Choroid plexus papilloma

Choroid plexus papillomas (CPP) are benign brain tumors.

These highly vascular tumors retain the physiological function of choroid plexus and thus lead to overproduction of cerebrospinal fluid (CSF), besides obstructing the CSF pathway.

Epidemiology

The overall incidence is less than 1% of all intracranial tumors.

They are predominantly located in the lateral ventricles in children, and in the fourth ventricle in adults.

They occur in the lateral ventricle in most cases, but have also been described in the third or fourth ventricles [1)]

Most of these tumors occur in the lateral ventricles in neonates.

Third ventricle location is uncommon, limited to a few case reports.

Tumors of the choroid plexus have been described previously in the fetus [2) 3) 4) 5)].

In two cases of choroid plexus papilloma reported at 21 weeks of pregnancy, the diagnosis was suspected because of ventriculomegaly. Both pregnancies were terminated and pathological examination showed bilateral papillomas of the choroid plexus [6)].

Classification

They are histologically classified as plexus papilloma, atypical plexus papilloma, and plexus carcinoma.

Atypical choroid plexus papilloma.

Choroid plexus papilloma in the posterior fossa.

Extraventricular choroid plexus papilloma.

Fetal choroid plexus tumor

Etiology

They can correspond to two different etiologies: papilloma, which is a benign tumor, and carcinoma of

the choroid plexus.

Diagnosis

Choroid plexus papilloma diagnosis.

Differential diagnosis

Choroid plexus papilloma differential diagnosis

Treatment

Choroid plexus papilloma treatment.

Outcome

Total removal is possible for 96% of papillomas and for 61% of carcinomas. Surgical procedures are associated with significant operative morbidity and mortality due to uncontrolled bleeding. The prognosis is widely variable between the two forms, with a survival rate at 5 years of approximately 100% for papillomas and 40% for carcinomas [7].

Case reports

A 43-year-old female with a hemifacial spasm of typical characteristics 6 months after onset. Upon clinical examination, a severe contraction of the orbicularis oculi, orbicularis oris, and superficial muscles of the neck displaying 50 crisis per hour was revealed. Brain magnetic resonance imaging showed the absence of the facial nerve vascular loop in the cisternal portion, with evidence of an intraventricular tumor in relation to the medial portion of the fourth ventricle at the facial colliculus level, indicating a secondary origin of hemifacial spasm. Preoperative electromyography demonstrated irritative electric activity in the muscular branches of the facial nerve. A telovelar approach was performed to the fourth ventricle with intraoperative electrophysiology monitoring, with immediate resolution of the irritative activity after complete tumor resection. The result of the histopathologic study was a choroid plexus papilloma.

Fourth ventricle tumors with extrinsic compression of the facial colliculus represent <0.6% of the causes of hemifacial spasm. Its relationship with choroid plexuses papilloma is being described as the first case reported in the literature. Clinical correlation, imaging, and intraoperative findings in conjunction with intraoperative electrophysiology recordings allow predicting the resolution of symptoms after resecting the lesion [8].

1)
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