Ventriculoperitoneal shunt dysfunction in a patient presenting with neurogenic pulmonary edema

Case report

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✓The authors report on a patient with dysfunction of a ventriculoperitoneal shunt who presented with two episodes of neurogenic pulmonary edema within the space of a few months. The edema resolved on correction of the shunt dysfunction. Because neurogenic pulmonary edema may be a rare consequence of shunt dysfunction, it is important to recognize this unusual association and provide appropriate diagnostic measures and treatment.

KEY WORDS • pulmonary edema • hydrocephalus • shunt malfunction

DYSFUNCTION of a CSF shunt usually becomes apparent with clinical signs and symptoms of increased ICP. Such signs and symptoms include headache, vomiting, impaired level of consciousness, gait disturbance, impaired cognition, and urinary incontinence. Neurogenic pulmonary edema is another well-known complication of increased ICP. It has been reported to occur in a wide variety of settings, most frequently in conjunction with craniocerebral trauma, hemorrhage, and after seizures.1,2 It has not yet been described, however, as a presenting symptom of VP shunt dysfunction. We present the case of a patient in whom neurogenic pulmonary edema developed twice due to dysfunction of a VP CSF shunt. The pulmonary edema resolved rapidly after revisions of the shunt system.

Case Report

History. This 29-year-old woman had a congenital myelomeningocele, which resulted in paraplegia distal to the L-3 level. A few weeks after she had been born, hydrocephalus associated with Arnold–Chiari malformation was treated by implantation of a ventriculoatrial CSF shunt. Six years later, that shunt system was replaced by a VP shunt. There was no history of cardiopulmonary disease.

Examination. At the age of 29 years, the patient was admitted to the hospital with symptoms including shortness of breath, headache, and neck pain. Chest x-ray films showed bilateral pulmonary edema, but there were no hints of pneumonia or pulmonary embolism. Admission CT scans of the head revealed enlargement of the ventricular system (Fig. 1 left).

Shunt Revision and Postoperative Course. Replacement of the obstructed VP shunt system resulted in rapid improvement of her headache and neck pain, as well as of her pulmonary distress. Diagnostic workup did not reveal a plausible cause for development of pulmonary edema at that time. She was discharged from the hospital 10 days after surgery and resumed her work as a secretary.

Readmission and Examination. Four months after the shunt revision, the patient was readmitted with dyspnea, headache, neck pain, and paresthesias of her arms. She was sleepy, but reacted adequately to painful stimuli. On pulmonary examination there was audible rale accompanied by bubbling noises. Chest x-ray films confirmed extensive bilateral pulmonary edema. Clinical and laboratory signs of cardiorespiratory failure were evident. The patient underwent intubation and received mechanical ventilation with 100% O2 and positive end-expiratory pressure of 14 cm H2O. Arterial blood gas analysis indicated marked respiratory acidosis: pH 7.08, PaCO2 63 mm Hg. Protein concentration in the pulmonary fluid was 47.3 g/L and it was 48 g/L in the serum, indicating protein exudation. Cardiac ultrasonography showed diffuse hypokinesia without left-sided ventricular dilation. The ejection fraction was reduced to 35%. There was no history of aspiration, intoxication, or trauma.

Repeated CT scans of the head again revealed asymmetrical dilation of the ventricles with dislocation of the ven-

Abbreviations used in this paper: CSF = cerebrospinal fluid; CT = computed tomography; ICP = intracranial pressure; VP = ventriculoperitoneal.
tricular catheter. No CSF could be aspirated by puncture of the Rickham reservoir.

**Second Shunt Revision.** On revision of the proximal part of the shunt, ICP measurement revealed increased pressure, recorded at 35 cm H₂O. With the aid of a neuroendoscope, it was demonstrated that only the tip of the ventricular catheter was located in the ventricle, but that the openings of the catheter were obstructed. A new ventricular catheter was inserted with endoscopic guidance. Bacterial cultures of the CSF were sterile.

**Postoperative Course.** After shunt revision surgery, the patient recovered gradually from cardiopulmonary failure. One week later, mechanical ventilation was removed with no problems. Follow-up CT scans obtained 2 months later demonstrated marked decrease of the hydrocephalus (Fig. 1 right). Two weeks later she returned home. There were no further episodes of pulmonary edema or shunt dysfunction during a follow-up period of 2 years.

**Discussion**

Neurogenic pulmonary edema after sudden increases of ICP due to brain injury, hemorrhage, or seizure is a well-reported condition. In contrast, only a few cases of neurogenic pulmonary edema associated with chronic hydrocephalus have been reported. One was in a patient with a colloid cyst, and another was in a patient who suffered from hydrocephalus due to aqueductal glioma. To our knowledge, neurogenic pulmonary edema has not yet been reported in a patient with dysfunction of a CSF shunt system. Pathophysiological concepts for the mechanisms of neurogenic pulmonary edema have been outlined in detail. Despite extensive clinical and experimental studies, however, no definitive, conclusive concept has emerged to explain the clinical manifestation of this condition. A sudden increase of ICP seems to play a pivotal role under these circumstances. Subsequent hemodynamic changes due to massive sympathetic discharge and increased venous resistance have also been discussed as factors in the development of neurogenic pulmonary edema. Neural structures that are thought to be involved include the hypothalamus, the nuclei of solitary tract, the area postrema, and areas A1 and A5 of the medulla. These structures form a network in the regulation of cardiovascular responses of the autonomic nervous system.

In our patient, another contributing condition may have been her Arnold–Chiari malformation, resulting in direct compression of the medulla oblongata during the periods of shunt dysfunction. As discussed by Rickert et al., sudden compression of neuronal pathways within the brainstem may result in subacute cardiopulmonary disturbances. The common pathway, finally, is the excessive increase of vascular resistance, left ventricular failure, and pulmonary edema. In addition, disturbance of endothelial permeability results in accumulation of protein-rich edema fluid. Non-cardiogenic pulmonary edema has a high protein content because the more permeable microvascular membrane has a reduced capacity to restrict the outward movement of larger molecules such as plasma proteins.

One of the remarkable aspects in our case is the repeated occurrence of neurogenic pulmonary edema, with shunt dysfunction on two occasions. We think it is important to emphasize that neurogenic pulmonary edema may develop in patients suffering from a condition that is not associated with the sort of structural brain damage that is seen in trauma.
ma, tumor, or hemorrhage. The temporal series of events that occurred twice and the subsequent resolution of pulmonary edema both times following shunt correction in our patient indicate a causal relationship rather than a coincidental association. Furthermore, her clinical course would suggest a relationship between the level of ICP and the severity of the neurogenic pulmonary edema. Such a relationship has also been observed in experimental studies.\(^7\) Whereas the first manifestation could be managed without any specific cardiopulmonary treatment, during the second event cardiopulmonary treatment including mechanical ventilation was necessary.

**Conclusions**

In rare cases, pulmonary edema may be considered an initial symptom of shunt dysfunction in patients in whom a CSF shunt system has been placed. Differential diagnostic workup in patients with pulmonary edema and shunt-treated hydrocephalus should include immediate examination of the shunt system.

**References**


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