A Rare Case of Shunt Nephritis in Ventriculoatrial Shunts: Case Report and Literature Review

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Abstract

Shunt nephritis is a rare complication in patients with ventricular atrial shunt (VAS) infection, used in the treatment of congenital or acquired hydrocephalus (1). Nevertheless, nowadays its incidence has decreased considerably, affecting only 0,7 – 2.25% of all the patients with ventricular atrial shunt (2,3).

It is thought that shunt nephritis is due to the formation of immune complexes, which deposits on the glomerular base membrane in the kidney, because of a chronic bacteremia (5).

A good prognosis of this pathology is achieved if an early antibiotic treatment is established, and the infected shunt is removed.

Introduction

Case Report

A 2 years old, female patient, arrive to the emergency department, with a 6 days history of productive cough, hyaline rhinorrhea and 38.5°C fever, associated to irritability. She had medical history of preterm birth (35 weeks of gestational age at birth), Grade III intraventricular hemorrhage, anemia, structural epilepsy, she was taken to a ventricular – peritoneal shunt at the age of three months in 2013, which got infected, so another ventricular – peritoneal shunt was implanted in May of 2014, which had to be exteriorized due to infection in July, 2014. On July 2014 the ventricular – atrial shunt (VAS) was implanted.

Physical examination showed body temperature of 39.2°C, bilateral ocular congestion, palpebral edema, signs of low perfusion, ant blood oxygen saturation of 87%.

During a second examination a greater swelling on both eyelids, distended abdomen, and edema in limbs. Blood tests revealed, white blood cell of 19100 cells/µl, neutrophils of 79.9%, hemoglobin 6.3 mg/dl, hematocrit of 21.7%, platelets count of 258000, based on this it is decided to start a transfusion of 2 red blood cell units.

Blood culture where positive for gram-positive cocci, identified as *Stremtococcus mitis.* Thorax X-Ray was normal. Abdominal ultrasound did not show any alteration.

Brain CT Scan showed hydrocephalus (Image 1). It was decided to perform a cerebrospinal fluid study which revealed white blood cells 10 per camp, 8784 fresh red blood cells, 90% of the where crenate, glucose 46 mg/dl, protein 106 mg, no microorganism where identified.
Urine analysis showed positive leucocyte esterase, ph: 6, and 30-60 leucocytes per camp. It was reported a hypertensive crisis with proteinuria, showing a nephrotic syndrome. During a further more deep interrogation to the patient´s mother, it was revealed that the patient had been presenting isolated fever episodes 2 – 3 times per week, during almost one year, that resolved spontaneously or in some occasion with a single acetaminophen dose, so shunt nephritis was proposed as diagnosis.

Due to this it was decided to start antibiotic treatment with vancomycin. Is was decided to take out the VAS and rather use a ventriculostomy, with external drainage while the infection is controlled. Consecutive CSF cultures are made, but no one showed any microorganism.

On august 11th of 2015, in association with the pediatric surgery department, a new ventricular shunt was implanted, to the sub phrenic space over the hepatic surface, by the liberation of abdominal adherences under direct visualization.

The patient presented a good post operatory evolution, high blood pressure tending to get normalized, no signs of intracranial hypertension, renal function got normalized, shunt nephritis was resolved and there were no signs of systemic inflammatory response.

Blood cultures got negative, and brain CT Scan revealed resolution of hydrocephalus (Image 2).A new x-Rayimagewastakenfewmonthsaftertheresolution of theeventshowingthe position of thecatheter (Image 3)

Discussion

Shunt nephritis is a rare complication, presented in patient with VAS infection, used for the treatment of congenital or acquire hydrocephalus (1). It was first described by Black, Challacombe and Ockenden in 1965, reporting in that time 148 cases, nevertheless nowadays its incidence has considerably decreased. VAS infection incidence is between 3 – 11%, from this patients, only 0,7 – 2.25% of them will have shunt nephritis (2,3).

