CASE REPORT

Subclinical meningoventriculitis as a cause of obstructive hydrocephalus

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SUMMARY

Communicating hydrocephalus may complicate infantile bacterial meningitis, typically presenting with systemic features of infection. We report a rare case of 'subclinical meningoventriculitis' causing obstructive hydrocephalus and its challenging management. A healthy 10-week-old immunocompetent male patient presented with failure to thrive and vomiting, secondary to presumed gastro-oesophageal reflux. The child was neurologically alert, afebrile with normal inflammatory markers. Progressive macrocephaly prompted an MRI confirming triventricular hydrocephalus secondary to aqueductal stenosis. An endoscopic third ventriculostomy was performed however abandoned intraoperatively due to the unexpected finding of intraventricular purulent cerebrospinal fluid. A 6-week course of intravenous ceftriaxone was commenced for *Escherichia coli* meningoventriculitis. However, the child was readmitted 18 days postoperatively with acute hydrocephalus requiring a ventricular washout and staged ventriculoperitoneal shunt insertion at 4 weeks. Serial head circumference measurements are paramount in the assessment of a paediatric patient. In an immunocompetent child, a subclinical fibropurulent meningoventriculitis can result in several management challenges.

BACKGROUND

Hydrocephalus is a pathological excess of cerebrospinal fluid (CSF) resulting in intracranial ventriculomegaly. It is typically classified as either communicating or obstructive/non-communicating and congenital or acquired.¹ The diagnosis can be made prenatally, perinatally and postnatally. Prenatal causes including intrauterine infections have been reported as the most common cause in term infants² with intraventricular haemorrhage the most common cause in preterm infants.^{3,4}

Hydrocephalus, classically the communicating type, is a well-recognised complication of bacterial meningitis in infancy. Although the presenting symptoms and clinical signs of infection can occasionally be subtle in early infancy (eg, lethargy, irritability, poor feeding, rash and diarrhoea), meningitis severe enough to cause hydrocephalus is usually clinically recognisable as infection with associated systemic features of sepsis. Hydrocephalus secondary to bacterial meningitis was observed in 5% of term infants² and 7% of preterm infants in a European series^{3,4} and must be promptly diagnosed and treated to prevent serious complications.

However, we present an atypical encounter: a rare subclinical presentation of intracranial infection which can have serious consequences if undetected. The unexpected initial intraoperative finding led to challenging neurosurgical management. The case is unique and important for the following reasons:

- ▶ The child never clinically behaved in a meningitic or septic manner despite being immunocompetent and having intraventricular pus. Neither was there any biochemical nor radiological evidence of intracranial sepsis.
- ▶ Meningitis classically causes a communicating hydrocephalus—not an obstructive one, which was unusually the case here and adds to the atypical nature of the presentation.
- ▶ The neurosurgical management of patients with intracranial sepsis causing hydrocephalus is not straightforward. The options of external ventricular drainage (EVD) and shunt surgery including the timing of this are discussed.

CASE PRESENTATION

A 10-week-old male patient was seen in the paediatric clinic with failure to thrive and vomiting, thought to be due to gastro-oesophageal reflux for which he was receiving alginate acid. Five days prior to presentation, the child's urine had been noted to be foul smelling and a 5-day course of amoxicillin was prescribed by the general practitioner for a presumed urinary tract infection (UTI). The child's medical history was unremarkable including a normal birth history, pregnancy and delivery. A maternal perineal abscess was diagnosed 4 days post partum and successfully treated conservatively with oral co-amoxiclav.

Clinical examination confirmed failure to thrive and revealed an increase in head circumference from 50th at birth to the 91st centile at 10 weeks. The child was alert with soft fontanelles, moving all four limbs spontaneously, afebrile and bedside observations were within normal limits. Cardiorespiratory and abdominal examinations were unremarkable. There were no other abnormal clinical findings or signs of raised intracranial pressure.

INVESTIGATIONS

The blood tests on admission including inflammatory markers were not indicative of infection (haemoglobin 9.8 g/dL, white cell count (WCC) $8.98 \times 10^9/L$, platelets $408 \times 10^9/L$, sodium

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Figure 1 MRI brain demonstrating triventricular hydrocephalus.

