Case Report

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Giant Choroid plexus papilloma in a child: A Case Report

Tumors of the choroid plexus are rare neoplasms of neuroectodermal origin, accounting for less than 1% of all intracranial tumors**.** We report a case of giant choroid plexus papilloma of the lateral ventricle extending into the third ventricle of a 3 years old female child. She presented with features of obstructive hydrocephalus and had undergone V-P shunt at the age 3months. She repeatedly underwent revision of the V-P shunt at the age of 6 months, 22 months and 36 months prior to the tumor excision. An anterior interhemispheric, transcallosal approach was performed and total excision was achieved with fenestration of lamina terminalis at the age of 37 months. Following the surgery the patient developed bilateral subdural effusion requiring bilateral subdural-peritoneal shunt and the non-functional V-P shunt was removed. Histological evaluation confirmed the diagnosis of the choroid plexus papilloma.

Key Words: Choroid plexus, papilloma, surgery

Tumors of the choroid plexus are rare neoplasms of neuroectodermal origin, accounting for less than 1% of all intracranial tumors. These tumors are primarily found in children, but they can be observed as well as in adults , infants and prenatal occurrence has been reported.[1] In children, tumors of the choroid plexus are located most often in the lateral ventricle, but they can also be found in the third ventricle and in the fourth ventricle.

Of particular surgical interest in cases of choroid plexus tumors are:1) the associated hydrocephalus, its pathophysiological characteristics and the way it interferes with removal of the tumor; [7] and 2) the increased vascularity of these tumors, which makes surgery challenging, particularly in cases of carcinoma because the patients tend to be of young age. Although papillomas are usually cured by surgery, carcinomas have a tendency to recur. The type of adjuvant treatment necessary remains open to discussion.[3]

Case Report

A 3-years-old female child presented with history of severe vomiting, decreased feeding, agitation, restlessness and altered consciousness for three days. She was a diagnosed case of obstructive hydrocephalus with giant intraventricular mass extending into lateral and third ventricle **(Figure 1)**. She had undergone V-P shunt at the age of 3 months and surgical excision of the mass was abandoned due to poor physical condition of the child believing that the major surgical intervention may increase the risk of morbidity and mortality. Repeatedly revision of the shunt was done at the age of 6 months, 22 months and 33 months. She was planned for surgery with total excision of tumor due to repeated complication of shunt block. Total tumor excision was performed through anterior interhemispheric, transcallosal approach with fenestration of lamina terminalis. Post operatively she had gradual fall in her conscious level and CT scan brain

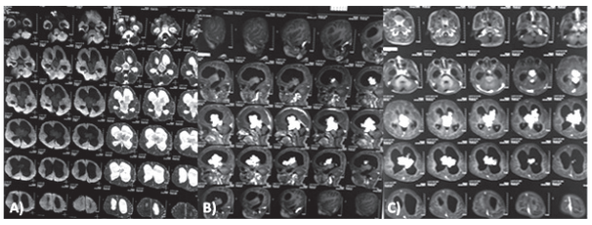

Figure 1: A) T1WI image showing hypo to isointense mass inside the lateral ventricle and T2WI isointense mass, B) Post Contrast sagittal image showing marked enhancement of the lesion, C) Post Contrast axial image showing marked enhancement of the intraventricular mass

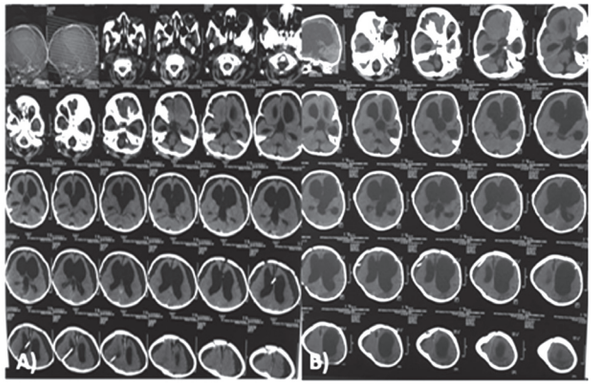

Figure 2: A)Post Op CT scan brain showing B/L subdural effusion with V-P shunt in situ, B)CT scan brain showing B/L subduro- peritoneal shunt and regressed subdural effusion

revealed bilateral subdural effusion. Repeated tapping of the subdural effusion was done but recollection occured after few days of each tapping, hence on 10th post-operative day she was planned for bilateral subduro-peritoneal shunt and removal of the non-functional V-P shunt **(Figure 2 and 3)**. Following subduro-peritoneal shunt there was gradual improvement in her conscious level and regressed the volume of subdural effusion. She made good recovery and was discharged on 19th day. Followed up CT scan brain showed ventriculomegaly with opening of sulci and gyri.Since she was asymptomatic we did not plan for CSF diversion surgery and recommended for regular follow up.