This conditions is associated with a chronical and steady infection of the VAS, and its produced by *S. epidermidis* in 70% of the cases (1,2), however there have been reported cases where the etiological microorganism is *S. aureus,* it is less frequently produced by gam negative and anaerobia bacteria. The pathogenesis seems to be related to a chronical bacteremia, that induce the production of immune complexes (antigen – antibody complex), which tent to migrate throughout the blood stream and finally deposit in the glomerular base membrane. Due to this process the complement system is activated and a chronical inflammatory reaction on the kidney, leading to the symptoms (2,4). The medium time for the onset of manifestations, since the moment the VAS was implanted, to the generation of the nephritis is 4,4 years, but it can be established from the second week after the surgery to the 14th year (2).

The medical profile is very variable, which delays diagnosis, in one study performed with 6 patients with shunt nephritis, the average time from the onset of symptoms to the moment of diagnose was 5 months (1). The signs and symptoms that are most frequently presented are recurrent fever (88%), hepatosplenomegaly (55%), non-thrombocytopenic purpura(20%), anemia (85%), within the manifestation of central nervous system compromise there are seen behavior changes, vomiting, and seizures. Renal manifestations include macroscopic or microscopic hematuria in 90% of the patients, elevates blood creatinine and ureic nitrogen, high blood pressure and heavy proteinuria, getting close to nephrotic range. Laboratory tests may show proteinuria in non-nephrotic range, high blood levels of creatinine andureicnitrogen. Cultures form blood or CSF samples tent to be positive in 90% of the patients, it can also be anemia, low complement levels and positive rheumatoid factor (1–4).

In this patients renal biopsy is indicated, and will show proliferative lesions such as: mesangiocapillary glomerulonephritis, diffuse endocapillary proliferation and mesangial glomerulonephritis. Electronic microscopy may reveal accumulation of electro dense deposits at mesangial and sub-epithelial levels, however there is no evidence con glomerular base membrane engrossment. Immunofluorescence shows accumulation of immunoglobulinIgM, in the 84% of the cases, IgG in 66%, and C3 fraction of the complement system in 94% of the cases.

Treatment of shunt nephritis is based on two pillars: the first one is the change of the VAS and the second one is antibiotic treatment. The infected shunt must be retired, while hydrocephalus should be treated by using other options, like scheduled CSF drainage through lumbar puncture, which should be used for at least two weeks before implanting a new shunt, which has to be set in a different place from where the infected one used to be. By the other side intravenous antibiotic treatment is recommended for at least 10 days. It is necessary to complete this two therapeutic pillars because it has been demonstrated that recurrence rate can be up to 50% in patient who were treated just with antibiotics (1,2).

Most of the times renal function is recovered totally after the event, there are few cases where it has evolved to a chronical kidney disease, this is why prognosis in these patients is good (2).

Conclusion

Nowadays shunt nephritis is a rare entity, which is presents in patients with VAS. Its pathophysiology is explained by the deposit of immune complexes con de glomerular base membrane in the kidney. It is a difficult diagnosis and a challenging one, generally is one of the lasts options of diagnosis, and the most specific tests are blood cultures.For the treatment it is recommended to start antibiotic treatment, exteriorization of the shunt, ventriculostomy or scheduled CSF drainage. It is needed to implant a new shunt, which should be implanted in a different location from the infected one. The case we present is not only rare nowadays, but also reveal a different etiologic agent from the common ones and the new shunt was placed in an anatomical space which is difficult to manage.

