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Abdominal cerebrospinal fluid pseudocysts in patients with ventriculoperitoneal shunts: 30 years of experience*

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ABSTRACT

Aim: We evaluated the treatment outcome of the patients having cerebrospinal fluid pseudocyst following ventriculo-peritoneal shunt. **Materials and Methods:** During the period of 1975 to 2005, 392 hydrocephalic patients underwent ventriculo-peritoneal shunt, of these eight developed abdominal cerebrospinal fluid pseudocyst. The medical records regarding the etiology of hydrocephalus, age of shunting, infectious screening, therapy and follow up were evaluated. **Results:** Cerebrospinal fluid analysis was normal in all except in 4 patients who showed high level of C-reactive protein. One patient had significant abdominal symptoms as pain, vomiting and diarrhea. All were treated by cyst excision, exteriorization of shunt and antibiotic treatment. A new shunt was placed once cerebrospinal fluid cultures were negative. **Conclusions:** cyst excision, appropriate antibiotic therapy followed by new shunt placement once cerebrospinal fluid cultures are negative constitutes the required treatment for these patients with abdominal pseudocyst.

KEY WORDS: Abdominal pseudocyst, Children, Hydrocephalus, Ventriculoperitoneal shunt malfunction

INTRODUCTION

At present, ventriculoperitoneal drainage of the cerebrospinal fluid (CSF) is the preferred treatment of hydrocephalus in children. Although this is the preferential method, it is not free from complications.[1-3] Abdominal pseudocyst formation is an infrequent complication and is characterized with signs of shunt malfunction and/or abdominal symptoms.

Within the last 30 years, 392 children who had hydrocephalus with various etiologies received a VP shunt. Abdominal CSF pseudocysts developed in eight patients. Five of them had recurrences over a period of 2 months to 3 years.

The treatment consists of the excision of the cyst, external drainage and then reconstruction of the entire shunt system.

MATERIALS AND METHODS

From 1975 to 2005, 392 hydrocephalic children were treated with ventriculoperitoneal (VP) shunts at the Department of Pediatric Surgery at the University of Innsbruck. The dysfunction of the shunt system that was caused by a peritoneal CSF pseudocyst was detected in eight patients. The cysts were confirmed by abdominal ultrasound. The medical records regarding the etiology of hydrocephalus, age, infectious screening, therapy and follow-up were evaluated [Tables 1, 2]. The antibiotic therapy was performed after the culture and antibiogram of the CSF and shunt tube.

RESULTS

Eight out of 392 hydrocephalic patients developed abdominal CSF pseudocysts and were treated by laparotomy. Cyst excision, externalisation of the catheter and antibiotic therapies have been carried out.

In five children (RP, SK, AB, BR and FV), a recurrence developed over a period ranging between 2 months and 3 years. The recurrences were similarly managed by

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Table 1: Characteristics of the patients (etiology of hydrocephalus, age at shunting, age at diagnosis of the abdominal cerebrospinal fluid pseudocyst and surgical treatment)

Patient/Sex	Etiology of hydrocephalus	Age at shunting	Age at diagnosis	Surgery
RP/M	Aquaeduct-occlusion unknown etiology	10 years	19 years	exc + ext. drain.
			20 years	exc + ext. drain.
TY/F	Hydrocephalus comm. int. (after perinatal asphyxie)	2 months	4 years	exc + ext. drain.
SK/M	Congenital hydrocephalus	5 months	24 years	exc + ext. drain.
			27 years	exc + ext. drain.
AB/F	Colloidal cyst III.ventricle (premature, esoph.atr.IIIb)	16 years	29 years	exc + ext. drain.
			31 years	exc + ext. drain.
SA/M	Hydrocephalus occlussus	2 months	3 years	exc + ext. drain.
BR/M	Hydroc. intern unknown etiology	7 months	8 years	exc + ext. drain.
			8 years + 2 months	exc + ext. drain.
PW/M	Hydrocep.ext and intern unknown etiology	2 years	22 years	exc + ext. drain.
FV/F	Occipital meningocele	1 months	10 years	exc + ext. drain.
			11 years	exc + ext. drain.

exc. + ext. drain. = excision + external drainage, M - Male, F - Female, Hydrocephalus comm. Int - Hydrocephalus communicans internus, Esoph.atr.IIIb - Esophageal atresia type IIIb, Hydroc. Int. - Hydrocephalus internus, Hydroc. Ext. - Hydrocephalus externus

Table 2: Laboratory findings

Patient/Sex	Age at diagnosis	CSF analysis	Culture of distal catheter	Blood analysis
RP/M	19 years	normal	negative	CRP = 9
	20 years	normal	*Propionibacterium acnes*	normal
TY/F	4 years	normal	*Streptococcus viridans* + *Enterococcus faecalis*	normal
SK/M	24 years	normal	*Coagulase negative Staphylococcus*	CRP = 20 Leucocytes 14600
	27 years	normal	negative	normal
AB/F	29 years	normal	negative	CRP = 21
	31 years	normal	negative	normal
SA/M	3 years	normal	negative	normal
BR/M	8 years	normal	*Staphylococcus albus*	normal
	8 years + 2 months	normal	*Staphylococcus aureus*	normal
PW/M	22 years	normal	negative	normal
FV/F	10 years	normal	negative	CRP = 8
	11 years	normal	negative	normal

CRP: C- Reactive Proteins

excision, shunt externalisation and antibiotic therapy [Table 1].

