Choroid Plexus Papilloma

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Choroid plexus papillomas are benign tumors arising from the choroid plexus epithelium. They are very rare and the incidence is less than 1% of all intracranial tumors. Choroid plexus papillomas are slow growing, well-differentiated neoplasms and are usually intraventricular. Reports of this tumor are sparse in Indian literature in recent years(l). We report one such case of choroid plexus papilloma.

Case Report

A $7^{1/2}$-year-old male child presented to us with history of, intermittent occipital headache and pain in the nape of the neck since six months. He also had truncal ataxia with weakness of all four limbs and a gradual history of being unable to walk and reach out for and hold objects and feed himself. On enquiry, there was history of irritability since 2-3 months, and occasional vomiting during the same period. He also gave history of bowel and bladder disturbance in the form of urgency for one week. There was no history of any visual disturbances, speech affection or any other cranial nerve involvement. He did not have any altered sensorium, convulsions, head trauma or tuberculous contact.

Examination revealed an averagely built boy with normal general examination. On examination of the central nervous system, the child was conscious, with normal speech. Fundoscopy showed evidence of bilateral papilledema; vision was grossly normal. There was no other cranial nerve affection. He had clasp kinfe spasticity of all four limbs, with grade III power across all joints. The deep tendon reflexes were exaggerated, with bilateral extensor plantars. Truncal ataxia was present along with intention tremors, dysmetria and dyssynergia. Macewan's sign was positive and neck stiffness was present. The other systems were normal. Our clinical diagnosis was a posterior fossa space-occupying lesion. Radiographs of the skull showed sutural separation with a silver beaten appearance. The magnetic resonance imaging of the brain revealed a large mass *(Fig. 1)* in the posterior horn of the right lateral ventricle attached by a pedicle, with moderate hydrocephalus-the picture being highly suggestive of a choroid plexus papilloma.

The child was operated through a right parietal craniotomy and the lateral ventricle opened by a cortical incision behind the post central gyms. The highly vascular friable tumor was exposed, its pedicle was clamped and cauterized and then the tumor was removed piecemeal. Histopathology revealed the characteristic features of choroid

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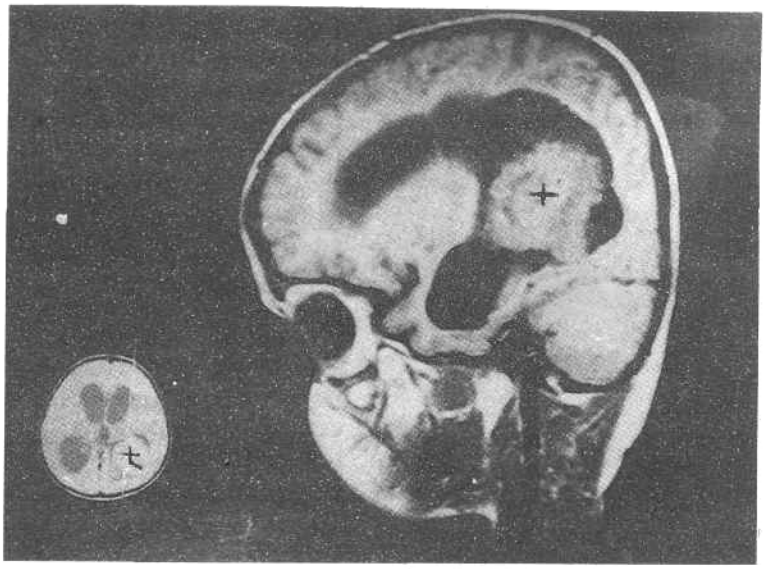

Fig. 1. Magnetic resonance imaging transverse and parasagittal sections showing the intraventricular tumor.

plexus papilloma, with branching papillae of vascular connective tissue stroma, lined by a single layer of columnar epithelium. A CT scan 2 weeks after surgery showed a decrease in the hydrocephalus, with normal appearing choroid plexus. All the signs and symptoms in the child had resolved postoperatively and he was discharged three weeks later.

Discussion

Choroid plexus papilloma is a rare epithelial neoplasm, accounting for less than 1% of intracranial tumors. The incidence reported in children is around 3% of all brain tumors(2). The age of incidence varies depending on the location and histology of the tumor. The fourth ventricle is the most common site of occurrence, followed by the lateral and third ventricles. In children, it occurs more commonly in the lateral ventricles, while the fourth ventricle appears to be the preferred site in adults. Benign tumors are more common in the fourth ventricle, while the anaplastic variety are seen mare often in the lateral ventricle(2).

On gross pathology, the papillomas are soft, pink, globular masses with irregular projections and are highly vascular and friable. Microscopically, they resemble the normal choroid plexus, with branching papillae of well vascularized connective tissue covered by a single layer of columnar or cuboidal epithelium(3). The ceils are histologically benign and properly oriented. Clumps of calcium may be seen occasionally.

Clinically, the course of this tumor is a slowly evolving one. Headache is the most common initial presenting complaint. Symptomatology of tumors of the lateral ventricle include convulsions, mental changes, papilledema leading to loss of

vision and focal deficits like hemiparesis(4). With tumors in the fourth ventricle, symptoms are headache, ataxia, nystagmus, cerebellar signs, dizziness, loss of vision, vomiting and diplopia(5). The clinical progression is usually one of gradual deterioration. Our patient had a history dating back to six months before he came to us. Surprisingly, though he had a tumor in the posterior horn of the right lateral ventricle, his symptomatology was like that of a posterior fossa tumor with raised intracranial tension. The weakness of limbs with long tract signs in our patient were probably due to long-standing hydrocephalus(6). The MRI clearly showed elevation of the cerebellum due to a large collection of CSF in the fourth ventricle and the cisterna magna. The presence thus exerted on the cerebellum could explain the cerebellar signs seen in our patient.

Complications that can occur with choroid plexus papillomas include spontaneous hemorrhage from the tumor, dissemination of tumor fragments and hydrocephalus(2). Accumulation of CSF leading to hydrocephalus results from excessive CSF production, intraventricular trapping of CSF or obstruction of extraventricular CSF outflow secondary to hemorrhage-induced arachnoiditis(7,8). Seeding in other areas of the central nervous system occurs m 20% of cases(2).

On investigations, CSF examination shows elevated pressure, increased proteins, xanthochromia and occasionally tumor fragments^), cytology of CSF in choroid plexus papillomas reveals single cells and papillary clusters of cuboidal cells with regular round to oval nucleus with evenly dispersed chromatin and ample cytoplasm(9).

A computed tomography scan, which is diagnostic, shows a hypodense, lobulated intraventricular mass, which may have finely speckled calcification and which enhances brightly after contrast. MRI will show, on Tl weighted images, the tumor to be hypo- or isointense relative to brain, but hyperintense relative to brain. Angiography reveals a tumor blush and AV shunts with enlarged choroidal arteries.

The treatment of choice for choroid plexus papilloma is operative intervention^). Gross total resection of the tumor can be obtained in 60-90% of the cases(10). However, operative mortality can be upto 20% because of the highly vascular friable tumor(ll). Hydrocephalus which persists after removal of the tumor requires ventriculo-peritoneal shunting(8). Radiation is advocated post-operatively to the tumor site, and also to the spinal cord, if seeding is detected, and in recurrent tumors(2).

The long term symptom-free prognosis is relatively good with gross total resection of the tumor and may even be a cure(ll). Recurrence requires a second operation.

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