Treatment of recurrent ventriculoperitoneal shunt failure associated with persistent cerebrospinal fluid eosinophilia and latex allergy by use of an “extracted” shunt

Case report

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Shunt failure is commonly associated with infection or mechanical obstruction of the shunt system. The presence of eosinophilia in the cerebrospinal fluid (CSF) has been associated with CSF shunt failure and may be related to both latex and shunt allergies. The authors describe the case of a child with a latex allergy who presented with 10 episodes of shunt failure over a period of 93 months. Cerebrospinal fluid sampling demonstrated persistent eosinophilia (3–36%) and negative cultures. Pathological examination of the ventricular catheter on 3 occasions demonstrated mechanical obstruction by inflammatory debris consisting largely of eosinophils and multinucleated giant cells. On the suspicion that the child might have some uncharacterized allergy to the shunt hardware, shunt replacement was performed using an “extracted” shunt system. The child has remained free of shunt malfunction for > 2 years since the last surgery. Immune responses to unpolymerized silicone are discussed. (DOI: 10.3171/PED/2008/1/3/237)

Key Words • cerebrospinal fluid eosinophilia • extracted shunt • shunt failure • ventriculoperitoneal shunt

For > 50 years, VP shunts have been used as the principle surgical treatment of pediatric hydrocephalus. The 2 most common factors contributing to shunt malfunction are infection and mechanical obstruction of the ventricular catheter by tissue from brain and inflammatory processes.11,18 One inflammatory phenomenon that has been repeatedly linked to shunt malfunction is CSF eosinophilia.2,25 The role of silicone allergy in shunt failure and CSF eosinophilia is poorly understood.

We describe the case of a child with a latex allergy who presented with multiple episodes of shunt failure associated with CSF eosinophilia and proximal shunt obstruction secondary to an eosinophilic immune reaction.

Case Report

This now 10-year-old boy had been born at 35 weeks’ gestation, and suffered bilateral Grade III intraventricular hemorrhages shortly after birth. Progressive ventriculomegaly and signs and symptoms of raised intracranial pressure led to the placement of a VP shunt on the right side. Blood and CSF cultures performed at the time of the first surgery grew Staphylococcus epidermidis for which the child received antibiotic therapy. Repeated cultures obtained 1 week later were negative.

Over the next 7 years, the patient presented with 10 episodes of shunt failure requiring shunt revision. At 1 presentation, an endoscopic third ventriculostomy was done but failed. After the second episode of shunt malfunction, the patient was found to have marked hypersensitivity to latex, and latex precautions were subsequently initiated for all surgical procedures. Cerebrospinal fluid cultures at the time of all shunt revisions failed to grow any organisms, however, there was CSF eosinophilia, which ranged from 3 to 36%.

On 3 occasions, microscopic examination of the shunt hardware was performed. The ventricular catheters were obstructed by inflammatory material consisting largely of eosinophils in a background of loose vascularized connec-
tive tissue. In addition, multinucleated giant cells and macrophages, CD4+ lymphocytes, and rare plasma cells were seen (Fig. 1). Similar tissue was adherent to the outer surface of the catheter. No glial tissue, bacteria, or fungi were identified.

Based on the pathological findings and persistent CSF eosinophilia, we suspected that the child might have an uncharacterized allergic response to the shunt hardware. As such, we planned in the event of another episode of shunt failure to use an “extracted” shunt. Extracted shunts are felt to be less likely to induce an allergic response. On the child’s next presentation with shunt malfunction, the shunt was removed and an antibiotic-impregnated external ventricular drain was placed in the right frontal horn. Three days later, once negative CSF cultures had been confirmed, a left occipital “extracted” shunt (Extracted Delta Valve and Extracted Ventricular and Peritoneal Catheters, Medtronic) was placed. After 2 more episodes of shunt malfunction within 2 days of surgery (1 immediate proximal obstruction presumably due to brain tissue debris, 1 distal air lock) and revision with similar hardware, the patient’s signs and symptoms resolved and postoperative computed tomography scans demonstrated successful reduction in ventricle size. Since placement of the “extracted” shunt, the patient has remained free from shunt malfunction for > 2 years.

