case report

Pseudotumor cerebri in a child receiving peritoneal dialysis: recovery of vision after lumbo-pleural shunt

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A 9-year-old boy with end-stage renal disease who was receiving continuous ambulatory peritoneal dialysis (CAPD) presented with acute visual loss and was found to have papilledema. Neuroimaging and cerebrospinal fluid (CSF) analysis were normal. The lumbar puncture opening pressure was 290 mm of water so the diagnosis of pseudotumor cerebri (PTC) was entertained. Medical treatment was not an option because of renal insufficiency; neither was lumbo-peritoneal shunting, because of the peritoneal dialysis. After a lumbo-pleural shunt was placed, there was marked improvement in symptoms. The lumbo-pleural shunt is a reasonable option for treatment for PTC in patients on CAPD who require a CSF divergence procedure.

Pseudotumor cerebri (PTC), or idiopathic intracranial hypertension (IIH), is characterized by headache, papilledema and increased cerebrospinal fluid (CSF) pressure, with normal CSF content and normal neuroimaging. It usually has a favorable prognosis, but if left untreated, may cause permanent visual loss. The exact pathophysiology remains uncertain, though current theories as to etiology revolve around elevated sagittal sinus pressure, believed to result from extracellular edema. This may cause partial compression of the major venous sinuses[1] and obstruction of CSF absorption or venous outflow.[2] In prepubertal children with intracranial hypertension, a number of risk factors have been described. These include obesity,[3] systemic lupus erythematosus,[4] anemia (including iron deficiency anemia),[5] sickle-cell anemia,[6] hypothyroidism[7] and hyperthyroidism.[8] Various medical treatments have also been implicated in the etiology of PTC (for example, growth hormone therapy,[9] vitamin A in hypervitaminosis A[10] and corticosteroid withdrawal.[11] There have also been reports suggesting that children with chronic impairment of renal function may be at greater risk of developing PTC.[12] Acetazolamide inhibits choroid plexus carbonic anhydrase and thus decreases CSF production and has been used widely as a medical treatment for PTC.[13] Surgical treatment includes lumbo-peritoneal shunting[14] and optic nerve sheath decompression (ONSD).[15] To the best of our knowledge, the use of lumbo-pleural shunting to divert CSF in a PTC patient has never been previously reported.

CASE

The patient was a 9-year-old boy with end-stage renal disease secondary to hypoplastic kidneys who was being treated by continuous ambulatory peritoneal dialysis (CAPD). He also had chronic hepatitis B and was deaf and mute. The patient's renal-replacement medications included lisinopril, minoxidil, amlodipine, propranolol, sodium polystyrene sulfonate (Keyexalate), 1-alpha calcidiol, calcium carbonate, erythropoietin and ferrous sulfate. One day prior to admission, the family noted that he seemed to have impaired vision. The mother noted that her son was running into objects and walls like a blind person. Also, he would not reach for objects when given to him, but when placed in his hand, he would handle an object properly. The mother also noted that he was pointing to his eyes, gesturing that he was not able to see her. There was no history of trauma

or vomiting or change in sensorium and he had no abnormal movements. There was a history of apparent headache; being unable to communicate well, he would point to his head and appear distressed.

On examination, he was alert, with a Glasgow Coma Scale (GCS) score of 15/15. His weight was 15.6 kg (<5th centile for age); height, 110 cm (<5th centile for age). His temperature was 36.8°C; respiratory rate, 18/minute; pulse rate, 110/minute; blood pressure, 110/67 mm Hg. The left pupil was 7 mm dilated and not reactive to light. The right pupil was 7 mm dilated and sluggishly reacting to light. Fundoscopy showed severe bilateral papilledema. Extra-ocular movements were normal, but he could neither fix nor follow a test object. He did not blink to shined light nor to a threatening hand waved before him. The other cranial nerves, the motor examination and the general physical examination were normal. A CT scan of the brain was normal. A lumbar puncture (LP) showed an opening pressure of 290 mm of water. The patient showed improvement in vision immediately after LP. He started making eye contact with the examiner, and was reaching for objects that were given to him. CSF analysis showed: WBC, 1; RBC, 13; protein, 0.18 g/L; glucose, 4.1 mmol/L; and culture was negative.

