Patients with Chiari Malformation Type 2 in VP Shunt Rare Complication

Chiari Tip 2 Malformasyonu Olan Olguda Nadir Görülen VP Shunt Komplikasyonu

VP Shunt Komplikasyonu / VP Shunt Complication

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Özet

VP shuntın sebep olduğu spontan barsak perforasyonu çok nadir görülen bir komplikasyon olup, farkedilmez ise ölümcül seyreder. Bu olgumuzda iki sene önce VP shunt distal uc revizyonu yapılan ayrıca infant döneminde meningomyelosel eksizyonu olan Chiari malformasyonu Tip 2' li vakada, laparoskopik olarak tespit edilen transvers kolon splenik fleksura düzeyinde nadir görülen spontan barsak perforasyonu sonrası peritoneal kataterin anal prolapsusu sunulmuştur.

Anahtar Kelimeler

Anal Prolapsus; Chiari Malformasyonu; VP Shunt

Abstract

Spontaneous perforation of the bowel caused by VP shunt is a very rare complication, and if unnoticed will be fatal. Two years previously our case with Chiari malformation type 2 had revision of the distal end of the VP shunt and also myelomeningocele excision at the end of the infant period. The case was examined laparoscopically and found to have rare spontaneous intestinal perforation at the transverse colon splenic flexure level, followed by anal prolapse of the peritoneal catheter.

Keywords

Anal Prolapse; Chiari Malformation; VP Shunt

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Introduction

Chiari malformation was described by Hans Chiari in 1891 as translocation of posterior fossa structures into the spinal canal. After Arnold reported a child with hindbrain prolapsed as well as myelomeningocele, it was named the "Arnold-Chiari malformation". It is a congenital anomaly that may also be seen to be acquired [1].

In Chiari malformation type 2, different to type 1, the medulla and 4th ventricle have changed places toward the caudal, and this syndrome is frequently accompanied by spinal and cerebral anomalies. This syndrome may occur together with myelomeningocele, meningocele, hydrocephalus, syringomyelia and many bone anomalies [2].

Treatment of Chiari malformation type 2 is posterior fossa decompression with meningocele or myelomeningocele excision in the same session. Additionally VP shunt surgery for hydrocephalus or endoscopic third ventriculostomy may be added, depending on the case [3].

VP shunt application is one of the standard methods to treat hydrocephalus, and complications accompanying this treatment may be listed as obstruction or disconnection of the proximal or distal end of the VP shunt, malfunction of the VP shunt reservoir system, CSF accumulation, overdrainage of VP shunt and formation of slit ventricle or subdural collection, VP shunt infection, intestinal obstruction, bowel perforation, VP shunt migration, and perforation of internal organs and important veins [4]. Colon perforation is very rare, only seen in less than 0.1% of patients. The mortality rate linked to colon perforation is about 15%. The duration between VP shunt insertions and anal prolapse varies from 2 months to 7 years. Initial symptoms are meningitis following shunt infection, acute abdominal findings, seizures, fever and increased intracranial pressure findings linked to shunt failure. Colon perforation may be observed without peritonitis or acute abdominal findings [5].

Our case had myelomeningocele excision and VP shunt system inserted for hydrocephalus in the infant period. After revision of distal end of the VP shunt about 2 years previous, the case developed very rare transverse colon perforation at the splenic flexure level and anal prolapse of the distal VP shunt. Laparoscopically-aided total removal of the VP shunt system surgically and their post operative treatment is presented.

Case Report

A 14-year old patient monitored for Arnold-Chiari malformation type 2 applied to the emergency service due to abdominal pain for the previous week, with nausea and vomiting increased and a white foreign object observed in the anal region in the previous two days. The female patient was admitted for advanced examination and treatment and was taken for emergency operation after imaging tests, routine and culture laboratory tests and requested consultations. The case's infection markers (Sedim: 85/100, CRP: 35, Leukocyte: 15.000/mm3) were high and temperature was subfebrile.

The operation in addition to both cranial and peritoneal incisions would check the abdomen in the same session, aiming to visualize the bowel perforation track by laparoscopic incisions by general surgeon. With the aid of laparoscopic abdominal surgery, firstly the perforation of the transverse colon at the level of the splenic flexure was observed, and no clear abscess or pseudocyst formation was observed in the abdomen. Later the proximal ventricular catheter tip and reservoir was accessed in the cranium while the distal catheter tip was accessed in the peritoneum. The VP shunt system was fully disconnected, and the distal and proximal VP shunt parts removed as single pieces were sent for culture. As the same case had revision of the distal end of the VP shunt on the right side due to intestinal abscess and pseudocyst formation two years previously, in the same session CSF was checked for meningitis, abscess and hydrocephalus risk. For monitoring a burr hole was opened at the right frontal Kocher point and an external ventricular drainage system was provided. CSF was sent to microbiology. After primary repair of perforation laparoscopically, a drain was placed in the operation site. After hemorrhage control, the layers were anatomically sutured, as before the operation the cases lower extremities were paraplegic and she was extubated and left the operating room.

