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Case report

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Shunt nephritis

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Summary
Shunt nephritis is a rare complication associated with the use of atrialventricular shunts and their chronic infection. The microorganisms that cause the infection are generally of low virulence, but they can produce long term infections that permit the production of immune complex that deposit in the kidneys leading to nephritis. Because of this we want to describe the case of an adolescent patient with hydrocephalus, as a consequence of meningitis in the childhood, and chronic use of atrialventricular shunts. She presented fever and child for a long time, these symptoms were secondary to infection of the device, and shunt nephritis was also present.

Key words: Shunt nephritis, immune complexes, and atrial ventricular shunt infection.

Nefritis del shunt

Resumen
La nefritis por shunt es una complicación poco frecuente que se asocia al uso de derivaciones ventriculoatriales y su infección crónica. Las bacterias que la producen, por lo general, son de baja virulencia, pero producen infección prolongada y persistente permitiendo así la producción de inmunocomplejos que se depositan a nivel renal. Queremos describir un caso de una adolescente con hidrocefalia posterior a un evento de meningitis, con uso de derivación ventrículoatrial quien ingresa con un cuadro de fiebre y escalofrío de larga evolución, en la cual se documentó infección del dispositivo y nefritis por shunt.

Palabras clave: Nefritis por shunt, inmunocomplejos, endocarditis.

Introduction

Shunt nephritis is an infrequent complication in patients who develop infection of the atrioventricular shunt used for congenital or acquired hydrocephalus. It was described by Black in 1965[1], at a time when 148 cases had been identified. [2,3] However, the occurrence of this condition has declined over the past few decades, to the point where it is a very unusual presentation nowadays. The global incidence of shunt infection ranges between 3% and 11%, and shunt nephritis occurs in close to 0.7-2.25% of patients presenting with an infected shunt.[4] The bacteria implicated are usually of low virulence and give rise to a chronic non-specific infection that favours formation of circulating immune complexes which, in turn, create subepithelial and mesangial deposits. When diagnosed early and treated adequately with antibiotics and device removal, the prognosis of this disease is good.

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Case description

A 16 year-old female patient with a history of hydrocephalus secondary to bacterial meningitis in childhood requiring a ventriculoperitoneal shunt at 3 years of age. Because of multiple mechanical complications, the shunt was exchanged for an atrioventricular shunt at 14 years of age. She presented with fever associated with general malaise, nausea and vomiting, with symptoms in the form of morning chills, general malaise and occasional diaphoresis with no fever, and dark, foul-smelling urine having started three months after the implantation of the atrioventricular shunt.

On physical exam, the patient was alert, hydrated, with a heart rate of 145 x min, respiratory rate of 23 x min, blood pressure of 110/75 mmHg, temperature of 39.5, and SO2 92%. There wasgeneralized mucocutaneous pallor, no signs of dehydration, palpableshunt device in the frontotemporal and right cervical regions showing no tenderness to palpation; no signs of infection, rhythmic heart sounds, no murmurs, preserved breath sounds with no aggregates. No palpable abdominal masses, no abdominal tenderness; grade II oedema of the lower limbs; bitemporal hemianopsia secondary to childhood meningitis; no motor or sensory deficit.

Tests were performed on admission to the emergency department, revealing the presence of leukocytosis and neutrophilia with no evidence of immature forms, elevated CRP, negative procalcitonin, normochromic normocytic anaemia, urinalysis with hematuria and proteinuria in isolated sample, not suggestive of infection. Electrolytes within normal limits, 4 blood cultures positive for slow-growth Gram positive cocci of the Micrococcus spptype, prompting antimicrobial treatment with vancomycin on hospital day 5. Renal function was normal. On transesophageal echocardiography: no evidence of infection foci affecting the cardiac structures, tricuspid aortic valve with mild regurgitation, normal systolic and diastolic function, unaltered atrioventricular diversion catheter in the right atrium. The chest X-ray showed an adequately positioned atrioventricular diversion catheter (Figure 1).

The only finding on abdominal ultrasoundwas mild splenomegaly. A lumbar tap was performed after 4 days on antibiotic therapy, showing opening pressure of 16 cm H2O, cerebrospinal fluid showing evidence of hypoglycorrhachia and hyperproteinorrhachia, culture and Gram, Ziehl-Neelsen, and Indian ink stains were all negative (Table 1). In view of persistent hematuria and proteinuria in the urinalysis, additional workup was requested including renal and urinary tract ultrasound, which were found to be within normal limits. The finding of depleted C3-C4 complements led the nephrology service to consider shunt nephritis. The infectious diseases team suggested vancomycin continuation and shunt removal.

Discussion

Shunt nephritis is an immune complex-mediated glomerulonephritis associated with chronic infection of the atrioventricular shunt. Incidence may be as high as 2.25%[4] in individuals carrying infected atrioventricular shunts, affecting mainly paediatric and adolescent patients.

This complication is exceptional in patients with ventriculoperitoneal shunts given that the peritoneum is considered to act as a barrier to bacteria, preventing the formation of immune complexes.[5] The time period over which this condition develops from the moment the shunt is placed and nephritis occurs varies significantly between 3 weeks and 14 years, with a mean of 4.4 years. In the case of this patient, the time course was 18 months.

