The Open Access Journal of Science and Technology
Vol. 3 (2015), Article ID 101133, 4 pages
doi:10.11131/2015/101133

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Case Report

Ventriculopleural Shunt Surgery in A Patient with Multiple Skeletal and Neurodevelopmental Anomalies: A True Perioperative Challenge

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Received 13 September 2014; Accepted 4 December 2014

Academic Editor: Adriano Bechara De Souza Hobaika

Abstract. *Introduction*: Ventriculopleural shunt placement for decompression of hydrocephalous is an uncommon procedure. The usual indications are adhesions, infection, thrombosis or obliteration of the more commonly done ventriculoperitoneal shunt or an anatomical non-availability of the peritoneum. We present a clinical case of a patient with congenital hydrocephalous complicated by multiple skeletal and neurodevelopmental anomalies who presented for thoracoscopic placement of a ventriculopleural shunt. The anesthetic challenges and perioperative management constituted a complex multi-disciplinary challenge. *Case presentation*: We present a clinical case of a 42 years old woman with congenital hydrocephalous complicated by multiple skeletal and neurodevelopmental anomalies who presented for thoracoscopic placement of a ventriculopleural shunt. Some of the perioperative concerns included initiation and maintenance of one-lung ventilation in a patient with short stature, dysmorphic skeletal features, severe kyphoscoliosis, restrictive underlying lung disease and multiple neurodevelopmental midline defects. *Conclusion*: Our presentation highlights the special challenges in this patient based on the need for lung collapse in a severely short statured individual with a dysmorphic severely kyphoscoliotic thoracic cavity along with underlying restrictive lung disease and background neurodevelopmental midline defects.

Keywords: Hydrocephalous; Ventriculopleural; Shunt; Surgery; Anesthesia

1. Introduction

Surgical management of hydrocephalus commonly entails a shunt procedure with drainage to the peritoneum or right atrium. A ventriculopleural shunt is an uncommon shunt sometimes used for CSF drainage in patients when conventional sites are not suitable due to problems such as adhesions, infection, thrombosis or obliteration [1]. Typical patients are adults with multiple failed ventriculoperitoneal shunts and contraindications to ventriculo-atrial shunt placement. The congenital variant of hydrocephalus amongst other causes also has an association with multiple neurodevelopmental delays and skeletal deformities. We present one such case of a patient with multiple skeletal and neurodevelopmental anomalies who presented for thoracoscopic

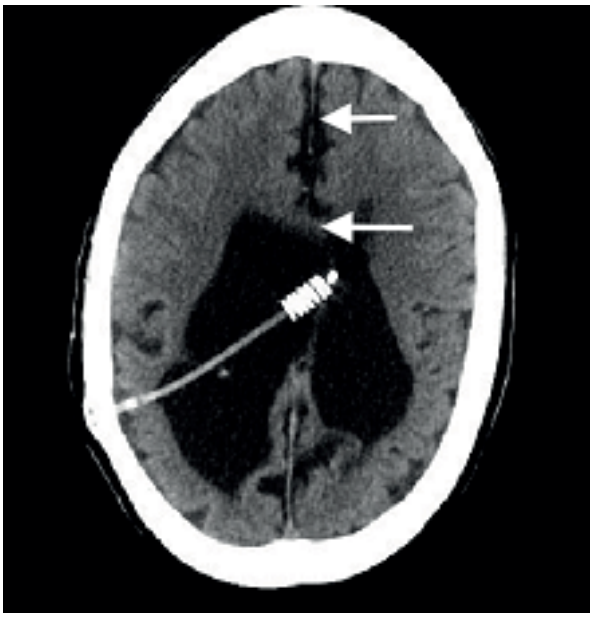

Figure 1: Axial CT scan of the brain showing dilated, parallel ventricles, with disproportionately large occipital and lateral horns of the lateral ventricles (colpocephaly). The non-functional shunt catheter is also seen within the ventricles. In addition, the inter-hemispheric fissure is almost in total continuity (white arrowheads). These are all characteristic of a partial agenesis of the corpus callosum.

placement of a ventriculopleural shunt. The anesthetic challenges and perioperative management constituted a complex multi-disciplinary challenge.

2. Case Report

A 42 year old, 125 cm tall, female with a weight of 35.5 kg (78 lb 4.2 oz) presented for a video-assisted thoracoscopic surgery (VATS) and simultaneous ventriculostomy for surgical placement of a ventriculopleural shunt. She has a past medical history of spina bifida with consequent paraplegia, type II Chiari malformation and partial agenesis of the corpus callosum (Figure 1). She had increasing shunt-dependent hydrocephalus since childhood. In addition, she also had dysmorphic skeletal features, severe kyphoscoliosis with underlying restrictive lung disease and generalized large joint contractures. Her baseline oxygen saturation (Spo2) was 92–95% on continuous 4 liter/minute home oxygen. Her prolonged and complicated surgical history started with a right occipital ventriculoatrial shunt placed around the time of her birth for hydrocephalus, which was revised at age 9 to a ventriculoperitoneal shunt. Recently, she had multiple shunt revisions with dehiscence and breakdown of the anterior chest wall wound and distal shunt malfunction.