Discussion

Ventriculoperitoneal shunting is the mainstay of treatment in children with hydrocephalus. Despite the successful institution of specialized intraoperative protocols,6,10 the use of antibiotic-impregnated shunt catheters,1 and advances in endoscopic placement procedures,12 the clinical course in children with shunts remains fraught with complications that often necessitate numerous revision surgeries.9 By far, the 2 most significant complications of shunt placement are infection and mechanical obstruction.

Ventricular catheters are commonly obstructed by brain tissue, including choroid plexus and associated connective tissue, vascularized astroglial tissue, hemorrhagic debris, and ependyma. In many cases, ventricular catheter obstruction is also accompanied by an inflammatory process that includes foreign body giant cells and lymphocytes.4 Patients who exhibit CSF eosinophilia have been found to experience more shunt malfunctions.2,13,14,25,26,28

Allergies to shunt hardware,9,22 ethylene oxide,17 and other foreign materials such as suture material, glove powder, and antibiotic irrigation have been implicated as causes of eosinophilia.24 Silicone hypersensitivity has been implicated in cases of both proximal and distal shunt obstruction,23 as well as rare complications such as abdominal CSF pseudocysts8 and colonic perforation.3 Snow and Kossovsky20 examined the shunt hardware in patients who had undergone multiple shunt revisions and concluded that there was evidence of hypersensitivity reactions with an abundance of eosinophils. Our case is unusual in that the shunt-obstructing mass was almost entirely composed of eosinophils rather than the scattered eosinophils mixed with other material that is often seen.

The authors of several studies have attempted to quantify the response associated with failed shunts. Blocked distal catheters have been found to have more protein deposition on the surface, and these children were found to have higher levels of autoantibodies against extracted proteins on the shunt surface.27 These findings suggest that immunologic responses mounted against antigens present on the surface of silicone shunt hardware may play an important role in aseptic shunt malfunction. A high prevalence of latex hypersensitivity has been established in children with spina bifida,10 but its relationship to repeated shunt malfunction is unclear. A patient with persistent CSF eosinophilia, an allergy to latex, and multiple shunt revisions was successfully treated with glucocorticoids.23 Another child with profound hypersensitivity to latex and high CSF eosinophilia (80%), underwent successful shunt revision in a latex-free environment.16

In our patient, on the suspicion that an allergy might be contributing to the recurrent shunt malfunctions, we inserted a shunt system composed of an “extracted” silicone elastomer. Silicone elastomers contain a small amount of unbound silicone oligomers, which might be able to interact with the immune system on the surface of the catheter.9 To remove most of the unbound silicone oil and other impurities such as platinum (which is used as a polymerizing catalyst and which might also be allergenic), the hardware is subjected to an extensive solvent extraction process. There have been prior suggestions that this is helpful,23 and unpublished feedback from surgeons suggests that this product works well in individuals who are apparently sensitive to silicone (personal communication, M.R.D. with Jeff Bertrand of Medtronic, 2006), however, there are very few documented cases of the successful use of an extracted system. The only other published case of reduced shunt complications with extracted silicone involved peritoneal reactions, and no histological evaluations were done.8 It should be noted that extracted shunts are not impregnated with radioopaque barium sulfate, which in rare circumstances has been implicated in allergic and inflammatory responses.19,21

Although a specific “shunt” allergy has not been demonstrated in this or previously reported cases, the presence of CSF eosinophilia is highly suggestive of an allergic response. This case is one of the few demonstrating actual obstruction of the shunt lumen with an inflammatory mass made up largely of eosinophils, as well as a long period of

Fig. 1. Photomicrograph showing material from the ninth shunt revision. Apparent are small blood vessels (filled with erythrocytes, patterned arrow) that are surrounded by eosinophils with multilobulated nuclei, and eosinophilic granules in the cytoplasm (black arrows). H & E; original magnification × 400. Bar = 20 μm.
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shunt survival after use of an extracted shunt system. We recommend further study of the use of extracted silicone shunt systems in circumstances of repeated early shunt failure and recommend histopathological examination of all catheters to identify intense noninfectious inflammatory processes. The allergens or proinflammatory chemicals involved in these processes need to be defined.

Disclosure

Dr. Del Bigio has served as a consultant to Medtronic for the development of shunt catheter technology, although not specifically related to extracted shunts. None of the authors have reported financial interests in these products.

References


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