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A corny cause of cerebrospinal fluid ascites: A case report and review of literature

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Abstract

Objective: To report a rare cause of cerebrospinal fluid ascites.

Methods: A 37-year-old female with history of intracranial hypertension and a ventriculo-peritoneal shunt was referred to liver clinic for evaluation of newly developed ascites.

Results: Initially, the cause of ascites was thought to be secondary to a liver etiology. However, this was excluded after a comprehensive evaluation including portal pressure measurements. We determined the ascites to be infected cerebrospinal fluid secondary to a rare commensal organism, Corynebacterium non-Jeikeium, which resolved after removing ventriculo-peritoneal shunt, appropriate antibiotics and conversion to a ventriculo-atrial shunt.

Conclusion: Cerebrospinal fluid ascites is a rare complication of VP shunts and since 1976 only 8 cases of Corynebacterium non jk VP shunt infections have been reported in the literature but none associated with ascites. Also this report highlights the beneficial role of transjugular portal pressure measurements in the evaluation of ascites.

Keywords

Gastroenterology, hepatology, infectious diseases, neurology

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Introduction

Cerebrospinal fluid (CSF) ascites is a rare complication in patients with a ventriculo-peritoneal (VP) shunt, and its pathophysiology is still not fully understood. It has been reported that CSF ascites could be due to increased CSF proteins,[1] excessive CSF production from tumors,[2] multiple shunt revisions[3] or infections.[4] The organisms that most commonly infect VP shunts are the *Staphylococcus* species.[5] We are reporting the first case of a common non-pathogenic organism, *Corynebacterium* non-Jeikeium (JK), causing a VP shunt infection resulting in CSF ascites.

Case

A 37-year-old female was referred to the liver clinic for new onset ascites on 13 February 2015. Her past history was significant for idiopathic intracranial hypertension (IIH) and a VP shunt placed in 2011 with a revision in 2012. A lumbar puncture revealed an opening pressure of 31 in September 2014, suggesting the shunt was not functioning. She was going to have the VP converted to a ventricular-lumbar shunt, but during her evaluation, a magnetic resonance imaging examination revealed a moderate amount of ascites and she was referred to the liver center for evaluation. She used to weigh 340 pounds, but over the past year, she intentionally reduced to 192 pounds. She had no other risk factor for liver disease denying alcohol abuse, illicit drugs, tattoos or family history. Medications included acetazolamide, topiramate, fluoxetine, dexlansoprazole and morphine. Review of system was non-contributory. The physical examination was normal except for body mass index of 30.99 kg/m^2 and rare spider angiomata on her chest. Abdomen was obese, soft, non-tender without organomegaly and no definitive fluid. Her VP shunt was tapped; the pressure was elevated and sent for culture that was negative for bacterial, fungal and viral etiologies. Therapeutic and diagnostic paracentesis revealed 5.1 L of clear, yellow fluid, negative for infection or malignancy; white blood cell (WBC) 288/μL; 25% neutrophils; 16 red blood cells/μL; albumin 2.2 g/dL (serum albumin 3.3 g/dL, serum albumin ascites gradients (SAAG) 1.1); and total protein 2.9 g/dL. Serum labs revealed alanine transaminase (ALT) 10 IU/L, aspartate transaminase (AST) 18 IU/L, total bilirubin 0.4 mg/dL, alkaline phosphatase 94 IU/L, total protein 8.1 g/dL and normal complete blood count and international normalized ratio. Further workup excluded viral, autoimmune and genetic

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etiologies. A transjugular portal pressure (TJPP) measurement with liver biopsy was performed. The right atrial (RA) pressure was 8 mmHg, free hepatic vein pressure (FHVP) 13 mmHg, wedge hepatic vein pressure (WHVP) 18 mmHg and hepatic venous pressure gradient (HVPG) 5 mmHg. The liver biopsy only had six portal tracts but was reported as normal except for mild sinusoidal dilatation of uncertain significance. The elevated FHVP and WHVP suggested a post-hepatic etiology for ascites. The normal RA pressure argued against a pulmonary or cardiac etiology that was confirmed by a normal echocardiogram. Concern for an inferior vena cava (IVC) or hepatic vein partial obstruction was considered. Repeat abdominal computed tomography scan revealed an increase in ascites and no obvious vascular obstruction; therefore, a second TJPP with liver biopsy was performed. A diminutive right hepatic vein with a 90-degree angle from the IVC was noted and felt to be suboptimal for performing the pressure measurement that was used to obtain the first TJPP values. The middle hepatic vein was cannulated and all pressures were normal (FHVP, 1 mmHg; WHVP, 5 mmHg; and HVPG, 4 mmHg). The second liver biopsy was adequate (16 portal tracts) and without hepatic pathology. Neurosurgery convinced her ascites was not from a hepatic or post-hepatic etiology and the VP shunt was converted to a ventriculo-atrial (VA) shunt. Following revision, her ascites resolved. However, 2 weeks thereafter, she developed intermittent fevers and was started on broad-spectrum antibiotics. Her condition deteriorated and the serum WBC count increased to 22,000/μL despite antibiotics and repeat blood cultures that were persistently negative. However, CSF fluid from the shunt grew *Corynebacterium* species non-JK group. The shunt was removed and she was treated with intravenous vancomycin and cefepime and oral metronidazole for 21 days. One month later, a new VA shunt was placed without any further complications.