The blood and CSF analysis are summarised in Table 2. The CSF cell count and the glucose concentration were in normal range in all the cases.

In four children, the blood analysis were normal whereas the other four showed elevated CRP and there was no relationship between bacterial growth and elevated CRP [Table 2].

The cyst formation was confirmed by abdominal ultrasound. Laparotomy was performed in all the cases. During laparotomy, cyst excision and externalisation of the catheter was conducted. The peritoneum and intestinal serosa localised around the tip of the peritoneal catheter were hyperaemic and oedematous in all the patients [Figure 1].

The aerobic and anaerobic culture of the tip of the peritoneal catheter revealed that in five cases, the following bacterial growths were found: *Propionibacterium acnes*, *Streptococcus viridans*, *Enterococcus faecalis*, *Staphylococcus albus*, *Staphylococcus aureus* and *Coagulase-Negative Staphylococcus* [Table 1].

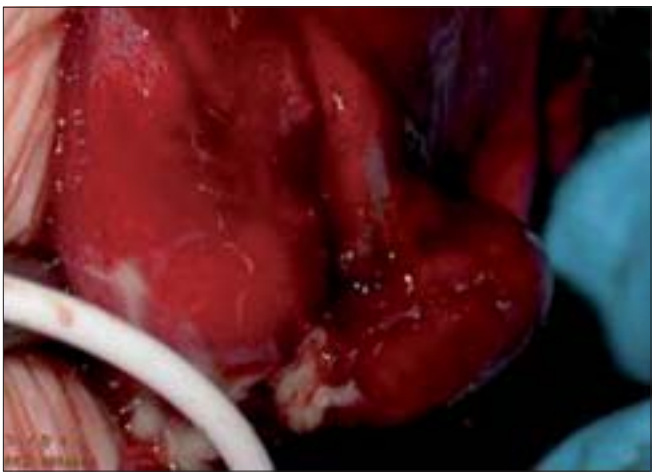

Figure 1: Inflammatory reactions around the tip of the catheter and at the pericecal region

DISCUSSION

The ventriculoperitoneal drainage of the CSF is presently the preferred treatment for hydrocephalic children; however, still there are complications. Abdominal pseudocyst formation with signs of shunt malfunction and/or abdominal symptoms is very infrequent, but they are important complications.[2-6] In the literature, the incidence of the formation of CSF pseudocyst ranged between 2 and 5%. Rainov described an incidence of 4.5%.[6] In our series, we observed an incidence of pseudocyst formation of 2.04%.

In young patients, the main symptoms are abdominal pain and the signs of elevated intracranial pressure, whereas the adult patients with a VP shunt present in the first instance with local abdominal signs.[6,7] In our patient population, we observed only a 4-year-old girl who primarily presented with abdominal symptoms such as abdominal pain and diarrhoea. The other children presented with symptoms of elevated intracranial pressure, as described in literature.

The term "pseudocyst" is used when the cyst is without epithelium and its walls consist of intestinal serosa and peritoneum.[4,7-10] As in other reported cases, the pathological examination showed chronic inflammation and a possible foreign body reaction.

There are many speculations about the etiology of the abdominal CSF pseudocyst formation; However, to date no clear reasons could be found.[5]

We assume that the "pseudocysts" are caused by the chronic irritation of the peritoneum and also a foreign body reaction could become important. The reaction of the peritoneum and the intestinal serosa is apparently noninfectious but a poorly understood inflammatory process.[6,8,10] In our patients, this inflammatory process was particularly located around the tip of the peritoneal catheter. The pseudocysts were histologically examined. Histological evaluation showed typical signs of pseudocysts with a cyst wall without epithelium and its walls comprise intestinal serosa, peritoneum and granulation tissue. In some supplements, fibrosis on the cyst wall and histiocytosis were observed.

An inflammatory reaction due to the starch granules from the surgical gloves has been discussed. There is a report with reference to silicon allergy; however, these children had recurrent skin breakdown over the shunt tract without any abdominal symptoms.[11] Since 1986, we are using latex-free gloves while inserting a VP shunt or excising pseudocysts in patients.

The cysts can be easily detected by ultrasound. In our studies, the intraabdominal cysts were located by ultrasound [Figure 2].

