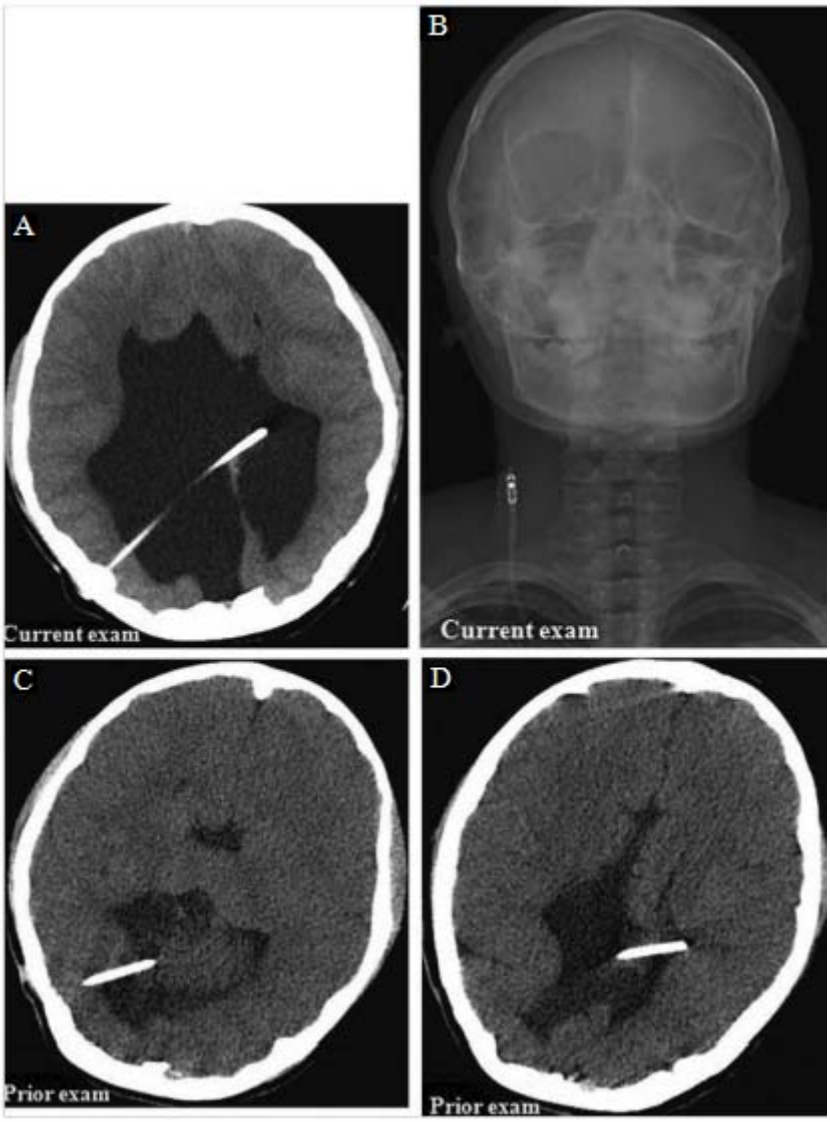
EANS/UEMS European examination in neurosurgery

Variants of questions with answers (compilation - Vyacheslav S. Botev, Department of Neurology and Neurosurgery, Ivano-Frankivsk National Medical University)

CASE HISTORIES IN SHUNT MALFUNCTIONS

Case 1

A 12-year-old boy with altered mental status and a history of Chiari II malfomation with ventriculoperitoneal (VP) shunting.



Questions

1. What are the findings on the CT scan, skull and neck x-ray?
2. What is the diagnosis?
3. What are the etiologies of slit ventricle syndrome?

Case 2

A 3-month-old child with a L5-level myelomeningocele and a ventriculoperitoneal (VP) shunt comes to the emergency department with 24 hours of progressive irritability and fever. The child had the spinal defect closed at 2 days of life and a VP shunt inserted for progressive macrocrania and hydrocephalus at 14 days of life. He has been on a program of home intermittent catheterizations since birth and no prophylactic antibiotics. The exam demonstrates a child with a fever of 39.5°C, bulging fontanel, somnolent, and irritable. The motor examination is unchanged with no plantar flexion in the feet, but otherwise normal. The child's incisions all look well healed. White blood cell (WBC) count is elevated, and the urinalysis shows positive bacteria and WBCs.

Questions

1. What is the differential diagnosis suggested by this child's presentation?
2. What investigations are appropriate and why?

