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Intraventricular Meningiomas in Adults –A Case Series and Review of Literature

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ABSTRACT

Intraventricular meningiomas account for 20 to 30% of all the tumours which are found in the intraventricular region, and its incidence is higher in childhood and adolescence. Intraventricular location of the meningioma itself is rare site and they accounts for nearly 0.5% to 5% of all intracranial meningiomas. We present our experience; nine patients were with operated with diagnosis of intraventricular meningioma from 1982 to 2013. The mean and median age was 44 and 42 years respectively (range 18- 69 years), 6 females and 1 male . Duration of symptoms range between 25 days and three years. All tumors were located in the lateral ventricles. Presentation, imaging features, surgical procedures performed, histological diagnoses, and postoperative outcomes were analyzed. Eight patients underwent the parieto- occipital Transsulcal/transcortical craniotomies and one patient underwent occipital approach. The tumour was seen in the left side in the seven patients and right sided in two patients. In seven patient, simpson grade I resection was achieved and grade II in two patients. Majority of the patients improved post operatively. One patient had transient memory loss which improved after 6 weeks. Two had persistent homonymous hemianopia and one improving hemiparesis. In our series there were three meningothelial meningiomas, two fibrous , one transitional , one anaplastic ,one atypital and one rhabdoid variety.

Keywords: NIL

INTRODUCTION

Intraventricular meningiomas account for 20 to 30% of all the tumours which are found in the intraventricular region, and its incidence is higher in childhood and adolescence. Intraventricular location of the meningioma itself is rare site and they accounts for nearly 0.5% to 5% of all intracranial meningiomas[(1)].A marked female predominance is seen in our series and in all other series discussed below [(1,8,9).] The intraventricular meningiomas are slow-growing tumors, hence they either present late or are silent at presentation [(2,28,35).]The anatomical distribution of these lesions are 77.8% in the left lateral ventricle, 15.6% in the third ventricle, and 6.6% in the fourth ventricle[(3).] As mentioned above the intraventricular meningioma are rare and many case reports have been published so far and only few case series in world. In this case series , the first case series in south india , we have presented our experience with 9 cases of intraventricular meningioma , based on the epidemiology symptomology ,approach and surgical outcome with discussion and review of literature till date.

MATERIAL, METHODS AND RESULTS-

We included all ventricle based meningiomas , excluding all the falcine,tentorial secondary tumors and operated nine patients with diagnosis of intraventricular meningioma from 1982 to 2013. The mean and median age was 44 and 42 years respectively (range 18- 69 years). There were six women and three men. Duration of symptoms range between 25 days and three years. All tumors were located in the lateral ventricles. Presentation, imaging features, surgical procedures performed, histological diagnoses, and postoperative outcomes were analyzed. Headache was present in 7 patients. In two , there were symptoms of intracranial hypertension (vomiting). Two patients presented with cognitive impairment at onset. Three patients presented with motor impairment, manifested as hemiparesis 2 and dysphasia 1 ,three patients presented with homonymous heminopia . Papilledema was seen in eight patients. In all the patients pre- operatively computerized tomography scan of the brain was done showing high density lesions , seven had it in left side and two in the right side. The contrast enhancement was homogenous in 7 and heterogenous in two patients.Six patients had perilesional edema. Two patients had trapped occipital horns. Magnetic resonance imaging (MRI) was performed in 7 patients. In all cases, the tumours were isointense on T1 weighted images and homogeneous gadolinium enhancement was present in 6 patients, and heterogeneous in 1 patient (Figure 1). On T2-weighted images the tumours were hyperintense. Signal voids because of calcification were visible in one cases. Carotid and vertebral angiography and embolization was not performed in any of our cases. Tumors were predominant on the left ventricle seven patients; two patients had it on the right side in the ventricular trigone out of 9 patients. Complete clinical, radiological, histological surgical approach and the outcome have been depicted in**.** The results were analysed for immediate post operative period and long term follow up as well .Eight patient underwent the parieto- occipital Transsulcal/transcortical craniotomies and one patient underwent occipital approach. The tumour was seen in the left side in the seven patients and right sided in two patients. In seven patient, simpson grade I resection was achieved and grade II in two patients. Majority of the patients improved post operatively. One patient had transient memory loss which improved after 6 weeks. Two had persistent homonymous hemianopia and one improving hemiparesis. In our series there were three meningothelial meningiomas, two fibrous , one transitional ,one anaplastic ,one atypital and one rhabdoid variety.

DISCUSSION

EMBROLOGY

Presence of arachnoid cell nests in the normal choroid plexus stroma has been discussed in the literature, and a microscopic examination of the choroid plexus usually reveals collections of these cells.[1,2,3]

The presence of these arachanoid nest cells in the choroid plexus has been explained in various texts. The choroid plexus initially develops from the invagination of a mesenchyme in roof of the myelencephalon during the 6th week of gestation.[4] Around 7th to 9th weeks, the telencephalic choroid plexus has initiated to develop a loose mesenchymal stroma, which is covered by a layer of cells derived from the ependyma.[5] This arachnoid tissue is then transported together with the choroid plexus as the ventricular system invaginates[6] and the end of 20 to 40 weeks, the central stroma of the choroid plexus contains meningocytes, connective tissue, and blood vessels.[7] Meningiomas arise from these arachnoid cap cells, which are specialized cells in arachnoid granulations.