The planned surgical procedure entailed an internalization of the distal shunt revision with ventricular pleural shunt creation via thoracoscopic assistance. Perioperative concerns

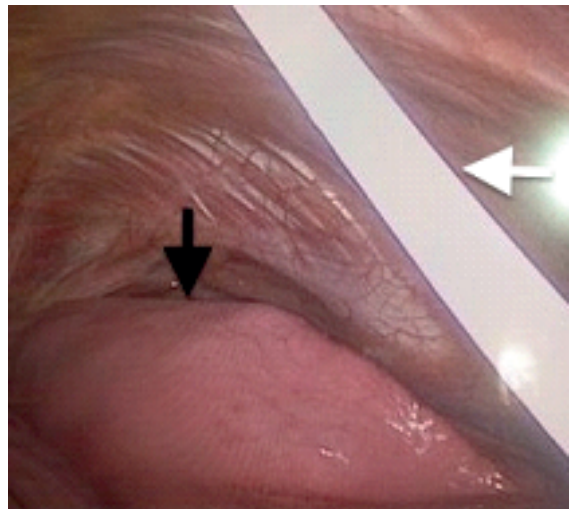

Figure 2: The patient as seen on the operating room table, with a 35 Left Double-lumen endotracheal tube. The short stature and severe kyphosis are also evident.

included initiation and maintenance of one-lung ventilation in a patient with short stature, dysmorphic skeletal features, severe kyphoscoliosis, restrictive underlying lung disease and multiple neurodevelopmental midline defects.

In the operating room a left radial arterial line and two wide bore peripheral intravenous access lines were secured awake. After adequate preoxygenation intravenous induction of anesthesia was carried out and a 35 French (Fr) 11.7mm Mallinckrodt Left sided Endobronchial Tube (Covidien, Mansfield, MA)was placed. Correct positioning of the tube was confirmed via bronchoscopic and clinical examination and the tube was secured at 25 cm at the lip. The patient was positioned in a left lateral decubitus position for the surgery and a final reconfirmation of correct tube placement and adequate lung isolation was done via a bronchoscope. Inhalational anesthesia was maintained with 1-1.5 MAC of sevoflurane in an air/oxygen 1:1 mixture. This was later switched to 100% oxygen after collapse of the right lung was initiated. Analgesia was supplemented with iv fentanyl 1–2 micrograms/kg as needed. As the neurosurgical team tunneled their shunt, the tunneling device was visualized thoracoscopically with carbon dioxide insufflation of the right pleural cavity and the thoracic surgery team then successfully completed catheter placement. (Figure 2) She remained stable throughout the procedure with acceptable cardiovascular and respiratory parameters. She was transferred intubated to the Post-Anesthesia Care Unit for delayed extubation after full reversal of neuromuscular blockade was confirmed. A small basilar right pneumothorax due to residual carbon dioxide was present; this resolved on serial imaging. Multiple subsequent shunt series radiology films confirmed correct placement of the shunt in the pleural cavity. (Figure 3) The patient made a progressive recovery and was discharged home on post-operative day one. We continued to follow

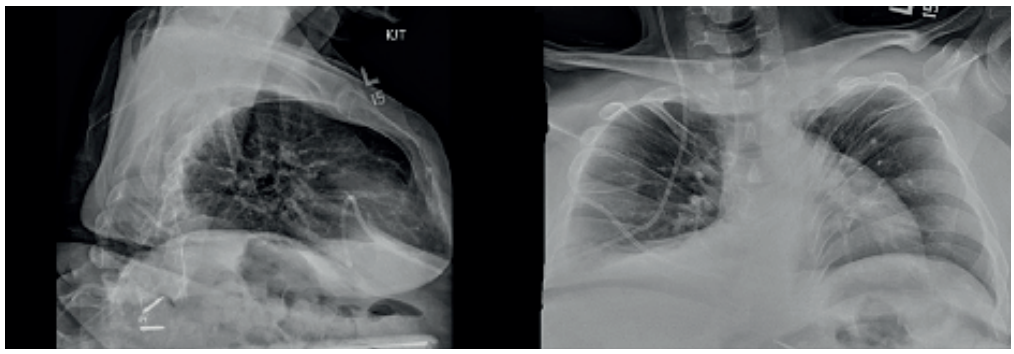

Figure 3: Thoracoscopic view of the distal shunt catheter (white arrowhead) with the collapsed right lung in the background (black arrowhead).