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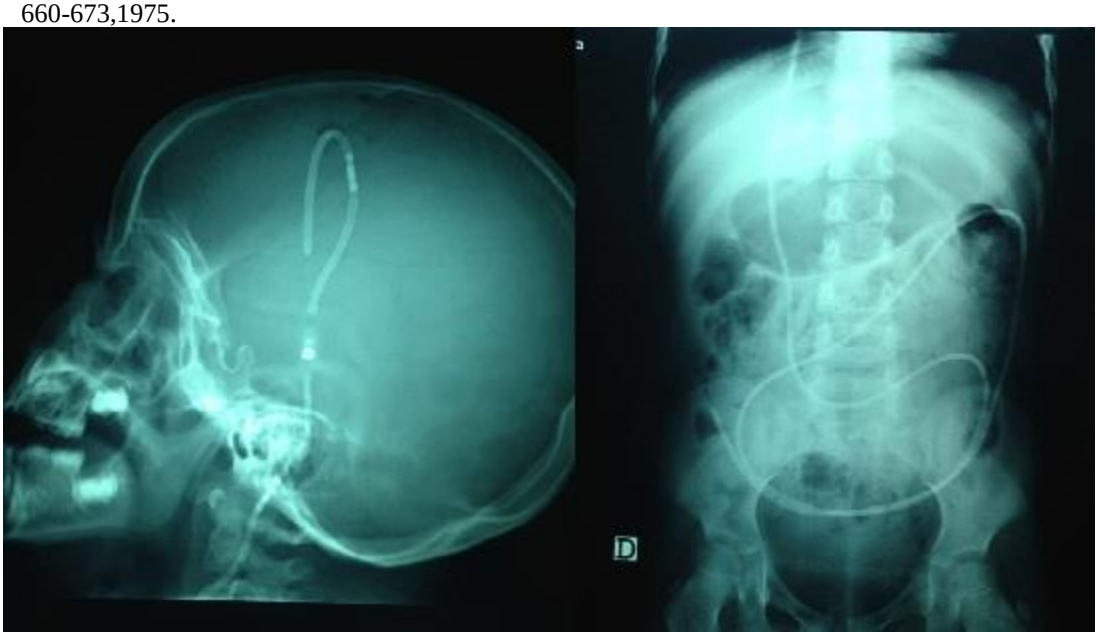

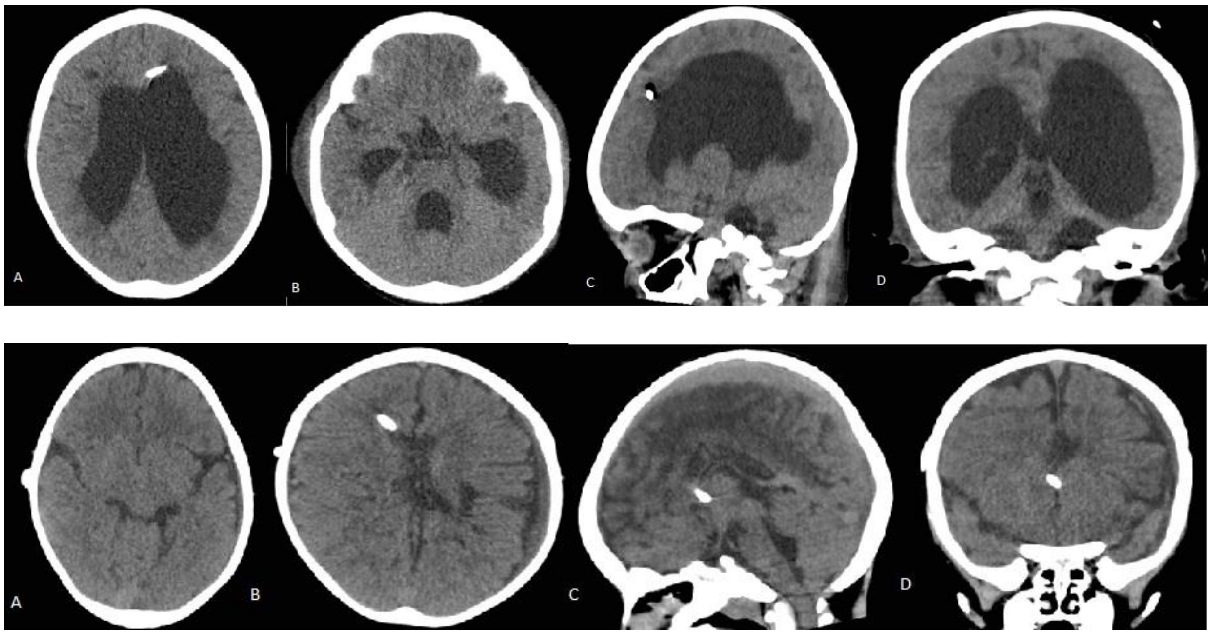
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