The most common infectious agent is S. epidermidis, followed by S. aureus, but cases of Gram negative rods, anaerobes and even fungi isolates have also been described.[6] Positive blood cultures are found in 90% of patients with infected atrioventricular shunts. This is in contrast with infections in other types of CSF shunts, in which the incidence of negative blood cultures is close to 80%. The clinical picture is characterized by fever, hepatosplenomegaly, anaemia and purpura.[7] Renal manifestations include macro and microscopic hematuria with proteinuria of varying degree[6] that may even reach nephrotic

Figure 1.

Chest X-ray showing adequately positioned atrioventricular shunt.

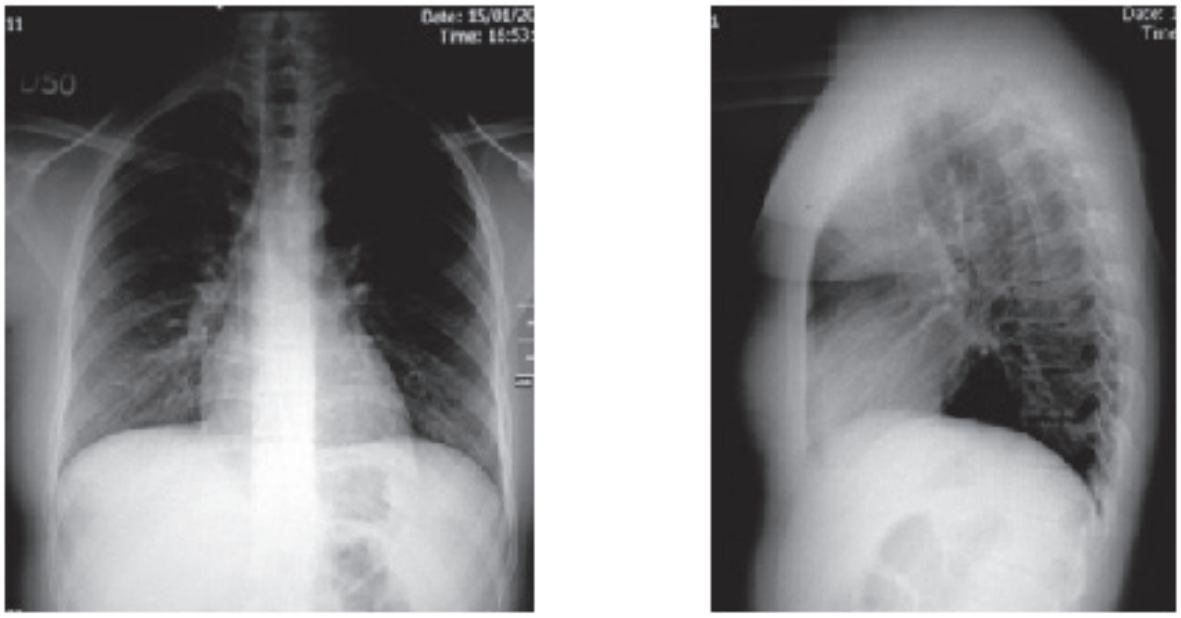

Table 1

Relevant test results during hospital stay

Complete Blood Count		CSF		Blood chemistry	
Leukocytes	14.500	Glucose	36mg/dl	Glycemia	85 mg/dl
Neutrophils	85.2%	Protein	84	BUN	16
Hemoglobin	10.6mg/dl	Cytochemical	Xantochromic limpid, leu: 4 mm3, RBC 15 mm3, 70% fresh	CreatininE	0,6
Hematocrit	32.2			Complement	
Platelets	211.000			C3	55.20
Cryoglobulins	1/8			C4	7.000

ranges. Patients may show an increase in nitrogen compounds, but advanced renal injury or high blood pressure are unusual at the time of the initial diagnosis.

Complement testing is part of the workup, as this type of nephritis is characterized by hypocomplementemia and the presence of circulating immune complexes and positive cryoglobulins, as evidenced in this patient. Rheumatoid factor positivity may also be seen occasionally. Tests usually return to normal once the infectious process is resolved. It is worth noting that shunt nephritis may be the earliest manifestation of shunt infection, with very bizarre preceding symptoms.

The persistent bacteremia caused by low-virulence microorganisms associated with this disease leads to the presence of circulating immune complexes that saturate the reticuloendothelial system and end up as deposits in glomerular capillaries, activating the complement cascade and the chronic inflammatory reaction that results in renal injury.[6-8]

Renal biopsy histologyshows proliferative lesions of a mesangiocapillary glomerulonephritis type, diffuse intracapillary proliferation and mesangial glomerulonephritis. Electron microscopy shows a granular pattern of electrodense deposits in the mesangium and the subendothelial space. Immunofluorescence reveals 84% and 66% IgM and IgG immunoglobulins, respectively, 94% C3 complement and bacterial antigens.[6] Because our patient met all the diagnostic criteria, no biopsy was performed.

Treatment of shunt nephritis is based on eradication of the infection and valve exchange. Antibiotic therapy is systemic and, in some cases, it may be combined with intrathecal antibiotic administration. For valve exchange, the recommendation is to place a new shunt in a site different from the old one.[9-10] Some authors recommend only intraventricular antibiotic treatment, although cure rates are only close to 50%.

In view of the above, despite being rare, this condition needs to be suspected, bearing in mind that the clinical diagnosis of chronic shunt infections is not easy and blood and CSF cultures may be negative. For this reason, this condition is frequently underdiagnosed, leading to delayed treatment by months or even years. Early diagnosis and adequate therapeutic management are key for symptom resolution and for reducing the risk of renal injury.[11]

Conflict of interest

The authors declare having no conflict of interest.

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