Intracranial calcified pseudocyst reaction to a shunt catheter

Case report

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A 9-year-old boy with spina bifida, Chiari II malformation, and hydrocephalus presented with signs of increased intracranial pressure consistent with a shunt malfunction. Radiological investigations revealed an intracranial calcified lesion along the ventricular catheter. A shunt tap revealed a translucent milky white fluid. The patient underwent a ventriculostomy and, eventually, a shunt revision. Pathology findings were consistent with the formation of dystrophic calcification and a pseudocyst around the shunt catheter. Postoperatively, the patient returned to his neurological baseline. This is, to the best of the authors’ knowledge, the first report of an intracranial calcified pseudocyst in a patient with normal renal function.

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KEY WORDS • ventriculoperitoneal shunt • calcification • pseudocyst • hydrocephalus

Long-term shunt complications may occur as a result of silicone degradation, which can cause dystrophic calcification of the shunt tubing. This shunt calcification most commonly occurs extracranially, in areas of mechanical stress, such as along the neck or anterior chest wall. However, intracranial calcification of a lateral ventricle wall from a shunt catheter in an adult has been reported. We report a rare case of dystrophic calcification and development of an intracranial pseudocyst in a pediatric patient.

Case Report

History and Examination. This 9-year-old boy had a history of spina bifida, Chiari II malformation, congenital talipes equinovarus, and hydrocephalus. He had undergone myelomeningocele repair and placement of a rightsided occipital ventriculoperitoneal shunt (VPS) at birth, with no revisions having been required. His mother was a 20-year-old primigravida woman, and his birth, after a full-term pregnancy, was done through a Caesarean section because of a breech presentation. His myelomeningocele was first detected at birth. At baseline, the patient had a neurogenic bladder, fecal incontinence, and used ankle foot orthoses when ambulating. He presented with a 1-day history of increasing lethargy and neck pain. On examination, the patient was asleep but arousable. He was otherwise neurologically at his baseline. The shunt valve compressed and refilled easily.

A shunt series did not show catheter breaks or disconnects but did show abnormal calcification of the catheter, especially around the clavicle (Fig. 1). A head CT scan showed increased attenuation in both lateral ventricles, occipital horns, and along the right temporal horn as well as along the shunt catheter before it entered the atrium of the right lateral ventricle (Fig. 2). The attenuation around the catheter was unusually high for acute blood and was associated with edema. The abnormal hyperintense mass along the track of the catheter communicated with the right lateral ventricle. Compared with a head CT scan obtained a year previously (Fig. 3), an interval increase in the size of the third and lateral ventricles was also noted.

Hospital Course. Due to the patient’s increasing somnolence and worsening hydrocephalus, he was intubated. A shunt tap was initially performed, and a milky white fluid was easily aspirated from the reservoir (Fig. 2). The patient received a ventriculostomy on the first hospital day to clear the white CSF from his ventricles. His intracranial pressures increased into the 40s shortly after drain placement and were managed with mannitol, sedation, and hypertonic saline. Brain MRI showed no pathological leptomeningeal or pachymeningeal enhancement or cystic process (Fig. 4). The patient’s status eventually returned to its neurological baseline and remained stable.

Abbreviation used in this paper: VPS = ventriculoperitoneal shunt.
for the rest of his hospital course. The ventriculostomy was discontinued once the CSF became clear. The patient underwent removal of the right VPS, which was extracted without difficulty and was not adherent to the surrounding brain tissue. The patient then underwent placement of a left occipital VPS with a medium-pressure valve (Fig. 5). The patient was discharged 2 days after the shunt revision, and he made a full recovery. Follow-up at 1 year showed the patient to be doing well with no signs of a VPS malfunction.

Pathological Findings. Laboratory investigations showed no abnormality of renal function or calcium metabolism. Blood urea nitrogen and creatine were 5 mg/dl and 0.3 mg/dl, respectively. The results of CSF studies are shown in Table 1. Of note, the calcium level was 88.5 mg/dl compared with a serum calcium level of 8.4 mg/dl. A Gram stain showed no organisms or neutrophils with large amounts of amorphous material. Bacterial, fungal, and viral cultures showed no growth at 4 weeks. Cytological examination of the CSF showed no erythrocytes or leukocytes, and glucose and protein levels of 56 mg/dl and 197 mg/dl, respectively.

