CASE REPORT

Subacute bacterial endocarditis and subsequent shunt nephritis from ventriculoatrial shunting 14 years after shunt implantation

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Accepted 30 May 2014

SUMMARY

Fourteen years after shunt implantation, a 26-year-old patient with myelomeningocele, concomitant hydrocephalus and a ventriculoatrial cerebrospinal fluid (CSF) shunt presented with brief but recurrent episodes of fever predominantly when taking showers or during physical exertion. After 4 years of inconclusive multidisciplinary investigations, the patient progressed into end-stage renal disease before an echocardiogram revealed a vegetative plaque on the tendinous chords of the tricuspid valve. CSF cultures were grown from the shunt valve, confirming bacterial growth of *Propionibacterium acnes* suspected of causing subacute bacterial endocarditis and subsequent shunt nephritis. The patient was successfully treated with antibiotics combined with ventriculoatrial shunt removal and endoscopic third ventriculocisternostomy (VCS). This case illustrates the nowadays rare, but potentially severe complication of subacute bacterial endocarditis and shunt nephritis. It also exemplifies the VCS as an alternative to implanting foreign shunt systems for CSF diversion.

BACKGROUND

Ventriculoatrial (VA) and ventriculoperitoneal (VP) shunts are used in order to treat hydrocephalus by diverting the cerebrospinal fluid (CSF). Infection rates are similar between VP and VA shunts, but VA shunts that divert CSF into the right atrium of the heart carry the risk of spreading bacteria from a potential shunt infection directly into the blood stream.¹ Accordingly, VP shunts widely replaced VA shunts as the latter was associated with severe complications such as septic infections, renal failure and cardiorespiratory complications.^{2,3}

VA shunt infections often occur during the first month after implantation, with infection rates of around 2.5–12% being reported.^{1,4} The presentation of such infections is often non-specific, with fever, lethargy, irritability and anorexia being common symptoms. A late complication is shunt nephritis, where a chronic low level infection of a VA shunt causes an immune-complex deposition in the renal glomeruli and subsequently glomerulonephritis.⁵ Even though VA shunts are quite rare, an infection may become severe and therefore it is important to quickly diagnose and treat these patients. We present a case of VA shunt infection with a particularly insidious course and severely delayed diagnosis, but we also describe how this patient was successfully treated.

CASE PRESENTATION

The patient, a woman with myelomeningocele, paraparesis, Chiari type II malformation and hydrocephalus had been treated with CSF diversion since infancy. She had undergone a total of nine shunt revisions during her childhood, either due to shunt malfunctions or shunt infections. The last revision was performed at the age of 12, when she received a VA shunt due to repeated failures of the peritoneal catheter of her VP shunt. As a young adult, the patient moved to the city that is served by our neurosurgical centre. Her medical condition was stable until the age of 26, when she started to experience recurrent fever episodes and myalgia occurring exclusively, but not always, when taking showers. After a few years these fever episodes also occurred during physical exertion but between these episodes she was completely well.

INVESTIGATIONS

Routine laboratory studies including biochemical markers, as well as blood and urine cultures showed no signs of ongoing infection. Initially, a neurosurgeon was consulted due to the presence of a shunt, and a lumbar puncture was recommended in order to detect possible signs of shunt infection. However, this lumbar puncture was for unknown reasons never performed.

Since symptoms only occurred during and after showers, a shunt infection was initially not considered a primary differential diagnosis. Initial investigations were performed by an infectious disease (ID) specialist and included cultures from shower water and repeated blood and urine cultures, all of which were negative. Even though no cultures were positive, suspicion was raised as to the possibility of a subacute endocarditis but a transoesophageal echocardiogram did not detect any signs of valvular vegetations at that point.

Owing to persistent symptomatology, a lumbar puncture and CSF analysis were eventually performed. This showed no pleocytosis, normal lactate and negative cultures. Further investigations with a CT of the thorax due to an unrelated pneumonia showed para-aortic lymph nodes of up to 15 mm in diameter, raising the question of possible malignancy. Nevertheless, supplementary laboratory investigations, bone marrow biopsy and biopsy of the lymph nodes were all normal.