Discussion

To the best of our knowledge, this is the first case report of ascites due to malabsorption of CSF in a patient with IIH and a VP shunt infection secondary to *Corynebacterium* non-JK species. Our evaluation for a hepatic etiology was primarily due to her significant history of morbid obesity and the concern for nonalcoholic fatty liver disease (NAFLD). Cirrhosis due to NAFLD can occur in the setting of normal liver function test[6] although ascites without any other evidence for portal hypertension is unusual. The neurosurgical team strongly felt the ascites was not due to a dysfunctional VP shunt and preferred a complete exclusion of all causes before undergoing removal. In our case, the SAAG was equal to 1.1 g/dL, potentially consistent with a hepatic or post-hepatic etiology. Also, the total protein of ascites was 2.9 g/dL and levels over 2.5 g/dL help to differentiate post-hepatic from sinusoidal portal hypertension causing ascites. TJPP measurements, although initially misleading in our case, eventually provided excellent exclusion for both post-hepatic and sinusoidal portal hypertension. An elevated FHVP, WHVP and normal HVPG suggest post-hepatic portal hypertension, whereas a normal FHVP, elevated WHVP and HVPG signify sinusoidal portal hypertension. In the current case, all pressures were normal. Additionally, the transjugular approach for a liver biopsy is preferred in subjects with ascites.

Table 1. Reported cases of CSF ascites after VP shunt.

S. no.	Diagnosis	Total
1	CNS tumors	17
2	Congenital hydrocephalus	13
3	Choroid plexus papilloma	6
4	Obstructive hydrocephalus	3
5	Hyperplasia of choroid plexus	1
6	Dandy–Walker malformation	1
7	Idiopathic intracranial hypertension	1

CSF: cerebrospinal fluid; VP: ventriculo-peritoneal; CNS: central nervous system.

A VP shunt is used to decompress excessive CSF fluid accumulation within cerebral ventricles, most commonly in congenital hydrocephalus. Common abdominal complications associated with a VP shunt are intestinal obstruction, volvulus and peritonitis.[7] CSF ascites is an uncommon complication, and in a retrospective study of 300 children, it was associated with 1.3% of shunt complications.[4] Our review of the literature revealed 42 cases of documented CSF ascites: craniopharyngioma and optic nerve glioma (n = 17), congenital hydrocephalus (n = 13), choroid plexus papilloma (n = 6), obstructive hydrocephalus (n = 3) and others such as diffuse villous hyperplasia of choroid plexus and Dandy–Walker malformation[1–3,7–9] as shown in Table 1. Although the incidence of VP shunt infection is 2.1%–21% worldwide,[10,11] the incidence of CSF ascites secondary to an infection over 10 years is only 1%.[4] Infected CSF fluid causes peritoneal inflammation and enhances flow within peritoneal blood and lymphatic vessels, increases microvascular permeability and further exudation of plasma proteins, thereby impairing lymphatic reabsorption resulting in ascites.[12]

In our case, the pathophysiology for CSF ascites was initially undefined. The opening pressure was high either due to IIH itself or the ascites increasing abdominal pressure limiting CSF drainage. After conversion to a VA shunt, our patient became overtly ill and *Corynebacterium* non-JK group was cultured from the CSF fluid. Patients who have undergone surgical revision are three times more likely to develop a shunt infection.[13] Normal skin flora such as coagulase-negative *Staphylococcus* and *Staphylococcus aureus* accounts for 50% and 33% of all VP shunt infections, respectively.[5] Rare pathogens include *Candida albicans, Corynebacterium* and *Mycobacterium*.[5] *Corynebacterium* non-JK group, a gram-positive rod, commonly colonizes skin and is rarely pathogenic. However, this group has recently emerged as an important pathogen in immunocompromised patients and patients with indwelling devices/catheters and is difficult to eradicate.[14] Since 1976, only 17 cases of *Corynebacterium* VP shunt infections have been reported in the literature, 8

Table 2. Reported cases of non-JK *Corynebacterium* species shunt infections.

Case no.	Reference	Age	Sex	Procedure	Species
1	Frame and McLaurin[16]	12 years	Female	VP shunt	*Corynebacterium* sp.
2	Hande et al.[17]	11 years	Male	VP shunt	*Corynebacterium* sp.
3	Gaskin et al.[18]	26 days	Male	VP shunt	*C. xerosis*
4	Hoy et al.[19]	20 months	Male	VP shunt	*C. striatum*
5		13 months	Female	EVD	*C. striatum*
6		6 years	Female	EVD	*C. striatum*
7	Arisoy et al.[20]	5 months	Female	VP shunt	*C. xerosis*
8	Miura et al.[15]	36 years	Female	VP shunt	*Corynebacterium* sp.
9	Present	37 years	Female	VP shunt	*Corynebacterium* sp.

EVD: external ventricular drain; VP shunt: ventriculo-peritoneal shunt.

from non-JK species[15] shown in Table 2. Treatment of choice is shunt removal and intravenous antibiotics.

In summary, the present case demonstrates an unusual cause of ascites due to CSF malabsorption secondary to a rare commensal organism causing a shunt infection expanding our differential diagnosis.

Declaration of conflicting interests

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Ethical approval

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Informed consent

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