After removal of the shunt, tissue adherent to the tip was sent for pathological examination. Results showed membranous reactive gliotic and fibrous tissue, with attached myxoid proteinaceous material with dystrophic calcification, with minimal chronic inflammation, most consistent with a pseudocyst. No epithelial-lined cyst or evidence of neoplasia was found.

Discussion

Shunt complications can be categorized as early (within the 1st year of implantation) or late (after the 1st year). The former relates to problems with technical problems, infection, and malfunction of the shunt itself while the latter refers to obstruction and degradation of the materials that comprise the shunt itself. In our patient’s case, he developed a long-term complication from his shunt catheter, most likely a reactive process related to the aging materials of his 9-year-old shunt.
Silicone is extensively used for the manufacturing of bio-products, such as shunt catheters due to its inert properties. However, there are numerous reports of adverse biological reactions to the Silastic tubing, especially in the periphery. Three responses of the body to silicone have been reported, including a local, granulomatous reaction, migration of the silicone, and human adjuvant disease with chronic exposure causing the development of a fibrous tissue pseudcapsule with little inflammatory response. Jimenez et al. suggested that the body may exhibit all of these reactions to silicone to some extent, depending on the patient’s own cellular and humoral immune responses based on his or her human leukocyte antigen genotype.

With regard to calcification of VPS catheters in the subcutaneous tissues, 2 mechanisms have been described. One mechanism is metastatic calcification that presents with elevated levels of calcium and phosphate. The other is dystrophic calcification that presents with normal levels of phosphorus and calcium but an abnormal cellular metabolism. Dystrophic calcification causes a dense fibrous reaction zone around the catheter. This reaction is associated with formation of nuclei from cellular debris that is propagated by flaws in the catheter or changes in the chemical composition of the catheter due to age (silanol group formation or element extrusion). Calcium phosphate deposition typically occurs with breakdown of the catheter and a reactive inflammatory response, most commonly calcification of the shunt catheter tract in the occipital or the lateral cervical regions rather than the chest wall. However, it has been reported that the cerebral parenchyma does not surmount a cellular immune response like subcutaneous tissues, and gliosis does not tend to occur in the brain as a reaction to the Silastic tubing. Finally, shunt calcification tends to occur more in younger patients due to the presence of a relative increased amount of serum phosphorus compared with that in adults. Interestingly, it has been noted that the cellular metabolism of calcium and phosphate does not need to be abnormal to have development of dystrophic calcification of the silicone catheter.

However, there is one example in the literature of a case of intracranial ventricular wall calcification in a 53-year-old man with end-stage renal disease and VPS in place for 11 years who presented with symptoms of a shunt malfunction. A head CT scan revealed ventricular calcification and a hyperdense CSF precipitate, with an inflammatory response noted in the ventricular tissue. This case seems to be a rare occurrence, and the patient seemed prone to abnormal calcium and phosphate metabolism due to his renal disease. The authors hypothesized that the dystrophic calcification started in the neck and extended into the brain due to his abnormal metabolism.

To the best of our knowledge, the current paper represents the first report of an intracranial pseudocyst with dystrophic calcification in a pediatric patient. This case report illustrates a unique case in which a young boy with no known abnormal calcium or phosphate metabolism is found to have had a reactive formation of a pseudocyst that developed in less than a year.

The authors hypothesize that the subcutaneous shunt calcification produced fluid that migrated intracranially. The patient’s immune system probably reacted to the
fluid by forming a pseudocyst. However, the pseudocyst broke and the fluid was then released into the ventricular system, causing signs and symptoms of a shunt malfunction. Although the patient does not share renal dysfunction that was present in the previously reported adult case, both individuals appear to have had similar reaction to the shunt’s Silastic tubing.

The dystrophic calcification seen in our patient may recur as a long-term complication since the shunt tubing is made of the same Silastic material as the original shunt, and his body may react to the material in the same way as it did to the previous catheter. Degradation of the catheter and dystrophic calcification were usually found in catheters that had been in place for 5 years or more, so our patient may eventually develop the same reaction years from now.1,2,6

Management for this type of problem should consist of external ventricular drainage of the abnormal fluid followed by complete removal of the affected shunt system if possible and placement of a new system distant to the affected system.

In conclusion, this case shows an unusual presentation of a not uncommon phenomenon associated with long-term shunting.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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