At the age of 30 (approximately 4 years after onset of her symptoms) the patient presented with increased creatinine levels, which rapidly

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To cite: Burström G, Andresen M, Bartek J, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2014-204655

Figure 1 Transoesophageal echocardiogram depicting a vegetation of the tendinous chords of the tricuspid valve (arrow).

progressed to a stage V chronic kidney disease. Complement components were analysed and indicated activation of the classical pathway. Further investigation led to the diagnosis of diffuse mesangiocapillary glomerulonephritis and the patient soon began haemodialysis. This again raised the suspicion of a latent shunt infection. A second echocardiogram was performed which showed a vegetation of the tendinous chords of the tricuspid valve (figure 1). The shunt valve was punctured for bacterial cultures, which were positive for *Propionibacterium acnes*, and the patient started treatment with ceftriaxone.

TREATMENT

Five years after symptom onset, and after having developed end-stage renal disease requiring haemodialysis, the patient underwent surgery with removal of all shunt hardware. At the same time an endoscopic third ventriculocisternostomy (VCS) was performed, which is a surgical opening between the third ventricle and the interpeduncular cistern resulting in continuous CSF diversion (figure 2). With all foreign material now explanted, the patient was treated with benzyl penicillin and subsequently ceftriaxone for a total of 4 weeks with full recovery of her infectious

symptoms. Haemodialysis was performed 3 times a week for 1.5 years until the patient received a kidney transplant.

OUTCOME AND FOLLOW-UP

After 2 years of follow-up, the patient remains without any signs or symptoms of hydrocephalus or systemic infection. The patient now lives independent of haemodialysis and without any CSF shunt implants.

DISCUSSION

As demonstrated in this case, shunt infections in VA shunts can be notoriously difficult to diagnose. Owing to the placement of the distal catheter in the right atrium, a VA shunt infection can give rise to severe late complications such as endocarditis and nephritis. Shunt nephritis was first described in 1965² and it is one of the most notorious complications of VA shunts. Low-virulence bacteria are typically responsible for the condition, since a chronic low-grade infection is necessary for immune-complex-mediated nephritis.⁶ In this case the infectious agent was not *Staphylococcus epidermidis*, the most common agent, but the less frequent *P. acnes* which has previously been reported in only a handful of cases.^{7–9} This may further complicate the diagnosis as *P. acnes* might be discarded as a contaminant in cultures.

The delay of diagnosis in this case was in part due to non-specific symptoms such as fever, lethargy and anorexia. The diagnosis was especially difficult to determine in this case; numerous investigations by different specialists continued for years and the suspicion of shunt infection was delayed. This highlights the importance of a well-functioning collaboration between neurologists, ID specialists and neurosurgeons in order to increase awareness of this rare condition. Furthermore, if complement activation is demonstrated in blood samples in a patient with a VA shunt it should lead to a high suspicion of a chronic shunt infection.¹⁰

Apart from illustrating the importance of being aware of VA shunt-associated infections, this case also illustrates that some patients previously considered to have a lifelong shunt dependency might instead be treated with a VCS. In case of an obstructive hydrocephalus a VCS may render CSF shunting to an extracerebral compartment (peritoneal cavity, atrium, etc)

Figure 2 Sagittal MRI sequences depicting the preoperative state (A) with an intact floor of the third ventricle (arrow) and (B) depicts the postoperative image, using flow-sensitive sequences, with a flow void artefact through the floor of the third ventricle (arrow) indicating a patent ventriculocisternostomy.

unnecessary. Hence, late infectious complications are nearly eradicated by avoiding the implantation of foreign material.

Learning points

- ▶ Patients with ventriculoatrial (VA) shunts can present with serious complications many years after surgery.
- ▶ Shunt infections, especially regarding VA shunts, can be difficult to diagnose due to non-specific symptoms.
- ▶ VA shunts are partially intravascularly placed implants and may give rise to subacute endocarditis and/or shunt nephritis in case of infection.
- ▶ Ventriculocisternostomy can be an alternative for patients previously considered to have a lifelong shunt dependency.

Contributors AF and JB contributed to the conception and design. GB acquired the data, and drafted the manuscript. MA, AF and JB performed analysis and interpretation of the data, and revised the manuscript for important intellectual content. All authors approved the final manuscript.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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