Case Report

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Abdominal Cerebrospinal Fluid (CSF) Pseudocyst: A Rare Complication of Ventriculo-Peritoneal (VP) Shunt

Shagun J Shah[1], Manavi KM[2], Milind S Tullu[3], Mukesh Agrawal[4]

Author Affiliation: [1]Assistant Professor, [2]Junior Resident II, [3]Professor (Additional), [4]Professor & Head, Department of Pediatrics, Seth G.S. Medical College & KEM Hospital, Parel, Mumbai 400012, Maharashtra, India.

Corresponding Author: Milind S Tullu, Professor (Additional), Department of Pediatrics, Seth G.S. Medical College & KEM Hospital, Parel, Mumbai 400012, Maharashtra, India.

E-mail: milindtullu@yahoo.com

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Abstract

Ventriculo-peritoneal (VP) shunt is a procedure performed for diverting the cerebrospinal fluid (CSF) flow in patients with hydrocephalus. The procedure may be associated with various complications including mechanical complications (shunt block, shunt obstruction or shunt migration) and biological complications (shunt infection). CSF pseudocyst, commonly called "CSFoma" is a rare but significant complication of VP shunt. We present a case of Crouzon syndrome (with hydrocephalus) operated for VP shunt, who developed a CSF pseudocyst within 3 months of insertion of the VP shunt. He presented with abdominal distension and the diagnosis was confirmed by ultrasonography followed by a contrast enhanced computed tomographic (CECT) scan of the abdomen. The patient underwent therapeutic fluid tapping for the abdominal distension (as it was causing respiratory distress) followed by exteriorisation of the VP shunt till the pseudocyst resolved. The VP shunt was replaced later with a ventriculo-atrial shunt. The child has been doing well with no further complications at 4 weeks of follow-up.

Keywords: Abdominal pseudocyst; Crouzon syndrome; CSFoma; Hydrocephalus; Ventriculo-peritoneal shunt.

Introduction

Ventriculo-peritoneal (VP) shunt is a common procedure performed for diverting the cerebrospinal fluid (CSF) flow in children with hydrocephalus. There can be various complications associated with this procedure; including shunt discontinuity, shunt malfunction, shunt obstruction, migration of the tip of the shunt, shunt infection, abscess or hematoma formation, intestinal perforation and abdominal pseudocyst ("CSFoma") formation.[1,2] CSFoma formation is a relatively rare complication of VP shunt, with an incidence of about 1% to 4.5% in the shunted patients.[3] Herein, we report a case of Crouzon syndrome with VP shunt in-situ, who developed the complication of a "CSFoma".

Case Report

An eight months-old-male child, a known case of Crouzon syndrome, had presented with increasing head circumference and vomiting along with signs of raised intracranial tension (ICT), three months back. A computed tomographic (CT) scan of brain done then had shown gross dilatation of all the ventricles (suggestive of communicating hydrocephalus). The child was operated for a right sided VP shunt with posterior vault repair. Three months later, the patient developed acute onset of abdominal distension (over six days), which was progressively increasing and was associated with respiratory distress (due to pressure effect). There was absence of history of fever, vomiting, excessive crying, lethargy, convulsions or new onset of focal neurological deficits. On physical examination, his vital parameters were normal,

without any signs of raised ICT. Anthropometry showed weight of 5.3 kg (z score < –3SD), length of 63 cm (z scores < –3SD) and head circumference of 43 cm (z score between –1 and –2). The VP shunt was functioning, when assessed clinically. His abdomen was grossly distended (abdominal girth of 47 cm), tense and dull to percuss. There was hypertonia (spasticity) in all 4 limbs (lower limbs were more affected than the upper limbs) with bilateral brisk deep tendon reflexes and bilateral extensor plantar response. Rest of the neurological and fundus examination was normal. The complete blood count and other routine investigations were normal. An ultrasonography of the abdomen revealed a pseudocyst with the VP shunt in-situ. A diagnostic abdominal (pseudocyst) fluid tap revealed absence of evidence of infection. A contrast enhanced computed tomographic (CECT) scan of the abdomen demonstrated a large 6.9 X 14.1 X 14.7 cm collection in the anterior peritoneum, with the VP shunt within it, suggestive of a "CSFoma" (i.e. a CSF pseudocyst) (Fig. 1). The fluid collection was displacing the transverse colon superiorly and rest of the bowel loops posteriorly. A therapeutic tap of the CSFoma was done to relieve the respiratory distress on the second day after admission. This was followed by exteriorization of the VP shunt. The draining CSF was clear with absence of growth in bacterial culture media. The child was treated with two weeks of intravenous antibiotic (ceftriaxone) and the VP shunt was later replaced by a ventricular-atrial (VA) shunt. Currently, the child is doing well with no further complications at four weeks of follow-up after the ventricular-atrial (VA) shunt.