137 mmol/L, potassium 4.4 mmol/L, urea 1.6 mmol/L, creatinine 12 µmol/L, prothrombin time 11.9 s, activated partial thromboplastin time 29.2 s, C-reactive protein 9 mg/dL).

A cranial ultrasound and subsequently MRI of the brain confirmed cerebral aqueduct obstruction causing third and lateral ventricle dilatation but not fourth ventricular dilatation (figure 1).

To surgically treat the hydrocephalus with imaging suggestive of underlying aqueductal obstruction, an endoscopic third ventriculostomy (ETV) was performed 4 days after presentation, during which time there had been no additional clinical concern. The procedure was abandoned due to the unexpected finding of purulent CSF preventing safe navigation of the ventricles (figure 2). CSF sampling demonstrated Gram-negative rods on microscopy, subsequently confirmed as *Escherichia coli*. Intravenous ceftriaxone was commenced via a peripherally inserted central catheter (PICC) for a planned total 6-week course of antibiotics. The child was investigated thoroughly for a source of infection including an ultrasound of the kidneys and MRI of the abdomen and pelvis which revealed no abnormalities. Given the unusual presentation of infection, an extensive immunological panel of investigations was undertaken which demonstrated no immunological abnormalities (serum complement levels,

electrophoresis, immunofixation, lymphocyte, T cell, CD4, CD8 and CD19 B cell differentials all within normal limits).

DIFFERENTIAL DIAGNOSIS

Cerebral aqueductal stenosis:

- ▶ Congenital—web (the most common), gliosis
- ▶ Acquired
 - Extrinsic—lesions of tectal plate/pineal gland/posterior fossa or a vascular malformation
 - Intrinsic—subarachnoid haemorrhage, infection

TREATMENT

Intravenous ceftriaxone was commenced via a PICC for a planned total 6-week course of antibiotics for *E. coli* meningoventriculitis. Eighteen days post-surgery the infant was admitted with vomiting, drowsiness and increasing head circumference. Ventricular tap demonstrated a high opening pressure (20 cmH₂O). A ventricular washout was carried out but ETV could again not be performed because of scarring preventing a good view of the floor of the third ventricle. Definitive CSF diversion in the form of a ventriculoperitoneal shunt (VPS) was inserted 4 weeks after initial surgery and after multiple negative CSF cultures post-antibiotic therapy.

OUTCOME AND FOLLOW-UP

Following completion of treatment, the child suffered from several further UTIs. A micturating cystourethrogram was performed to investigate this and demonstrated bilateral vesicoureteric reflux. This was treated with bilateral STING (subureteral Teflon injection) and there have been no further recurrences of UTI.

DISCUSSION

Infantile hydrocephalus may be due to prenatal congenital factors such as aqueductal stenosis or perinatal/acquired causes such as infection and haemorrhage. Although acute bacterial meningitis causing obstructive rather than communicating hydrocephalus has been very rarely noted,⁵ the two cases (one of whom died) were acutely unwell with features of sepsis and acutely raised intracranial pressure—unlike the subclinical aseptic presentation of our infant.

Hydrocephalus is termed 'obstructive' if the CSF blockage is at the level of the foramina of Munro, third ventricle, cerebral aqueduct (of Sylvius), fourth ventricle or foramina of Luschka and Magendie. It is termed 'communicating' if the site of obstruction is at the arachnoid villi. Classically, bacterial invasion leads to neutrophil migration into the subarachnoid space with purulent exudate accumulating in Rolandic and Sylvian fissures and basal cisterns where the subarachnoid spaces are deepest with sluggish flow. This exudate then impairs absorption at the arachnoid villi causing communicating hydrocephalus. Typically, an obstructive picture is noted to occur at the end of week 2 when neutrophils degenerate and fibroblasts proliferate in the exudate.⁵ One would expect this and leptomeningeal involvement to be a highly immunogenic process.