Blood cultures are collected and urine cultures are sent. A computed tomography (CT) scan is done, which shows a stable ventricle size compared with the last scan done after the shunt was inserted. Shunt tap reveals 1500 WBCs and 10 red blood cells (RBCs) with no bacteria seen on a gram stain.

3. What is the diagnosis?
4. What are the usual organisms involved?
5. What are the treatment options for this child?
6. What antibiotic regimen would you choose?
7. What is the incidence of shunt infection after an initial procedure?
8. What is the time frame over which these infections usually develop?
9. What is the incidence of shunt infection after an initial shunt infection?
10. What maneuvers have shown benefit in reducing shunt infection?

Case 3

A district general hospital refers a 30-year-old woman who presented to the emergency department following three generalized tonic–clonic seizures. She has a VP shunt which was inserted following traumatic intracranial haemorrhage at 15 months of age. She has had numerous shunt revisions, the last of which was 3 months ago. For the past 2 days, her family noted that she was quiet and not her usual self, although she herself did not complain of any symptoms. On arrival at the local emergency department she was post-ictal and drowsy but responsive with a GCS of 12/15 (E3, V3, M6).

During assessment she deteriorates to a GCS of 3/15 and is intubated. Her pupils are size 3, equal and reactive. Her CT scan is shown below.

Questions

1. Describe the appearances on the scan. What would be your advice over the telephone?

2. A scan from a recent hospital admission (during which the shunt was revised) is shown. Comment on the appearances and its relevance to the current presentation.

3. The patient arrives at your hospital intubated and ventilated. Where will you direct the patient and what will your immediate actions be?

4. At surgery, the shunt tubing is found to be patent throughout and the CSF pressure is low. A sample of CSF was sent to the microbiology laboratory intraoperatively, and the following results were obtained: red cell count, 167; white cell count, 1035 (92 % polymorphs); Gram stain, Gram-negative rods. What is the management?

5. List the routes by which internal ventricular catheters may become infected. What is the difference between meningitis and ventriculitis and which affects patients with infected shunts?

6. Are there any evidence based guidelines for the treatment of shunt-associated ventriculitis/meningitis?

Answers

Case 1

1. Unenhanced axial CT image (A) demonstrates significant ventricular enlargement compared with the baseline examination (C, D). A right posterior approach ventricular drainage catheter is identified. Frontal radiograph of the skull and neck from a shunt series (B) shown discontinuity of the VP shunt within the soft tissues of the neck on the right.

2. Shunt catheter malfunction.

VP shunting remains the most common treatment for chronic hydrocephalus. VP shunts consist of the intraventricular drainage catheter, an extracranial valve and reservoir, and a catheter that extends throughout the superficial soft tissues and into the peritoneum.

Shunt malfunctions are unfortunately common in chronically shunted patients. Malfunction may be mechanical (most common) or due to an underlying infection. Common presenting symptoms indude headache, irritability, lethargy, nausea, and vomiting. Additional symptoms include fever and localizing pain. Young children with open sutures often present with increasing head size. Clinically, slowed filling of the shunt reservoir is suggestive of malfunction.

Mechanical failure may result from obstruction of the shunt catheter, disconnections or breaks in the catheter tubing system, and migration. Obstrcution is most common at the intraventricular end of the shunt catheter and may be caused by proteinaceous material (hemorrhage, infection, tumor), brain parenchyma, or choroid plexus obstructing the inflow of CSF.

Distal obstruction at the peritoneal end is less common and typically results from peritoneal adhesions near the catheter tip. Occasionally, an intraventricular cyst or abdominal pseudocyst may obstruct CSF flow. Nuclear medicine radioisotope or contrast-enhanced computed tomography (CT) or fluoroscopic shunt studies can be used to evaluate for obstruction within the VP shunt system.

Disconnection of shunt components most often occurs where catheter tubing connects to the reservoir. Breaks occur in regions of increased mobility, such as the neck. Migration typically affects the proximal or distal end of the VP shunt catheter. When interpreting a plain film shunt study, it is important to know the type of catheter used, particularly with respect to the portions that are normally radio-opaque and radiolucent, to avoid mistaking normal radiolucent portions of the tubing as regions of discontinuity. If needed, cross-sectional imaging can be used for problem solving. A focal fluid collection may be identified at the site of malfunction.