In the post operative period after requested consultations with the Infectious Diseases and Pediatric Disease departments, urine culture found Escherichia coli, blood cuture found Staphylococcus epidermidis while VP shunt catheter culture found Pseudomonas aeruginosa. According to antibiogram results intravenous combined antibiotherapy (vancomycin+ meropenem +cephazolin) was completed in 15 days. After removing sutures and when CSF culture produced no microbiology the case was discharged. (Figure 1,2,3,4).



Figure 1. Pre operative abdominal X-Ray

Discussion

Bowel perforation by the peritoneal catheter of VP shunt is a very rare situation. The first case was recorded in 1996. Perforation may occur in any segment of the gastrointestinal system; generally the colon is the most frequently perforated region. The majority of cases are asymptomatic, and the catheter may

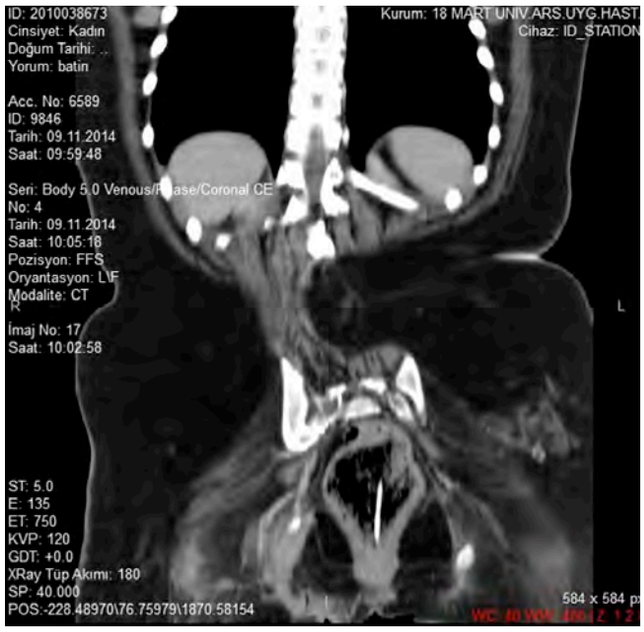


Figure 2. Pre operative abdominal CT



Figure 3. Pre operative abdominal CT

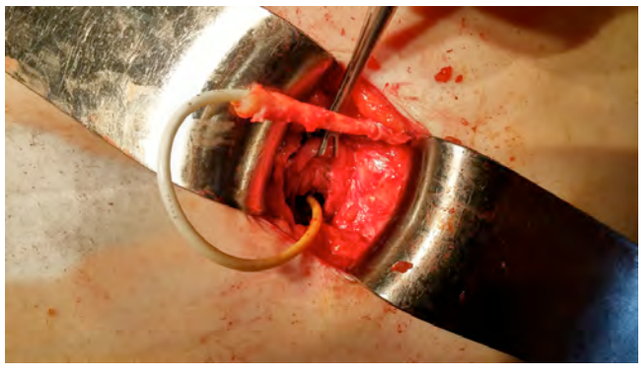


Figure 4. Appearance of distal end of VP shunt from the perforated colon

protrude from the anus. After perforation the mortality rate is relatively high at 15-19% [6].

The etiology of bowel perforation is not fully clear. It is thought that local inflammatory reactions and fibrosis around the distal catheter may cause it to adhere to an area of the bowel, and puncture the bowel with a stabbing effect [7].

It is known that previous abdominal surgery and adhesions may cause the catheter to be caught in a certain region, and form a risk of corroding the colon wall. Additionally myelomeningocele and congenital hydrocephalus patients are more prone to perforation due to weakened intestinal walls linked to weakened innervation.

Protrusion of the catheter from the anus makes diagnosis easy. Apart from this, for patients with abdominal complaints, meningitis, infection findings on the skin along the catheter pathway, and catheter failure, internal organ perforation should be considered.

When there is intestinal perforation linked to VP shunt, nausea, vomiting, peritoneal findings and intraabdominal abscess may develop. After perforation severe peritonitis develops. Patients with VP shunt and meningitis or ventriculitis caused by enteric microorganisms should be investigated for intestinal perforation.

There are three ways to remove VP shunt; through the anus, endoscopically and surgically. Using laparoscopy, a minimally invasive surgical method, allows diagnostic laparoscopy and correction of complications linked to the catheter.

After the VP shunt catheter is removed, external ventricular drainage and antibiotic treatment is very important for the case, with repeated use of the peritoneum one of the controversial topics [8].

In conclusion, our case had definite diagnosis as the distal catheter of the VP shunt protruded from the anus and was immediately taken for surgery. Laparoscopically the site of perforation was identified and primary repair undertaken. The VP shunt system was completely disconnected with the distal part removed through the anus. To allow monitoring of CSF, in the same session external ventricular drainage was given and according to the results of catheter, urine, blood and CSF cultures and antibiograms, combined intravenous antibiotic treatment was administered. On the fifth day the external drainage system was removed and on the fifteenth day CSF microbiology, biochemistry and infection markers were normal. All sutures were removed and the case was discharged with general situation stable. We believe these types of cases require a multidisciplinary approach.

Competing interests

The authors declare that they have no competing interests.

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