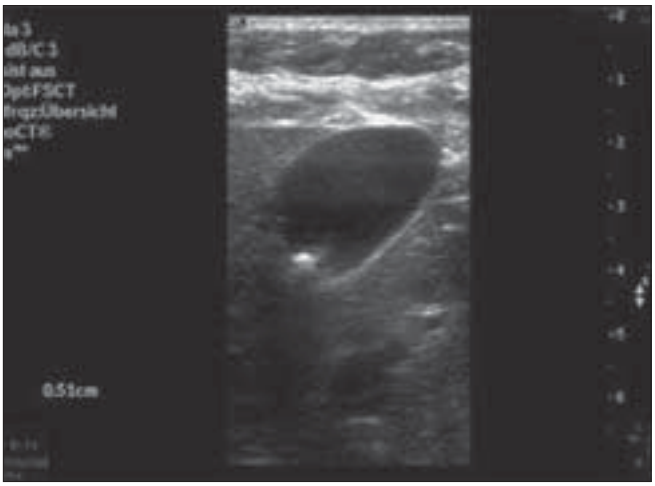

Figure 2: Sonographic view of the abdominal CSF pseudocysts

The operative management of the CSF pseudocyst comprises cyst excision and externalisation of the catheter. After appropriate antibiotic therapy, the catheter can be replaced into the peritoneal cavity; moreover, the transfer to a ventriculoatrial system is possible.[4-7,9]

Many authors accept this schedule and agree with the concept that distal catheter can be replaced in almost all cases in the peritoneal cavity after adequate treatment with the externalisation of the catheter and antibiotic therapies.[4,7-9]

The excision of the pseudocysts had been carried out in all our cases. The excision of the cyst is complicated; therefore, it is essential to operate it very meticulously. After the externalisation of the catheter, we favour to insert a VP shunt to protect the heart from an overload that could be caused by a fairly long term of the ventriculoatrial shunt system.

The literature search revealed that there are different outcomes of the excision of pseudocysts. In some studies, the recurrence is very rare whereas other authors described several cases.[4,6-8] In our study, we have a large number of relapses of pseudocysts. Five patients had recurrences over a period of 2 months to 3 years.

There are few reports in the literature that show the sterile culture of the CSF and the tip of the distal catheter.[4,7] In case of an infection, *Staphylococcus epidermidis* or *Propionibacterium acnes* could be detected.[6-8,10,12,13] In order to distinguish between an infected and a noninfected pseudocyst, it is important to be aware of the fact that *Staphylococcus epidermidis* grows slowly and requires prolonged incubation.[4,7,10]

Prolonged aerobic and anaerobic cultures should be performed to detect any organism. In our series, *Streptococcus viridans, Enterococcus faecalis, Coag. Neg. Staphylococcus, Staphylococcus albus, Staphylococcus*

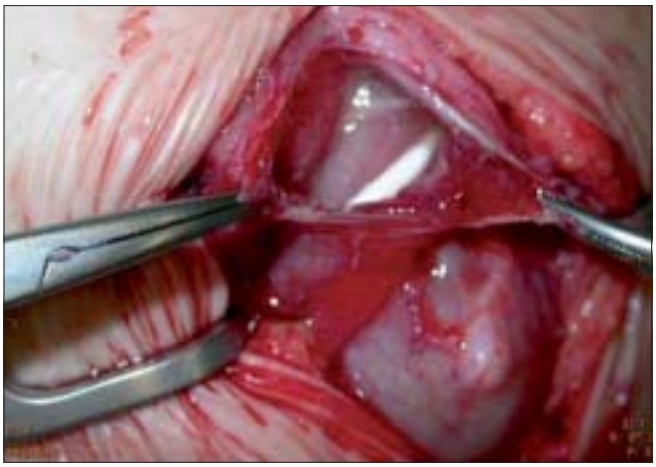

Figure 3: 4-year-old girl showed predominant abdominal symptoms; operative view of the thickened peritoneum and cyst wall and inflammatory reactions

aureus and *Propionibacterium acne*s in the distal catheter tip could be detected [Table 2].

One of our patients, a 4-year-old girl (TV), showed predominant abdominal symptoms such as high fever (39-39.5°C), abdominal pain and diarrhea. During her operation, we could observe inflammatory reactions along the abdominal catheter, and the thickening of the peritoneum could be established [Figure 3].

The hematological signs of infection such as high CRP and leucocytosis could be shown only in four cases. The relationship between the bacterial growth and a high level of CRP could not be found [Table 2].

In all cases, the spinal protein, CSF cell count and glucose concentrations were normal. The follow-up over a period of 3-7 years did not show any recurrence and no other complications were developed. In all our cases, the same operation technique were performed: cyst excision, externalisation of the catheter followed by appropriate medical antibiotic therapy over a period of of 10 to 15 days. A new VP shunt is placed after getting two sterile consecutive CSF cultures.

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