

Case Report

Cystoperitoneal Shunt Malfunction in Setting of Rifampin Allergy Presenting as Cerebrospinal Fluid Eosinophilia

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Abstract

Background: Cerebral spinal fluid eosinophilia (CSFe) is correlated with increased risk of cranial shunt malfunctions. Although antibiotics coated shunt catheters have significantly reduced the incidence of infections and associated shunt malfunction, on rare occasion they may contribute to a shunt allergy and become the source of subsequent malfunction.

Objective: We present a very rare case of a patient with an arachnoid cyst treated with cysto-peritoneal shunt (CPS), complicated by rifampin allergy manifesting as sterile CSFe and subsequent shunt malfunction.

Case Report: A 17-months-old-male with symptomatic arachnoid cyst underwent uneventful craniotomy for cyst fenestration and CPS placement. At 2-month follow up he was neurologically stable; however, he developed an expanding pseudomeningocele that was palpable at the site of the prior craniotomy. Although shunt and pseudomeningocele taps were both negative for infectious etiology, the CSF analysis was notable for isolated CSFe. Given the persistence of symptoms, and well as subsequent emergency room visits for abdominal swelling, low-grade fevers, and intermittent presence of pseudomeningocele, patient was admitted for further workup. Radio-Allergosorbent testing (RAST) was performed and revealed newly diagnosed rifampin allergy. CPS was then removed and patient was treated with a course of steroids until resolution of CSFe, and underwent placement of a new CPS with non-antibiotics coated catheter. One week after discharge, patient returned with Staphylococcus Aureus related shunt malfunction, for which the shunt was externalized. He underwent a course of antibiotics treatment and was subsequently trialed to be weaned off the CPS, in light of recent cyst fenestration. A

ventriculogram revealed complete occlusion of the prior cyst fenestration, thus patient underwent repeat craniotomy for more aggressive cyst fenestration. He was eventually weaned off the shunt and did not require further catheter placement.

Discussion: CSFe is associated with underlying infection, inflammatory changes, and hypersensitivity. This can be seen in up to 30% of shunt malfunctions. In the above case, the Rifampin allergy contributed to elevated levels of CSFe and secondary shunt failure. The inflammatory response may have contributed to the closure of initial fenestration, shunt failure, and granulomatous response.

Conclusion: Classic presentations of the allergy to the shunt catheter or its associated antibiotic coating are nonspecific and difficult to distinguish from primary infection. In complex cases, where an underlying cause is undetermined, particularly in the setting of isolated elevated CSFe and persistent non-specific shunt related symptoms, further workup is warranted. This case also highlights the need for improved biocompatibility of shunt hardware to avoid conditions leading to shunt malfunction.

Keywords: Rifampin allergy; Shunt malfunction; CSF eosinophilia; Arachnoid cyst; Hydrocephalus

1. Introduction

Shunt failure remains an ongoing problem in neurosurgery and is commonly associated with infection or mechanical obstruction. In certain cases of allergy or hypersensitivity reactions, the shunt system may become obstructed with inflammatory debris, ultimately leading to overt shunt malfunction [1-4]. Clinical symptoms of shunt allergy or hypersensitivity reactions include abdominal pain, fever, recurrent skin breakdown along the shunt tract, abdominal distention, peritonitis, and non-specific complaints along the shunt tract [2, 3, 5-7]. After detecting a shunt malfunction, it is important to perform a thorough workup in assessing for both infectious and noninfectious causes [1, 4, 5, 8-13].

Eosinophils are rarely seen within the cerebrospinal fluid, and when found they are usually associated with parasitic, bacterial or mycotic infections of the central nervous system [1, 12-16]. However, CSF eosinophilia may also be detected in up to 30% cases of allergic or aseptic shunt malfunctions [15]. We present the first described case of an arachnoid cyst treated with a cystoperitoneal shunt (CPF), which was subsequently complicated by sterile shunt malfunction secondary to CSFe and hypersensitivity reaction to Rifampin-Minocycline coated catheter, due to a previously undiagnosed Rifampin allergy.

2. Case Report

A 17 months-old-male with symptomatic arachnoid cyst underwent craniotomy for cyst fenestration of a large, compressive, temporal lobe, arachnoid cyst into the basilar cisterns, with temporary intracystic catheter placement, skin to that of an external ventricular drain. Patient went on to require further drainage via the intracystic catheter, for which he underwent a CPS placement. Immediately postoperatively he did well and was discharged home. In the

weeks postop, he presented with intermittent low grade fevers of unknown origin and transient abdominal distention in the setting of negative work-up for shunt malfunction, peritonitis, or other infectious etiology. Patient remained neurologically stable, however, over the ensuing two months he developed an expanding pseudomeningocele at the cranial surgical site. The pseudomeningocele was intermittent in nature; on occasion it was minimally appreciated and at times it was noted to be bulging and tense. Shunt and pseudomeningocele taps were both negative for infectious etiology (including cell analysis, protein, glucose, gram stain, culture, and prolonged broth evaluation) on separate occasions. Both taps were revealing for elevated CSF, which raised a concern of associated inflammatory reaction or concern of potentially indolent infection. Ultimately, he was admitted and further workup of CSF eosinophilia via Radio-allergosorbent testing (RAST), revealed new diagnosis of rifampin allergy. Cystoperitoneal shunt was removed and replaced with a temporary non-antibiotic coated external intracystic drain and a course of steroids until near complete resolution of eosinophilia, prior to placement of a permanent shunt.