We propose that in our patient a subclinical meningoventriculitis syndrome caused an inflammatory aqueductal obstruction secondary to webbing with further development of intraventricular septa. Ventriculitis may result from haematogenous spread of infection via glycogen-rich choroid plexus, which may facilitate local bacterial growth and act as a bacterial reservoir exhibiting relative antimicrobial resistance.⁶ This undoubtedly adds to the difficulties in treatment—achieving adequate penetration

Figure 2 Intraoperative fibropurulent exudate during attempted endoscopic third ventriculostomy.

and therapeutic levels of antimicrobial therapy. Meningoventriculitis secondary to *E. coli* infection, described as the main cause of purulent neonatal meningitis,⁷ often involves a fibropurulent inflammatory process. Similarly, a fibrinous exudate was noted in our case intraoperatively. This may accumulate without causing acute rises in intracranial pressure if the fontanelles are open.

In essence, the undiagnosed subclinical meningoventriculitis led to obstructive hydrocephalus with progressive macrocephaly. In retrospect, perinatal risk factors for sepsis could have been the vertical transmission of pathogens from the maternal perineal abscess and/or the presence of bilateral vesicoureteric reflux causing recurrent UTIs—both of which are commonly caused by coliforms.

A literature review highlights one series of ‘unrecognised meningoventriculitis’ presenting with hydrocephalus.⁸ This retrospective study from India considered 72 paediatric patients from 1991 to 1998. Thirteen patients had hydrocephalus associated with meningitis or ventriculitis of which 10 patients presented early with increasing head circumference. All 13 patients had risk factors (including being preterm, low birth weight, multiparty, signs of neonatal sepsis, prolonged neonatal hospitalisation and antibiotic use in the perinatal period) with 10 patients requiring postnatal antibiotics. Nine patients showed evidence of aqueductal stenosis on CT imaging. Six patients were reported as being clinically well at the time of presentation but all had significant risk factors—unlike our case which involved a term immunocompetent infant. Furthermore, in a population with relatively reduced access to screening, primary/secondary healthcare and treatment compared with the UK, it is conceivable that there may have been factors contributing to delayed presentations and partially treated infections which may confound the true incidence of subclinical meningoventriculitis in this retrospective series.

The management of these patients with subclinical meningoventriculitis presenting with hydrocephalus is not straightforward. Firstly, a high level of clinical suspicion is required to diagnose meningoventriculitis. The presence of CSF infection in our case was only discovered due to the unexpected purulent appearance of the CSF during endoscopic intervention subsequently confirmed with microscopic culture as *E. coli*. If a VPS had been inserted as first-line treatment of the obstructive hydrocephalus, the appearance of the CSF may not have been noted. Thus, the shunt would have been inserted into an infected system leading to shunt infection and likely failure. It is therefore imperative to send a CSF sample at the time of shunt insertion to exclude occult subclinical meningitis.

A course of appropriate antibiotics is essential to treat the underlying infection. In the Indian study,⁸ infants were treated with antibiotics for a mean of 32.8 days. CSF diversion is often required. This can be in the form of an EVD as a temporising measure or via a VPS as a long-term solution. Four of the six subclinically presenting patients in this study received EVDs. They later underwent VPS insertion 1–5 weeks postadmission or

repeat ventricular taps via the fontanelle to drain excess CSF. The latter option was used in our case as the patient decompensated at a very slow rate after each tap and did not require continuous CSF drainage to remain clinically well. The main advantage of this option is the decreased risk of superimposed central nervous system (CNS) infection from an EVD. The timing of definitive CSF diversion via a VPS is also important and is a balance between the risks of continuous external CSF drainage and the risks of inserting a shunt before the CNS infection has been fully treated. In our experience, serial negative CSF cultures and decreasing CSF WCCs are essential to make this judgement.

Learning points

- ▶ Clinicians should remain vigilant when considering the aetiology of obstructive hydrocephalus.
- ▶ Serial head circumference measurements are always clinically relevant when assessing a paediatric patient—especially in the face of diagnostic uncertainty—to identify progressive macrocephaly.
- ▶ Immunocompetent patients can present with subclinical meningoventriculitis resulting in obstructive hydrocephalus—the indolent yet fibropurulent pathophysiology of which presents several management challenges.
- ▶ A high level of clinical suspicion is always required to diagnose meningoventriculitis in such patients.
- ▶ It is imperative to send a cerebrospinal fluid sample at the time of shunt insertion to exclude occult subclinical meningitis.

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