ORIGINAL ARTICLE

Post ventriculoperitoneal shunt abdominal pseudocyst: Challenges posed in management

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ABSTRACT

Background: In patients with hydrocephalus, the abdominal cavity has been used for absorption of cerebrospinal fluid (CSF) since 1905. Ventriculoperitoneal (VP) shunt operation is followed by abdominal complications in about 5-47% cases. Abdominal CSF pseudo cyst is an uncommon, but well described complication.

Aim: This survey was conducted to study the clinical profile and management of this entity. We present our experience with cases of CSF pseudo cyst in children.

Materials and Methods: Retrospective analysis of 4 cases diagnosed to have abdominal pseudo cyst following VP shunt between 2008 and 2013. All the four cases were suspected clinically and diagnosis was confirmed by abdominal ultrasonography.

Results: In three patients, the cyst was multilocular and of varying size. Fourth one had a unilocular cyst at the lower end of VP shunt. All the four patients had features of varying degree raised intracranial pressure and a two patients had abdominal signs also. All the patients needed open exploration. Cyst fluid was drained and partial to complete excision of the cyst was done along with the repositioning of the shunt in abdominal cavity in three patients and exteriorization of shunt in one patient. Patients were followed for any further complication over a period of 1-year.

Conclusion: Abdominal pseudo cyst is a rare complication after VP Shunt and could result in shunt malfunction or abdominal symptoms and signs. Whenever suspected it should be confirmed by imaging, followed by open exploration and repositioning of the shunt.

Key words: Abdominal pseudocyst, cerebrospinal fluid, intracranial tension, ventriculoperitoneal shunt

Introduction

Use of the peritoneal cavity for cerebrospinal fluid (CSF) absorption in ventriculoperitoneal (VP) shunting was introduced in 1905, since then VP shunting is among the most frequently performed operations for the management of hydrocephalus.[1,2] Abdominal pseudo cyst, with CSF collecting and being poorly or not absorbed across the serosa.[3,4] Intracystic pressure increases and interferes with optimal shunt function. Clinical presentation include abdominal pain with/without a palpable mass, abdominal distension with/without tenderness, nausea and/or vomiting, decreased appetite, constipation, fever and signs of shunt malfunction such as lethargy and headache. The cyst wall may evolve from an inflammatory reaction to a focal peritonitis or low-grade infection.[5,6] treatment is usually cyst excision and shunt relocation.

Materials and Methods

This was a descriptive study based on the retrospective analysis of four patients diagnosed with abdominal pseudo cyst following VP shunt between 2008 and 2013.

Access this article online	
Quick Response Code:	Website: www.asianjns.org
	DOI: 10.4103/1793-5482.145539

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How to cite this article: Hamid R, Baba AA, Bhat NA, Mufti G, Mir YA, Sajad W. Post ventriculoperitoneal shunt abdominal pseudocyst: Challenges posed in management. Asian J Neurosurg 2017;12:13-6.

Case 1

A 4-year-old male child with a history of VP shunt in the neonatal period for congenital hydrocephalus, presented with abdominal distention and vomiting. Abdominal examination revealed a moderate distension, visible bowel loops and a lump in the abdomen. Systemic examination was normal. Abdominal skiagram showed few air-fluid levels with gross ascites. Ultrasound examination of the abdomen revealed gross amount of encysted fluid with internal septations and tip of shunt catheter within it. Size of the cyst approximately was 10 × 10 cm. Laparotomy was done for intestinal obstruction, and shunt malfunction. There was a large cyst with internal septations, adherent to sigmoid colon and terminal ileum with proximal intestinal dilatation. The tip of shunt catheter was within the cyst cavity and. Near total cyst excision, adhesiolysis and repositioning of tip of shunt in right sub diaphragmatic space was done. CSF obtained from cyst and shunt was sterile. Postoperative period was uneventful. He is doing well, until date with a follow-up period of 8 months.