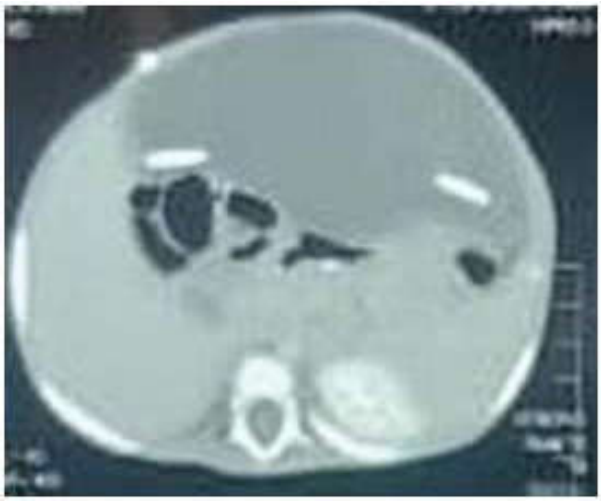

Fig. 1: Abdominal CECT showing 6.9 X 14.1 X 14.7 cm collection in the anterior peritoneum, displacing the transverse colon superiorly and rest of the bowel loops posteriorly, with the ventriculo-peritoneal shunt within it, suggestive of a CSFoma (i.e. a CSF pseudocyst).

Discussion

Crouzon syndrome, an autosomal dominant inherited cranio-synostosis syndrome may be associated with hydrocephalus due to the failure of absorption of the CSF from the subarachnoid spaces.[5] A shunt surgery may be required for the same, if the child develops signs of raised ICT (as was seen in our patient). Ventriculo-peritoneal (VP) shunt is a commonly used procedure for this purpose. Like other procedures, a VP shunt may be associated with variety of complications; with a CSF pseudocyst ("CSFoma") being an uncommon but well recognised complication. This CSF pseudocyst develops typically after months to years after the VP shunt placement surgery.[4] The CSF pseudocyst forms around the shunt tip and it is lined by fibrous tissue without an epithelial lining (hence the term 'pseudocyst" is used) with CSF within the pseudocyst. The pseudocyst can move freely within the peritoneal cavity, or it may adhere to small bowel loops, serosal surface of solid organs or the peritoneum.[6]

The mechanism of formation of a CSF abdominal pseudocyst is not very clear, but has various predisposing factors like - underlying inflammation or infection (which may frequently remain undiagnosed) affecting CSF absorption, adhesions due to previous surgeries, hypersensitivity to the shunt material and a high protein content in the CSF.[2,4] Egelhoff et al.[7] has stated that even if there are no signs of infection, presence of an abdominal pseudocyst in a patient with a VP shunt indicates possibility of an underlying infection. The most common organisms causing the infection include-staphylococcus epidermidis, staphylococcus aureus and propionibacterium acnes.[7]

The patients may present with signs of raised ICT (like headache, vomiting, convulsions, and altered sensorium) due to shunt dysfunction or with abdominal complaints. Salomao et al. studied 18 retrospective cases of CSF abdominal pseudocysts and reported various abdominal complaints like abdominal pain, abdominal mass, anorexia, constipation and subphrenic abscess.[8] Most of these cases can be diagnosed by an ultrasonography, which commonly shows a sonolucent mass (with or without septae) producing a "rail road sign" with the shunt tip in the cyst.[9] An abdominal CT scan can also be used for confirmation of the diagnosis. Both these imaging modalities can be used for diagnosing the size, position and type of cyst, along with the position of the shunt as well as for facilitating percutaneous drainage.[10]

The CSF abdominal pseudocyst can be treated using different methods like external ventricular drainage, conversion to ventriculo-atrial or ventriculo-pleural shunt, CT-guided needle aspiration, laparotomy with opening or removal of the cyst, and repositioning of the catheter tip in a different abdominal quadrant.[10] Most patients are given antibiotics after exteriorisation, prior to the shunt revision (as was done in our patient). Coley et al. reported that radiologically guided aspiration of the fluid can be done to treat the acute symptoms while waiting for the shunt surgery.[11] This process was followed in our patient to treat the respiratory distress.

Conclusion

CSF abdominal pseudocyst ("CSFoma") is an infrequent complication of a VP shunt and can be clinically suspected when the patient develops shunt dysfunction or abdominal complaints. The diagnosis can be confirmed by ultrasonography or CECT of the abdomen. Treating the infection with antibiotics and shunt revision by placing the distal tip in a new location in the peritoneum/ atrium/ pleural cavity (after the pseudocyst resolves) is the modality of treatment.

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