CT or magnetic resonance imaging (MRI) (preferred) of the brain most often demonstrates ventricular enlargement with shunt malfunction; however, this finding is not entirely sensitive or specific. False-negatives occur with scarring along the ventricles or decreased parenchymal compliance. Conversely, ventricular enlargement may be seen with normally functioning shunts. Secondary findings of shunt malfunction incldude sulcal and cisternal effacement and transependymal flow of CSF, which suggest uncompensated, acute hydrocephalus.

Shunt infections most often occur within the first few months of catheter placement. Patients present with signs of shunt malfunction, as well as fever. Imaging may be normal or demonstrate findings associated with ventriculitis or meningitis, to incdude abnormal T2/fluid-attenuated inversion recovery signal intensity and enhancement along the margins of the ventricles and meninges. Replacement of the infected shunt is often necessary.

3. Complications of VP shunting also include slit ventricle syndrome and subdural fluid collections. Etiologies of slit ventricle syndrome include overshunting, fibrosis, decreased parenchymal compliance, and intracranial hypotension. Overdrainage may result in subdural hematomas or hygromas. Typically, subdural collections are self-limited.

Case 2

1. What is the differential diagnosis suggested by this child's presentation?
• This child is presenting with the clinical signs of an infectious illness, and clinical signs of shunt malfunction, though the fever itself may be causing the child's irritability independent of shunt malfunction, and a severely irritable child may present with a bulging fontanel.
• The cause of the febrile illness may be viral or respiratory, but in this case, we would be worried about shunt infection or wound complication, as well as urosepsis.

2. What investigations are appropriate and why?
• This child needs cultures of blood, urine, a complete blood count, chest x-ray as well as viral swabs of an upper respiratory tract-related secretions and a full exam to look for skin abrasions, wound complications, or other pertinent physical findings.
• CT scan is indicated due to the clinical signs of increased intracranial pressure (ICP).
• A tap of the shunt reservoir to obtain cerebrospinal fluid (CSF) is clearly indicated in this child given the presentation and relatively recent shunt surgery.

3. What is the diagnosis?

• The cell count of the CSF points toward shunt infection even in the absence of an initially positive gram stain.

• The culture of CSF will probably grow eventually.

4. What are the usual organisms involved?

• Shunt infections are usually from skin colonization occurring at the time of surgery – gram-positive cocci are most common.

• In a child of this age, and especially one with urinary catheterizations, gram-negatives and coliforms are also possible.

5. What are the treatment options for this child?

• This child needs antibiotics and some type of shunt externalization procedure.
• Very few shunt infections will respond to antibiotics alone without hardware removal.
• Many centers will externalize the shunt by removing the distal catheter from the abdomen and then treat until the CSF is sterile before replacing the entire system.
• In the event of continued positive cultures, the entire shunt system should be converted to an external ventricular drain (EVD).
• Some centers would externalize the EVD upfront with removal of the whole shunt system.
• The duration of antibiotic treatment before reinternalization is debatable; it averages 10 to 14 days, but three consecutive negative CSF cultures is a common standard.

6. What antibiotic regimen would you choose?

• Initial antibiotic regimen needs to include broad spectrum CSF penetrating coverage, and good antistaphylococcal coverage until the organism is known.
• Typical initial regimens would include a thirdgeneration cephalosporin with vancomycin plus or minus an aminoglycoside.

7. What is the incidence of shunt infection after an initial procedure?

• Shunt infection rates per procedure after an initial shunt placement are approximately 8 to 10% in most large studies of shunt insertions in children.

8. What is the time frame over which these infections usually develop?

• Most shunt infections are procedure-related and present within the first 6 months of surgery.
• Other risk factors for shunt-related infection includes wound breakdown and CSF leak.

9. What is the incidence of shunt infection after an initial shunt infection?

• Shunt infections may occur up to 25% of the time when a shunt is replaced after an initial infection.

10. What maneuvers have shown benefit in reducing shunt infection?
• There are a great many studies attempting to demonstrate protocols to reduce shunt infection rates.
• Efforts to reduce shunt infection rates have includes perioperative antibiotics, various procedure-related technical issues such as double gloving, short duration of surgery, surgery timing, reduced mechanical manipulation of the hardware, and more recently antibiotic-impregnated shunt catheters.
• Many of these techniques including antibiotic-impregnated catheters are still the subject of some debate as to their relative efficacy.