Case 2

Nine year old male diagnosed case of tubercular meningitis with VP shunt since 3 years, complaints of progressive abdominal distension and pain in the abdomen for 1-month. Patient was having features of raised Intra cranial tension (ICT) (altered sensorium) due to shunt malfunction. Abdominal examination revealed mild distention without any tenderness. A vague lump was also felt on palpation of the abdomen. Systemic examination was normal. X-ray abdomen showed two air-fluid levels with mild distension of gut loops. Ultrasound examination of the abdomen revealed gross amount of encysted fluid with internal septations and tip of shunt catheter within it. On exploration, cyst was roughly the size of 6 × 7cm without internal septations. Clear fluid was drained from the cyst with total excision and relocation of the shunt. CSF drained was sterile. In follow-up period at 1 year, child was doing well.

Case 3

A 8-year-old male was diagnosed as a case of congenital hydrocephalus at 1-year of age, for which VP shunt was inserted. He presented in accident and emergency department in altered sensorium with features of raised ICT. The child was admitted, base line investigations done, CSF was sterile, abdominal examination revealed the presence of a cystic lump occupying the right upper quadrant and lumbar area. Ultrasonography (USG) abdomen showed a cystic lesion in relation to the lower end of the shunt. On the exploration, a thick walled and tense pseudo cyst was seen in abdomen size of 10 × 10cm. Excision of the cyst along with the drainage of the fluid was done. Shunt was repositioned in the peritoneal cavity which started functioning optimally and patient was discharged. After 11 months, the patient was doing well.

Case 4

A 5-year-old female child had undergone a VP shunt for congenital hydrocephalus at 6 months of age. 1-year after operation she was readmitted in our department with low-grade fever, poor appetite, light headache, abdominal pain and back pain. An abdominal pseudo cyst (APC) as a complication of the VP shunt was highly suspected, which was confirmed on USG. USG showed a septated APC lesion of 7 × 8 cm in size. A distal externalization of the peritoneal end of the shunt with excision of the pseudo cyst was performed. CSF culture demonstrated a *Staphylococcus* epidermis infection and adequate antibiotic treatment was administrated. After 2 months a new VP shunt was placed, and the child is doing well.

Results

Two cases had mild to moderate abdominal symptoms and signs, which included abdominal pain, vomiting abdominal distension and lump in addition to features of raised ICT. All the patients' diagnosis was suspected clinically and confirmed on USG of the abdomen. USG was found to be a good investigation and corroborated well with intraoperative findings. USG showed internal septations in three patients. It provided useful information regarding size position and relation with solid organs in the abdomen. In the case; 1 and 2, X-ray abdomen showed mild distension of the gut with few air-fluid levels. CSF examination was sterile in three patients.

Table 1: The salient features of APC and management

Case	Age in years/sex	Duration of shunt	Indication of VP shunt	Presentation of APC	USG appearance and size of the cyst	Surgical procedure done	CSF culture
1	4/male	4 years	Congenital hydrocephalus	Pain abdomen, vomiting, abdominal distension, abdominal lump	10 cm×10 cm, septated	Excision of the cyst and the repositioning of the distal end of the shunt in the abdominal cavity	Sterile
2	9/male	3 years	Tubercular meningitis	Altered sensorium, pain abdomen and distension	6 cm×7 cm, septated	Excision of the cyst and the repositioning of the distal end of the shunt in the abdominal cavity	Sterile
3	8/male	7 years	Congenital hydrocephalus	Altered sensorium palpable lump	10 cm×10 cm	Excision of the cyst and the repositioning of the distal end of the shunt in the abdominal cavity	Sterile
4	5/female	4.5 years	Congenital hydrocephalus	Pain abdomen, poor appetite, fever	7 cm×8 cm	Temporary exteriorization	Grown *Staphylococcus aureus*

APC – Abdominal pseudocyst; CSF – Cerebrospinal fluid; USG – Ultrasonography; VP – Ventriculoperitoneal

The salient features of abdominal pseudocyst and management have been summarized in Table 1.

The time duration between insertion of the shunt and the presentation was 3 years in case; 2 and 3, 4 years in case; 1 and 1-year in case; 4. The surgical management was drainage of the cyst with partial to complete excision and the repositioning of the shunt in the abdominal cavity in three patients and exteriorization of shunt in one patient. All the patients are doing well on follow-up.