He was admitted to the pediatric intensive care unit (PICU) for observation, and developed hypotension, with the blood pressure falling to a low of 70/40 mm Hg, which was treated by fluid boluses and later a dopamine drip to a maximum of 10 μg/kg/min. There was no clear explanation for the hypotension, though hypovolemia was suspected. While in the PICU, he developed another episode of apparent deterioration in vision, which improved after another LP. Dopamine was weaned slowly over 3 days. Echocardiography was normal. Ophthalmology evaluation revealed bilateral papilledema, flat retina and macula. Brain magnetic resonance imaging and venogram were normal, with no signs of venous thrombosis. His condition stabilized, and he was discharged from the PICU to a regular ward. Laboratory studies showed the following: hemoglobin, 9.3 g/dL; blood urea nitrogen, 31 mmol/L (normal range, 2.1-7.1 mmol/L); creatinine, 857 μmol/L (normal range, 27-62 μmol); serum glucose, 5.5 mmol/L; thyroid-stimulating hormone, 1.01 MIU/L (normal range, 0.35-5.5 MIU/L); free T4, 20 pmol/L (normal range, 11.5-22.7 pmol/L); cortisol level, 866 nmol/L (normal range, 119-618 nmol/L); vitamin A level, 3.13 μmol/L (normal range, 1.02-2.72). Parathyroid hormone was 97 pg/mL (normal range, 12-72); serum iron, 4 μmol/L (normal range, 9-21 μmol/L); serum ferritin, 129 μg/L (normal range, 22-322 μg/L). Subsequently, he developed two other episodes of loss of vision. In each of these, the symptoms improved after LP. Management options were limited because of renal failure and peritoneal dialysis. The renal failure precluded the use of acetazolamide. Similarly, lumbo-peritoneal shunting was not an option because of the peritoneal dialysis. The procedure of optic nerve sheath decompression was not available in our institution. A lumbo-pleural shunt was eventually performed. In the immediate postoperative period, there was improvement in his vision. He was discharged after 5 days in stable condition. Follow-up chest x-ray showed no evidence of pleural effusion. Twelve months later, the papilledema had completely resolved and his vision had returned to normal.

DISCUSSION

There are a few case reports that mention uremia as a predisposing factor in the development of PTC.[12] These suggest that the fluid overload, increased cerebral blood flow and anemia, associated with the condition, may be responsible for the development of PTC. Other factors associated with uremia (for example, chronic hypertension, episodic hypotension, uremic optic neuropathy and vitamin A toxicity) may contribute to the visual impairment. This makes the treatment of PTC in uremic patients especially challenging.[12] In addition, our case was particularly challenging because the patient was a young child who was deaf, thus making communication difficult. Also, at presentation, there was loss of vision, indicating that the condition was already significantly advanced. The initial LP findings tended to confirm the diagnosis of PTC. The hypotension that developed within the first 3 days of presentation was probably due to depleted intravascular volume. Its improvement with fluid replacement and inotropic support and the absence of any other etiology would support this diagnosis. His symptoms recurred, but they responded to repeated lumbar punctures. Medical treatment with acetazolamide is contraindicated in renal-failure patients. Although an established and effective treatment for PTC,[14] lumbo-peritoneal shunt was excluded because of the high risk of infection in the presence of peritoneal dialysis. Our patient was young and also deaf. Loss of his vision would be particularly devastating, since he had few other ways of communicating. Thus a CSF diversion procedure was considered vital for saving his vision. Optic nerve sheath decompression (ONSD) was an option, but in a group of predominantly adult patients treated with this modality, one third were reported to have developed complications, which included diplopia, ophthalmoplegia and traumatic optic neu-

ropathy.[15] In another relatively small study, ONSD was described as being a safe procedure for the treatment of childhood PTC patients.[16] However, the literature is sparse regarding its efficacy in treating young children, especially in the prepubertal age group. The anatomically small optic nerve and orbital cavity may make the procedure difficult. In addition, the relative rarity of the syndrome in children limits the procedural experience of many pediatric ophthalmologists. In the management of hydrocephalus, ventriculo-pleural shunting has proven to be a viable alternative to peritoneal and atrial shunts when for various reasons, these sites are unsuitable.[17] We were not aware of any previous reports of experience in draining CSF from the lumbar region to the pleural cavity. However, in this case, where the therapeutic options were limited, this seemed a reasonable approach. Accordingly, a lumbo-pleural shunt was created. The procedure proved to be easy, effective and safe, requiring no revision since installation. The patient had immediate improvement in his vision, which went on to return to normal over the next 12 months. Although this procedure apparently has not been reported previously, it would seem to be a valuable therapeutic option in the treatment of similar cases.

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