Discussion

CPP is a slow-growing, benign neoplasm that arises from the choroid plexus epithelium. It constitutes about 3% and 0.5% of all intracranial tumors in children and adults respectively.[2] It usually occurs in the first and second decades of life, but has been reported in other age groups also. CPP commonly occurs in the lateral ventricles in children, and in the fourth ventricle in

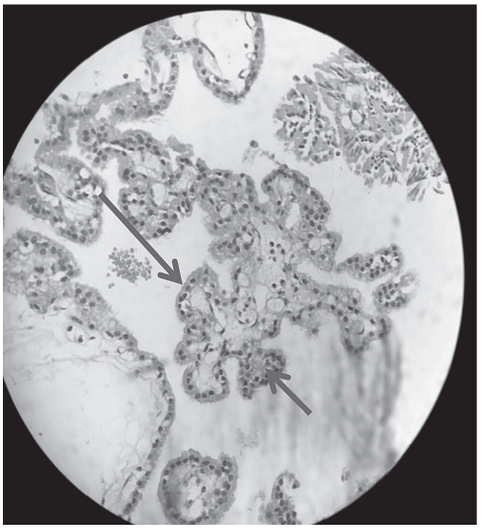

Figure 3: Histopathological picture, long arrow above showing papillary projections and short arrow showing columnar/cuboidal cells (lining epithelium)

adults; other uncommon sites are the third ventricle and the cerebellopontine angle. CPP has also been reported in the suprasellar region, cerebellum, and the frontal lobe. [8] In our case, the tumor was located in the lateral ventricle and 3rd ventricle.

CPP predominately presents with features of raised intracranial pressure in the form of headache, vomiting, and visual deterioration. Progressive enlargement of the head and excessive crying are present in majority of the infants. Raised intracranial pressure is due to hydrocephalus as a result of obstruction of the CSF pathway, excessive production of CSF, and recurrent occult bleeding due to the tumor, leading to arachnoidal fibrosis and adhesions. Seizures and mental changes may often be the only clinical features in these lesions and fourth ventricular tumors may also have cerebellar signs. Cerebellopontine angle CPP has been reported to present with cranial nerve deficits.[4,10] The predominant clinical feature in our case was due to raised intracranial pressure.

The classical CT findings of CPP have been described as a well-defined, homogeneously enhancing, lobulated mass with an irregular frond-like pattern, resulting in a cauliflower-like appearance typically seen in cases of intraventricular tumors. Fine, speckled calcification is often seen within the tumor. MRI may reveal hypo- or isointense lesion on T1WI and hyperintense lesion on T2WI with marked enhancement on contrast. Varying degrees of associated hydrocephalus is also present in almost all the cases.[9, 12, 13] Calcification is present in some cases.

Grossly, choroid plexus papillomas appear as pink or reddish globular masses with a rough, irregular

surface resembling a cauliflower. They are very vascular and sometimes, significant calcification is noted. Microscopically, they resemble the normal architecture of the choroid plexus and show papillae composed of a single layer of columnar or cuboidal epithelium lining a stroma of vascularized connective tissue. Features of microscopic invasion, mitotic activity, and pleomorphism should raise the possibility of malignancy, even when the general architecture indicates a well differentiated papilloma.[6] The histopathology results for our case was characteristic of CPP, with no evidence of malignancy.

The treatment of choice of CPP is surgical excision of the tumor. Total excision should be the aim and is usually achievable. As the prevention of bleeding is a major consideration during surgical excision of these lesions in pediatric patients, gentle coagulation of the tumor under constant irrigation (to shrink the tumor and remove it in totality), is preferred to its piecemeal excision. The thalamostriate vein was meticulously preserved in the lateral ventricle anterior to the tumor. The vascular pedicle supplying the tumor was coagulated and divided during the last part of tumor removal as retraction of a bleeding pedicle may result in profuse ventricular hemorrhage and brain edema. Hydrocephalus is generally relieved following excision of the tumor and fenestration of the floor of 3rd ventricle just after excision of tumor may prevent subsequent CSF diversion surgery for hydrocephalus but in some cases with persisting postoperative hydrocephalus (usually due to arachnoidal adhesions at the level of subarachnoid spaces and arachnoidal granulation leading to the communicating hydrocephalus), a permanent CSF diversion procedure may be required.[5] Postoperative radiation therapy is indicated in cases with incomplete excision although its validity is doubtful. Radiotherapy also has a role in cases where histopathology is suggestive of features of malignancy.[11] In our case, child underwent total tumor excision and did not required postoperative radiotherapy.

Conclusion

CPP is the rare neoplasm of the childhood. Raised intracranial pressure is the most common clinical presentation. Repeated shunt block is the most common complication .Total surgical excision of the tumor gives complete cure, achievable with minimum morbidity and provides an excellent long-term outcome. Hydrocephalus usually resolves following the total tumor excision due to opening of CSF pathway and Fenestration of the floor of 3rd ventricle after total excision of CPP may resolve hydrocephalus not requiring CSF diversion surgery in future.

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