Choroid plexus hyperplasia

Choroid plexus hyperplasia (CPH), also known as villous hypertrophy of the choroid plexus, is a rare benign condition that is characterized by bilateral enlargement of the entire choroid plexus in lateral ventricles without any discrete masses. This can result in overproduction of CSF and communicating hydrocephalus.

Pathophysiology

Despite the current knowledge about hydrocephalus, we remain without a complete understanding of the pathophysiology of this condition. glymphatic system (GS) could be more important than the conventional concept of reabsorption of CSF in the arachnoid villi, therefore GS could be a new key point, which will guide future investigations [1].

Pathology

Histology shows an increased number of normal-sized cells.

Radiographic features

MRI

This is best diagnosed by MRI which demonstrates a diffuse enlargement and homogeneous enhancement of choroid plexuses in a patient with communicating hydrocephalus [2].

Treatment

It is a rare condition that may necessitate unusual treatment paradigms.

Although some authors recommend choroid plexus excision or coagulation, ventriculoatrial shunt insertion is a simple and effective treatment modality in cases of diffuse villous hyperplasia of the choroid plexus [3].

It can be seen in trisomy 9p where coexisting congenital heart disease additionally may complicate the therapeutic approach [4].

Case reports

At 20 months of age, a Caucasian girl with trisomy 9 and a family history of an older brother and twin

sister having the same syndrome displayed signs of congenital hydrocephalus due to increasing head circumference. Magnetic resonance imaging revealed enlarged lateral ventricles and a prominent choroid plexus and the girl was treated with a ventriculoperitoneal shunt, which 2 days later had to be replaced with a ventriculoatrial shunt as cerebrospinal fluid production greatly exceeded the ability of the patient's abdominal absorptive capability. At 16 years of age, the patient was diagnosed with cardiomyopathy and diminished ejection fraction. Some months later, she was admitted to the neurosurgical ward showing signs of shunt dysfunction due to a colloid cyst in the third ventricle. Cystic drainage through endoscopic puncture only helped temporarily. Revision of the shunt system showed occlusion of the ventricular drainage, and replacement was merely temporary alleviating. Intracranial pressure was significantly increased at around 30 mmHg, prompting externalization of the drain, and measurements revealed high cerebrospinal fluid production of 60-100 ml liquor per hour. Thus, endoscopic choroid plexus coagulation was performed bilaterally leading to an immediate decrease of daily cerebrospinal fluid formation to 20-30 ml liquor per hour, and these values were stabilized by pharmaceutical treatment with acetazolamide 100 mg/kg/day and furosemide 1 mg/kg/day. Subsequently, a ventriculoperitoneal shunt was placed. Follow-up after 1 and 2 months displayed no signs of hydrocephalus or ascites.

High cerebrospinal fluid volume load and coexisting heart disease in children with trisomy 9p may call for endoscopic choroid plexus coagulation and pharmacological therapy to diminish the daily cerebrospinal fluid production to volumes that allow proper ventriculoperitoneal shunting [5].

A 1-year-old patient was diagnosed with communicating hydrocephalus; ventricle peritoneal shunt (VPS) is installed and ascites developed. VPS is exposed, yielding volumes of 1000-1200ml/day CSF per day. MRI is performed showing generalized choroidal plexus hyperplasia. Bilateral endoscopic coagulation of thechoroid plexus was performed in 2 stages (CPC) however the high rate of CSF production persisted, needing a bilateral plexectomy through septostomy, which finally decreased the CSF outflow.

New knowledge about CSF physiology will help to propose better treatment depending on the cause of the hydrocephalus. The GS is becoming an additional reason to better study and develop new therapies focused on the modulation of alternative CSF reabsorption. [6].

In these patients, intractable ascites can occur after a ventriculoperitoneal (VP) shunting operation. However, shunt-related hydrocele is a rare complication of VP shunting. Previous reports have indicated catheter-tip migration to the scrotum as a cause of hydrocele. Here, we present the first documented case of choroid plexus hyperplasia that led to intractable ascites after shunting and a resulting hydrocele without catheter-tip migration into the scrotum [7].

[1], [6]
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