SYMPTOMS AND SIGNS-

Majority of the clinical symptoms of intraventricular meningiomas are to due raised intracranial pressure. Mass effect due to direct compression on adjacent brain structures is another common clinical manifestation. These tumours are slow and reach a substantial size prior to becoming symptomatic as the ventricle accommodates lesion for a longer time . The symptoms may occur earlier if the tumor causes obstruction of the cerebrospinal fluid (CSF) outflow [8,9,10]. On reviewing the literature, duration of symptoms ranged from a few days to several years. Major symptoms were signs of increased intracranial pressure (86%), corticospinal tract signs (43%), visual field defects (36%), cognitive changes (29%), and seizures (7%) [11,12,13]. We had similar clinical presentation in our case series were increased

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intracranial signs (77%)the corticospinal tract signs (33%),visual field defects (33%) and dysphasia in 11%

There are few case series reported of intra-ventricular meningioma in adults [14,15]. Due to the rarity of these tumors, autopsy findings are less[16] A is rare association of the intraventricular meningiomas with intracranial hemorrhage has also been reported . [17,18,19]. Subarachnoid hemorrhage at presentation was described in a few case series [20]. In our case series no patient presented with intraventricular haemorhage.

Differential diagnosis

The most important differential diagnosis is the intraventricular solitary fibrous tumor [21,22,23]**).** Intraventricular meningioma are the most common in the trigone of lateral ventricle , in adults **(v).** The differential diagnosis of lateral intraventicular trigonal tumors ,include choroid plexus papilloma in patients below 10 years of age; low-grade gliomas,as ependymoma, oligodendroglioma, and low-grade astrocytoma, in patients ranging from 10 and 40 years of age; and metastases and lymphoma after the fourth decade of life [24,25,26].

Pathology

Histopathological features of these tumors are similar to those seen in meningiomas in other locations **[24].** Bertalanfy discovered in his series that most of intraventricular meningiomas were meningothelial, transitional (mixed), or lymphoplasmacyte- rich meningiomas (81%) and three tumors were classified as atypical (19%) **[9].** Rare pathological types described are rhabdoid, osteoblastic and chordoid [25,26,27]. In our series the majority were benign lesions (7/9). There were three meningothelial meningiomas, two fibrous and one transitional, one anaplastic, one atypital and one rhabdoid variety.

Surgery

Bhatoe et al, in 2006 presented a series of 12 intraventricular meningiomas (IVM) in his experience, a parieto-occipital (trigon) craniotomy should be performed through for lateral ventricular, transcortical- transventricular route for third ventricular, and sub-occipital craniotomy for fourth ventricular tumors [24]. Trigonal meningiomas are commonly resected via intra parietal/inter-parietal or parietal-occipitalapproach. Neuro-navigation preoperatively can be very useful for pin point localization and avoidance of neurological morbidity. Bertalanfy et al utilized neuronavigation in 8/16 of their patients [11]. The surgical management of intraventricular meningiomas requires a proper and careful planning. Tumors involving the dominant lobe, preoperative neuropsychological testing should be obtained. Meningiomas are solid, discrete lesions that can be totally excised. The aim should be, to reach the feeding vessels with minimal brain resection, and coagulation of the tumor debulking the lesion, which can be achieved by defining the plane between the tumour and the normal parenchyma followed by internal decompression, and cutting off the feeding vessels. Piecemeal tumor removal can be easily achieved. Special intra-operative attention should be paid to the choroid vessels [28]**.** Placement of the post operative extra-ventricular drainage, can avoid complications arising due to hydrocephalous [29]. In our series 7/9 patients underwent parieto-occipital transulcal approach,initially plane between the tumour and the normal parenchyma defined then the feeding vessels were identified ,the blood supply cut and the tumour was debulked in piece meals. All patients had external venricular drainage post operatively for 3-5 days

Approach related complications

Transient memory disturbance after removal of an intraventricular meningioma by a parietal-occipital inter-hemispheric pre-cuneous approach was described in literature [29]. In our series only one case presented with transient memory disturbance after parietal-occipital approach which improve later.

Outcome- One of our cases had the diagnosis of rhaboid meningioma ,3 months later patient came to us with malignant brain edema. Drop metastasis in the subarachnoid of intraventricular meningiomas has been detailed in the literature [30,31,32]. Hence clinician should continuous follow-up for the cases for new or recurrence of the neurological symptoms in- spite these tumors being usually benign.

Conclusions

Intraventricular meningiomas in adults are rare with predominance in woman.These tumour remain silent till they acquire considerable sizes to produce symptoms and signs when they cause hydrocephalous

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aor mass effect. There are no pathognomonic signs or clinical features which can be attributed to intraventricular meningiomas.A proper clinical assessement and the radiological information can lead to a precise surgical planning and in turn better outcome. Keys to surgery are defining the tumour plane early, isolation of the control of the feeding vessels, periodic debulking and post operative extraventricular drainage. Recurrences are rare after complete excision. Histologically these tumors are same as that of meningiomas elsewhere.

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