LATERAL VENTRICLE CHOROID PLEXUS IN INFANT. CASE REPORT AND REVIEW OF THE LITERATURE.

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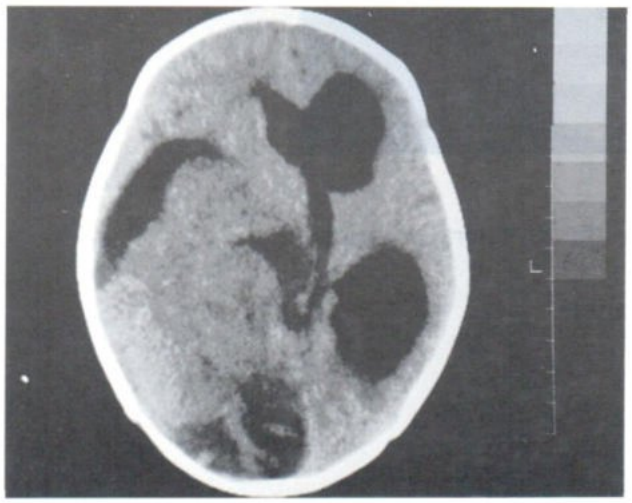
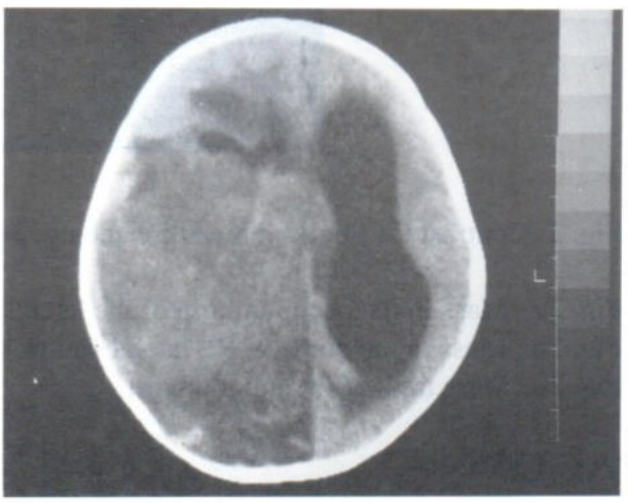
Choroid plexus papillomas are rare intracranial tumours of childhood. They usually occur within the first decade of life. Choroid plexus papillomas are benign lesions that arise from the epithelium of choroid plexus. We report a case of choroid plexus papilloma arising from the right lateral ventricle in a 6 months-old boy. The chief presenting signs and symptoms are drowsiness, poor feeding and papilledema. CT scans reveal a large intensely enhancing mass occupying in the right lateral ventricle with markedly hydrocephalus. The tumor was removed surgically and the pathological diagnosis was choroid plexus papilloma. In this report; we also discuss the pathology, radiologic manifestation by reviewing relevant literatures.

CASE REPORT

This 6 months-old boy was admitted at Suratthani Hospital with the symptoms of drowsiness and poor feeding. On physical examination, he had symptoms and signs of increased intracranial pressure such as projectile vomiting and papilledema, etc. The vital signs were normal. Plain skull films show no definite abnormalities. Cranial CT scan demonstrated a large intense enhancing well defined lobulated hetrogenous mass; about 6x9 cm, filling in the Rt. lateral ventricle and causing communicating hydrocephalus. The patient showed clinical improvement after the tumor was totally removed.



1a



1b

Fig. 1 Pre-contrast CT reveals a large heterogenous isodense lobulated mass, 6x9 cm. In size filling in Rt. lateral ventricle with shifting of midline structure to the Lt. and markedly diffuse hydrocephalus.
Fig. 1a Mass in the Rt. occipital horn
Fig. 1b Mass in body Rt. lateral ventricle.

