



## Choroid Plexus Papilloma Causing CSF Shunt Ascites: A Rare Presentation

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### Abstract

Choroid Plexus Papillomas (CPPs) are congenital intracranial tumors of neuro-ectodermal origin. Choroid plexus neoplasms constitute about 0.5% of all intracranial neoplasms. Majority are found in lateral ventricles. Most of these neoplasms are benign papillomas, while one-fifth are malignant carcinomas. The present communication describes a rare case of a choroid plexus papilloma leading to CSF ascites following Ventriculoperitoneal (VP) shunt.

### Case Presentation

A 5 year old boy presented to us with complaints of progressively increasing abdominal distension from past 6 months and respiratory distress for 2 days. There was no history of jaundice or bleeding manifestations. Patient was a known case of hydrocephalus for which medium pressure VP shunt (chabra shunt) was placed at the age of 3 years. On examination child was having massive ascites with positive fluid thrill sign. There was no hepato-splenomegaly and other signs of hepatocellular failure. Neurologically the child was conscious and oriented and there were no signs of shunt dysfunction. Shunt bulb was palpable and soon gets refilled after compressing the bulb. Paracentesis showed clear transudate fluid with no evidence of infection (WBC= 5 cells/mm<sup>3</sup>, all lymphocytes, sugar = 72 mg/dl and protein = 24 mg/dl). Ascitic fluid culture was sterile and was negative for Acid fast bacilli. In addition, cytology was negative for malignant cell. Liver and renal function test were essentially normal (serum bilirubin = 0.6 mg/dl, SGOT = 36 U/l, SGPT = 11 U/l, serum albumin = 3.8 gm/dl, urea = 27 mg/dl, creatine = 0.8 mg/dl). Echocardiography revealed a normal functioning heart. The patient tested negative for HIV. Abdominal Ultrasound and CT scan showed no abnormality in relation to abdominal viscera or peritoneum. CECT head showed gross communicating hydrocephalus with choroid plexus papilloma (Figure 1) in bilateral lateral ventricle which was later confirmed by histopathological examination of postoperative specimen.

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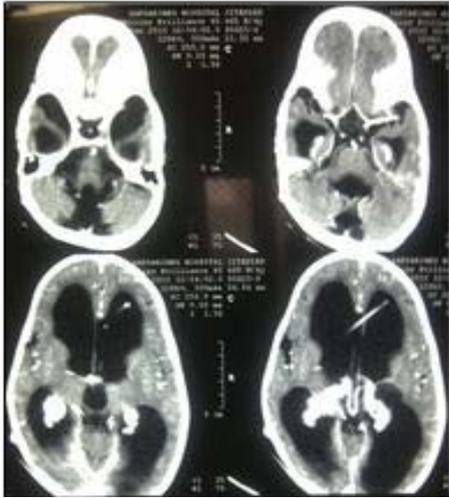
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Despite diuretic treatment and peritoneal tapping the ascites re-accumulated. Based on impression of CSF ascites the lower end of the shunt tip was exteriorized and was maintained as ventricular drain so as to relieve intracranial and intra-abdominal pressures, which continued to drain 1200-1500 ml of CSF daily and ascites resolved within 2 weeks postoperative. Surgical resection of choroid plexus papilloma was done on both sides of lateral ventricles and shunt was removed. The postoperative period was uneventful. Sections from both right and left tumour tissue show multiple fragments composed of delicate fibrovascular fronds lined by single layer of monomorphic cuboidal cells with basal round nucleus. No necrosis, mitosis or pleomorphism seen. S-100 immunostain shows positivity in tumour cells. Above histological features are suggestive of choroid plexus papilloma (Figure 2).

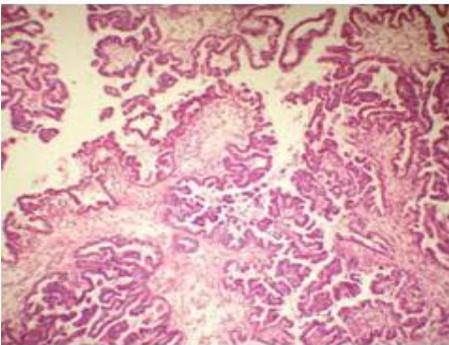
### Discussion

Ascites has been defined as accumulation of excess fluid within the peritoneal cavity [1]. The commonest cause of ascites is cirrhosis of the liver, closely followed by other serious hepatic diseases [2]. In children, hepatic, renal and cardiac diseases are the most common causes. CSF ascites is a rare complication of Ventriculoperitoneal (VP) shunts. VP shunts are usually placed for obstructive or progressive hydrocephalus. Occlusion of the shunt tube and infection are frequently observed as V-P shunt complications. Overproduction of the CSF will be the likely possibility once the shunt infection had been ruled out. Early detection of shunt ascites (noninfective) which is an uncommon occurrence and its aetiology will be helpful for better management.

Different intervals (2 months- 13 years) between shunt placement and symptomatic ascites



**Figure 1:** CEFT head showing gross hydrocephalus with choroid plexus papilloma.



**Figure 2:** Photomicrograph (H&E) showing fibrovascular fronds lined by single layer of monomorphic cuboidal cells with basal round nucleus of choroid plexus papilloma.

have been reported [3-6]. Our case develops ascitis after two and half years of shunt placement. Several etiologic factors had been discussed in literature, but it is the imbalance between peritoneal absorption capacity and amount of CSF Production is the major cause. By this definition, patients with excessive amount of CSF production like choroid plexus papilloma are at risk of developing CSF ascites following VP shunt [3,7,8]. On the other hand, patients with high CSF protein due to chronic infection (tuberculous meningitis) [9] or brain tumors –especially optic glioma [4,8,10] may have difficulties in CSF absorption through peritoneum. Under such circumstances, inflammation has been associated with an increase in leukocytes, impairment of lymphatic flow, and a subsequent increase in intraperitoneal protein concentration due to impaired protein absorption causing ascitis.

Peritoneal inflammation due to repeated shunt revisions [5] or non-specific inflammatory response to shunt material [9], play role in the other side and decrease absorptive ability of peritoneum. Also in brain tumors, especially in astrocytoma and glioblastoma, increased vascular permeability can cause microvascular extravasation of plasma into the peritoneal cavity and cause ascites [11-14]. A large series of twenty-eight patients with cerebrospinal ascites have been reported [5]. Their ages ranged from 10 days to 53 years, but most

patients were children, especially infants. Common etiological factors responsible were congenital hydrocephalus, obstructive hydrocephalus, choroid plexus papilloma, craniopharyngioma and posterior fossa tumour. In our patient choroid plexus papilloma was found to be the cause of CSF ascites. This is thought to be due to imbalance between excess production and its absorption.

Treatment for cerebrospinal ascites is revision of the V-P shunt to ventricular-atrial shunt but in choroid plexus papilloma revision will only relieve ascitis with associated risk of congestive heart failure and bacteraemia. Surgical resection of the papilloma is the definitive cure.

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