Discussion

Abdominal pseudo cyst is an uncommon manifestation of a VP shunt.[7] Several predisposing factors have been suggested, including infection, multiple shunt revisions, obstruction or dislodgement, peritoneal reaction to stranger body reject, but the pathophysiology is still unknown.[8] In most cases, the etiology of an APC is not identified, may be due to loss of absorptive capabilities for the CSF fluid because of adhesions and/or subclinical peritonitis. The infection rate has been reported to be from 17% to 80% and S. epidermis, and *Staphylococcus aureus* are the most commonly cultured micro-organisms as was demonstrated in case no 4 in our series.[9] McLaurin and Frame suggest that the presence of an APC in a patient with a VP shunt indicates shunt infection, even in the absence of clinical evidence of infection. It is possible that a low-grade infection of the shunt may be under-diagnosed with a single CSF culture or that the infection be transient or latent.[8-11]

In our series three patients had undergone VP shunt operation for congenital hydrocephalus. One patient (Case 2) had VP shunt for hydrocephalus following TBM. Formation of CSF pseudo cyst in our cases was probably due to sub clinical infection which could not be documented in three of our patients.

The clinical feature of abdominal pseudocyst was abdominal pain (three patients), Abdominal lump (two patients), features of raised intra cranial tension were present in three patients. Children mainly complained the symptoms of elevated intracranial pressure, such as headache and nausea while adults predominantly suffered local abdominal signs.[9] In our case series, the age group included was only <10 year, and the main presenting features was raised ICT and pain abdomen.

Plain radiographs are useful to rule out other causes of acute abdomen and help determine the continuity of the catheter tube. However, they are often normal[12] or as demonstrated in our three cases, do not contribute any additional valuable information. USG are a method of choice in the evaluation of the pseudo cyst and other complications at the distal end of the VP shunt.[5,12,13]

We believe computed tomography (CT) is not needed in every case as USG is sufficient to diagnose the pseudo cyst in addition to clinical examination as suggested by Rajendra *et al*.[14] USG has also the advantage being less costly and radiation free. CT should be done when infection is suspected, and diagnosis is in doubt. The cyst is surrounded by a wall of nonepithelial tissue, such as intestinal serosa and peritoneum. Histopathological evidence demonstrates the presence of inflamed serosal surfaces, fibrous tissue lined with acute and chronic inflammatory cells and granulomatous tissue with fibroblasts, collagen and inflammatory cells. It has been suggested that both the lack of epithelial lining and the presence of inflammatory cells may hinder CSF absorption.[15-18]

Surgical treatment options include repositioning the distal peritoneal catheter in a different abdominal quadrant, shunt removal, external ventricular drainage and conversion to either a ventriculoatrial (VA) or ventriculo pleural shunt system.

In 1995, Kim *et al*. first described the laparoscopic management of a CSF pseudocyst, which involved excision of a portion of the cyst and repositioning the catheter within the peritoneal cavity. Another way of treating CSF pseudocyst peritoneal cavity is simply conversion of VP shunt to ventriculo - the pleural shunt or most frequently to VA shunt.

Treatment must be individualized. The main indication of exploration was shunt malfunction in three cases and gut obstruction in one case. Open exploration takes care of both shunt malfunction as well as pressure, obstructive effects on gut as described in our case. We feel when a patient presents with pseudocyst and features of raised ICT, CSF exam should be done if it is normal cyst should be excised surgically and cyst fluid sample should be taken. as this cyst is the cause of shunt malfunction as described by Grosfeld *et al*., Raghavendra *et al*. that with CSF collecting and being poorly or not absorbed across the serosa results in increased pressure within the cyst, reducing forward pressure gradient and optimal shunt function.[3,4] Thus, cyst excision is important for optimal shunt function as was done in our cases. In three of our patients shunt was repositioned in the peritoneal cavity at the same exploration and patients did well one patient needed temporary exteriorization, followed by placement inside the peritoneum. In all cases, the pseudo cyst was dealt with open exploration. Advantages of open exploration are multiple, it confirms the diagnosis, multilocular cyst can be dealt effectively, simultaneously shunt can be either repositioned or exteriorized, adhesionolysis in case cyst is causing gut obstruction.

Conclusion

Abdominal pseudo cyst formation at distal end of VP shunt can result in both features of shunt malfunction and abdominal signs and symptoms. Post VP shunt pseudo cysts whenever suspected should be evaluated properly by imaging. Open

exploration should be performed to manage the cyst and relocate the shunt.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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