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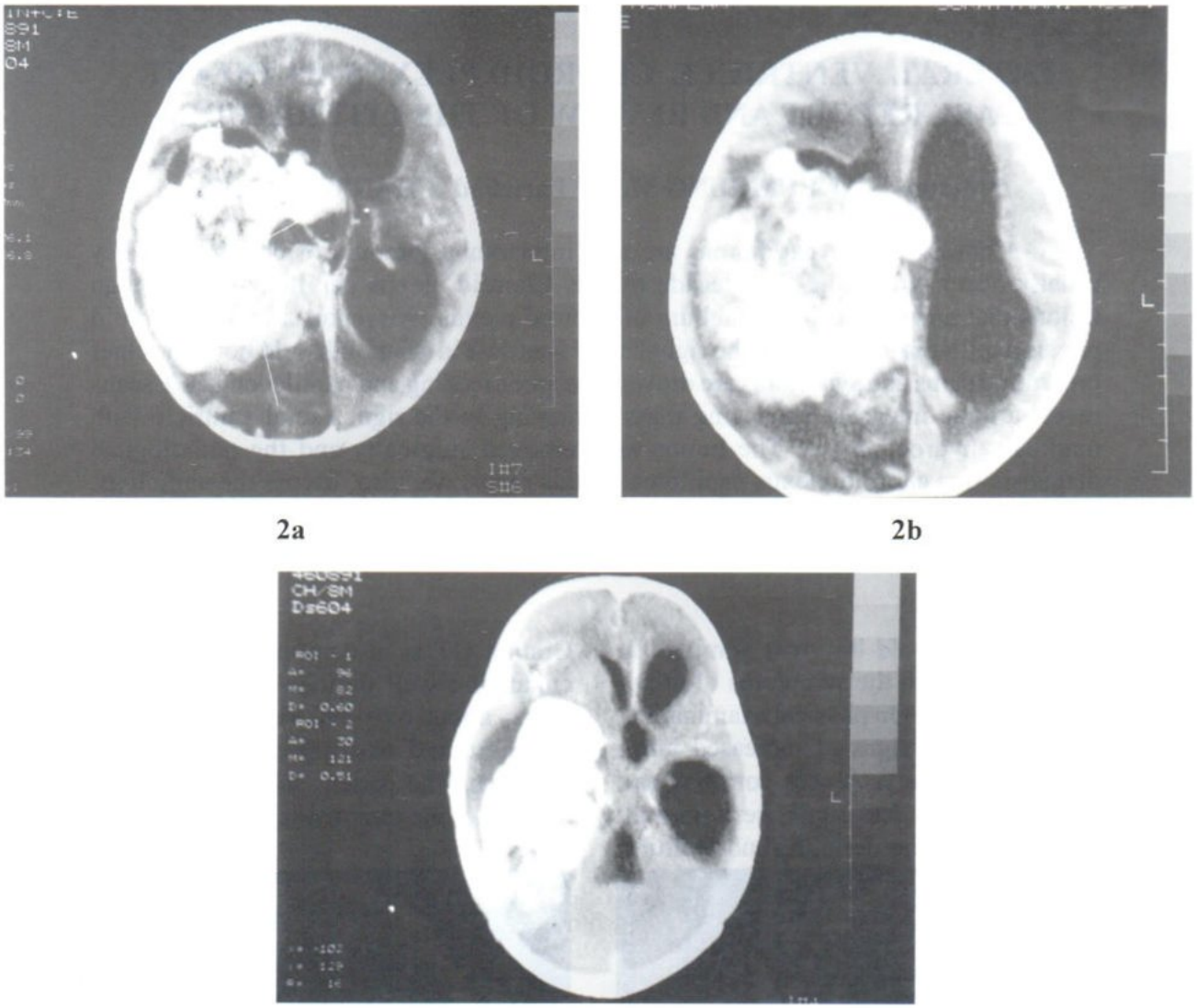

2a 2b

2c

Fig. 2 Contrast CT shows intense enhancing mass in the Rt. lateral ventricle

Fig. 2a Mass in Rt. occipital horn of the lateral ventricle
Fig. 2b Mass in the body of Rt. lateral ventricle
Fig. 2c Mass in the Rt. temporal horn.