Case 3

1. Describe the appearances on the scan. What would be your advice over the telephone?

The third ventricle and the occipital horns of the lateral ventricles are dilated. A ventricular catheter traverses the right lateral ventricle and lies in the third ventricle. Some low density, which may represent transependymal flow and raised intracranial pressure, is seen around the frontal horns.
Clinical deterioration, ventriculomegaly, and transependymal flow are consistent with a blocked shunt. This patient requires urgent transfer to the neurosurgical department for evaluation. The local clinicians may be asked to aspirate some fluid from the shunt or perform an LP if there is likely to be a delay in transfer. If available, previous imaging should be consulted to see if there has been any interval change in the size of the ventricles. Blood tests should be processed at the local hospital so that the patient can proceed directly to theatre on arrival with reference to those results.

2. A scan from a recent hospital admission (during which the shunt was revised) is shown. Comment on the appearances and its relevance to the current presentation.

The ventricles are larger on the previous scan. A shunt catheter is seen with its tip in the right frontal horn of the lateral ventricle. There is more transependymal flow compared with the current scan. In other words, the current scan looks ‘better’ than the previous one. In this situation it is important to establish whether the old scan was performed before or after a shunt revision. In any case, a current scan that looks ‘better’ than a previous scan does not necessarily

exclude shunt blockage because there may have been initial improvement followed by deterioration.

A blocked shunt must always be suspected (regardless of radiological appearances) if there has been clinical deterioration.
Seizures or coma from a metabolic cause are also a possibility and should be considered, although priority should be given to exploring the shunt. Since a shunt revision operation has been performed only 3 weeks ago, infection needs to be excluded as shunt infections are more common in the postsurgical period, with their likelihood decreasing progressively with time since last revision.

3. The patient arrives at your hospital intubated and ventilated. Where will you direct the patient and what will your immediate actions be?

The patient should be transferred to the operating theatre immediately as an operation can rapidly and simultaneously address diagnosis (blocked shunt or infection) and management (revise shunt if blocked or remove shunt if infected). If the shunt is found to be patent and the CSF sterile, another cause for the neurological deterioration must be sought. The pupillary responses should be checked on a regular basis, in this case on the patient's arrival in the department and before surgery.

4. At surgery, the shunt tubing is found to be patent throughout and the CSF pressure is low. A sample of CSF was sent to the microbiology laboratory intraoperatively, and the following results were obtained: red cell count, 167; white cell count, 1035 (92% polymorphs); Gram stain, Gram negative rods. What is the management?

These findings are consistent with bacterial infection. The management for a shunt-associated infection is as follows.

Remove the entire shunt system if feasible (if the patient is not fit for surgery, antibiotics may be used with the shunt left in situ, but this is associated with a worse outcome).

Provide interim CSF drainage (e.g. with an external ventricular drain). This enables continuous drainage of CSF, CSF sampling to monitor the response of infection to antibiotic therapy, and administration of intrathecal antibiotics if necessary.

Identify the source of infection.

Commence intravenous antibiotics.

Re-insert the shunt when the CSF is sterile.

5. List the routes by which internal ventricular catheters may become infected. What is the difference between meningitis and ventriculitis and which affects patients with infected shunts?

Contamination of the shunt by the patient's skin flora at the time of insertion (most common)

Breakage of the skin overlying the shunt

Contamination of the distal end of the shunt (e.g. peritonitis for ventriculoperitoneal shunts)

Hematogenous.

By definition, meningitis involves inflammation of the meninges, whereas ventriculitis involves inflammation of the ventricular ependyma. Patients with an infected shunt or external ventricular drainage device initially develop ventriculitis. They may or may not develop meningitis depending on the extent of residual communication between the ventricular system and the subarachnoid space.

6. Are there any evidence-based guidelines for the treatment of shunt-associated ventriculitis/meningitis?

Better outcomes are achieved by removing the shunt system rather than leaving it in situ. There are no evidence-based guidelines for the type and duration of antimicrobial therapy, although recommendations have been published. Intravenous antibiotics should generally be continued for 7–14 days and negative CSF cultures should be obtained throughout this period before the shunt is re-inserted. Intrathecal antibiotics may be considered in resistant cases or when the shunt apparatus cannot be removed.

Pearls

• Shunt malfunction is common in chronically shunted patients and may be mechanical or related to infection.

• It is important to know the type of catheter used and which portions are normally radio-opaque and radiolucent

• CT or MRI (preferred) most often demonstrate ventricular enlargement in the setting of shunt failure.

• Overshunting may result in slit ventricle syndrome and subdural hematomas or hygromas.