Within one week of new shunt placement and subsequent discharge, patient developed persistent low-grade fever and shunt tap revealed presence of *Staphylococcus Aureus* organism; therefore the shunt was removed and replaced with external intracystic catheter. Patient completed a course of proper antibiotic treatment prior to re-internalization of the shunt. In the interim, a ventriculogram was performed to assess patency of prior fenestration; there was complete occlusion of prior fenestration. A repeat craniotomy, more aggressive cyst resection and fenestration was performed with subsequent ventriculogram study revealing complete opening of the cyst into the basal cisterns. Intraoperatively, the patient's arachnoid cyst walls had thickened relative to his initial surgery and he had developed multiple adhesions within the cyst that were not previously present. After re-fenestration, an external intracystic catheter was placed in the cyst cavity and it was eventually weaned off as the fenestration site remained patent.

3. Discussion

Underlying CSF eosinophilia has been reported as both an incidental and benign finding in neurosurgical patients [8, 9, 13, 14, 16-18]. However, when CSF eosinophilia is present, it may be an indicator of an underlying pathology associated with, or possible cause of shunt malfunction. In patients presenting with concern of shunt malfunction in the setting of elevated CSF eosinophil count, clinicians should consider workup of infection, benign underlying inflammatory reaction, or both [1, 2, 4, 6, 7, 9, 10, 12-14, 18]. CSF eosinophilia has been reported in association with infection such as parasitic, bacterial, or mycotic infection of the central nervous system [1, 7, 9, 12, 13, 15, 16, 19]. Additionally, CSF eosinophilia has also been reported in association with inflammatory reaction to antibiotic coated shunt catheters, as reported in the case of rifampin and minocycline impregnated shunt catheters [1-6, 8, 11, 14, 20, 21]. Minocycline has been more commonly reported culprit in cases of shunt malfunction. However Rifampin has also been reported, as seen in this case report.

Patients, who present with signs of shunt malfunction with coexisting CSF eosinophilia, in the absence of shunt infection, should be considered for obstruction of the shunt system with inflammatory cell debris [2, 6, 7]. In the above case however, the arachnoid cyst contents, as well as a rifampin allergy could potentially contributed to high levels of CSF eosinophilia and secondary shunt failure [1, 22]. Spikes in eosinophilia were noted in the immediate

cyst fenestration, subsequent shunt placement, and repeat fenestration in this particular case report, as they have been previously discussed in the literature [16]. The inflammatory response likely contributed to the closure of initial fenestration, thickening of the cyst wall, and a granulomatous response, even prior to presenting with infection. Variations of eosinophilia in the CSF also correlated with congruent changes of CSF in the blood of these particular patients.

It is important to monitor patients with hypersensitivity reactions associated with sterile shunt malfunction as they are more likely to have multiple episodes of shunt failure [6, 9, 11, 17]. Treatment of aseptic shunt failure may include removal of the current shunt system and replacement with a hypoallergenic “extracted” shunt and in cases where CSF eosinophilia has been determined, a course of steroids may ultimately reduce the inflammatory reaction and prolong shunt survival [14, 23]. This case demonstrates the importance of recognizing CSF eosinophilia and identifying other noninfectious causes of shunt failure in order to treat patients successfully and avoid future shunt complications.

4. Conclusion

In complex cases of shunted hydrocephalus, where a clear underlying cause is not determined, further workup to rule out infection, inflammatory changes, and hypersensitivity may be warranted. Recurrent presentations to the hospital after shunt hardware placement should prompt clinicians to conduct studies beyond routine CSF analysis and infectious process workup, as seen in this case. Elevated CSF eosinophil count, with or without elevated blood eosinophil count, could be a potential tool in the surgeon’s armamentarium, when evaluating patients with such clinical concerns. Although, the specificity and sensitivity of CSF eosinophil count is insufficient to use it as a screening tool, when interpreted in conjunction with clinical presentation, it can shine light on the underlying pathology of shunt failure [19]. This case also highlights the need for improved biocompatibility of shunt hardware to avoid conditions leading to shunt malfunction.

Conflict of Interest

No disclosures or conflicts of interest to be disclosed for all authors involved in this manuscript.

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