DISCUSSION

Choroid plexus are rare and represent about 5% of supratentorial tumors and less than 1% of intracranial tumors in children.[2,3] They usually occur within the first decade of life and 20% -30% occur within the first year of life. They are most common in the neonate. These tumors arise from the epithelium of the choroid plexus.[3,4] The most common site is in the lateral ventricle. Ninety percent arise from the glomus of the lateral ventricle. The remaining papillomas arise from the fourth and third ventricle.[3,4] Histologically, these lesions consist of hyperplasia of both

the surface epithelium of the choroid plexus and the underlying vascular connective tissue core.[5] Occationally, the epithelium may exhibit malignant changes, and such tumors are then classified as carcinomas; these may extend into the adjacent brain parenchyma.[6] 5-10% of choroid plexus papilloma degenerate into carcinomas.[7] The symptoms are related to the large size of the lesion that produces a form of obstructive hydrocephalus. The literature states that choroid plexus papilloma produce an excess of CSF which contributes to the hydrocephalus. This is extremely rare. In the majority of cases, the hydrocephalus from choroid plexus papillomas is secondary to obstruction by the large mass effect of the lesion or results from hemorrhage and adhesions developing in the pathway of the flow of CSF.[8,9]

RADIOLOGIC FINDINGS:

On ultrasound,large choroid plexus papillomas appear as highly echogenic masses with irregular borders related to the glomus ofa the choid plexus and as associated with hydrocephalus. When tumor located in the third ventricle, the tumor can mimic acqueductal stenosis and the diagnosis may prove difficult, particularly in the prenatal period, unless the echogenic mass is visible.[8,9,10] Occationally, intratumoral cysts may also be seen on ultrasound in choroid plexus papillomas. If calcifications are present in the tumor, echogenic foci with shadowing may be seen. Typically, Dopplor ultrasound will show a biphasic flow on choroid plexus papillomas because of their highly vascular nature. This finding has been thought to be specfic for choroid plexus papillomas.[10-12]

CT:

On CT, choroid plexus papillomas may appear as well defined, homogeneous, usually hyperdense intraventricular masses. Tumor margins can be smooth (29%), lobulated (19%), or irregular (52%).[7] Irregular margins are found in 71% of malignant tumors and 43% of benign lesions. Papillomas are iso-dense or hyperdense to parenchyma in 75% of cases, and 25% are hypodense or heterogeneous.[11-13] Tumor calcification on CT has been reported in 24% of choroid plexus tumors.[14] Hemorrhage within the tumor may also be seen. In most case, there is fairly intense, homogeneous enhancement after infusion of contrast medium.[15] Most of the tumors are associated with marked hydrocephalus; occationally, a large mass in the trigone can cause entrapment of the ipsilateral temporal horn. Choroid plexus papillomas may show limited parenchymal invasion[13-17] and it may be difficult to distinguish benign from malignant choroid tumors on imaging studies.

MRI:

On MRI, the tumors appear as an intraventricular lobulated mass isointense or hypointense on T1-weighted images and hypointnse, isointense or hyperintense on T2-weighted images. If hemorrhage is present, the MRI signal intensity will depend upon the age and degree of hemorrhage. After administration of Gd, there is marked homogeneous enhancement.

The differential diagnosis of intraventricular tumor in children includes choroid plexus papilloma, glioma, choroid cyst,ependymoma, teratoma, dermoid, epidermoid ,metastasis and tumors deposited by CSF seeding. Metastasis are unlikely in a young patient. Choroid cyst, epidermoid, dermoid, teratoma, and most craniopharyngiomas can be excluded by their CT and magnetic resonance features.[18-22] Supratentorial ependymomas usually have an intraventricular portion smaller than the component of the tumor mass invading the parenchyma. Choroid plexus papillomas and gliomas possess similar features and may be confused with each other radiographically.[23-25] Choroid plexus papillomas have a lobulated appearance and can be partially differentiated from intraventricular meningioma based on it appearance. Intraventricular meningiomas are round, well-circumscribed lesions, where as choroid plexus papillomas demonstrated a more lobulated appearance.[7]

SUMMERY

Choroid plexus papilloma is a rare tumor found in children which causes communucating hydrocephalus from the results of complex interaction of CSF overproduction and partial restriction of CSF flow as confirmed by previous literatures. The typical radiologic manifestations are intense enhancing lobulated intraventricular mass. The findings may be difficult to be differentiated from other intraventricular masses. Cranial ultrasound is the first image modality in fetus. CT and MRI also offer diagnostic tools to confirm the type of hydrocephalus and tumor evaluation.

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