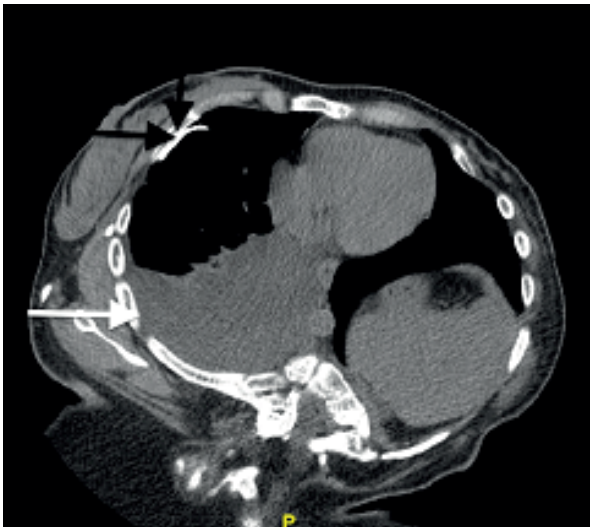

Figure 4: Post procedure chest x-rays (AP and lateral films) showing correct placement of the ventriculopleural shunt catheter in the right thoracic cavity.

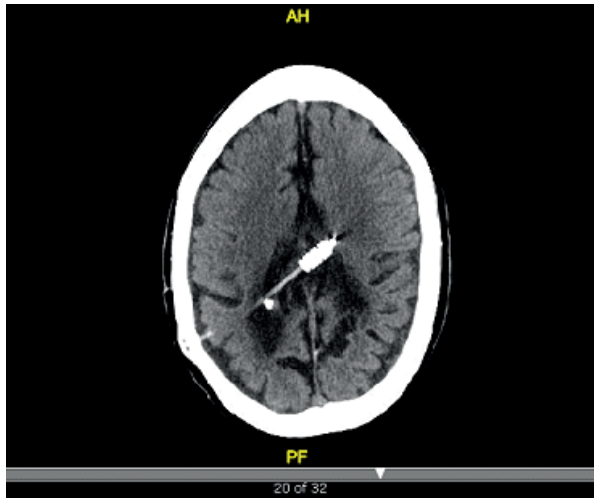

Figure 5: Post-procedure (2weeks) CT chest without contrast showing a moderate right-sided pleural effusion (white arrowhead). Also seen is the dorsal end of the shunt catheter at the level of the right middle lobe (black arrowheads).

up our patient, who returned to the primary service with complaints of respiratory impairment after two weeks. On imaging a moderate size right pleural effusion resultant lung base compression with the shunt catheter in proper position was seen on a CT scan of the chest (Figure 4). This necessitated an interventional radiology guided thoracentesis of the effusion with consequent improvement in ventilatory status.

3. Discussion

Ventriculopleural shunts are uncommon shunt procedures, typically performed as salvage procedures after multiple failures of the commonly done ventriculoperitoneal shunts. Though both open surgical incision and a thoracoscopic approach are described, at Cleveland Clinic most of these procedures are done via the Video Assisted Thoracoscopic (VATS) technique. Our patient had several perioperative issues that made this a special perioperative challenge. First and foremost, achieving and maintaining adequate lung isolation in a short stature patient (125cm) , with a severely kyphotic and scoliotic vertebral anatomy was difficult. Our choices were between a regular double lumen endobronchial tube (DLT), a bronchial blocker or a regular endotracheal tube guided with a flexible bronchoscope to the left main bronchus. We decided to proceed with a 35 left-sided double lumen tube for our first attempt, with the bronchial blocker and the regular tube as “Plan B” and “Plan C,” respectively. This decision was based on the familiarity of our anesthesia team in placement and management of lung isolation with a regular double lumen tube. There is no absolutely accurate method for selecting the correct size tube. A properly sized DLT is one in which the main body of the tube passes without resistance through the glottis and advances easily within the trachea and in which the bronchial component passes into the intended bronchus without difficulty [3]. It has been seen that left bronchial or tracheal width measurement on CT

radiography or using an ultrasound are superior methods of selecting an appropriately sized double lumen tube [4, 5]. Traditional teaching has used height as the major correlation with correct sizing of the double lumen tube. Based on this premise we select 35F and 37F in female patients (< and >160cm respectively) and 39F and 41F in male patients (< and >170cm respectively) at our institution [6]. Though we had the option of starting with a 32 F tube we choose to go with a relatively larger tube first and see if we could position it correctly without trauma. A 35 Fr tube also allowed us ease of maneuverability of our 3.5mm flexible bronchoscope and better respiratory compliance parameters later on.

Secondly, our patient had restrictive pulmonary disease and severe kyphoscoliosis. In view of this, we maintained a pressure-controlled mode of ventilation, and titrated our airway pressures to prevent barotrauma in the setting of poor baseline ventilatory compliance. This was at the cost of some permissive hypercapnia that the patient tolerated well.

Thirdly, initiation of one lung ventilation and carbon dioxide insufflation into the right thoracic cavity was a further insult to the patient's poor pulmonary function. However, we were able to maintain an acceptable oxygen saturation of 92–95 percent on a 100 percent oxygen (which was similar to the patient's baseline) all through the procedure.

Fourthly, we were concerned about post-operative recovery and post-extubation problems. Based on the fact that there was some airway instrumentation and possible airway edema consequent to that, we waited till the patient was totally awake and reversed of muscle relaxant in the PACU, before doing a watchful extubation with the difficult airway cart nearby. A post-procedure chest x-ray with basilar pneumothorax was seen, but since the patient was generating good tidal volumes and maintaining oxygenation we extubated the airway and followed up with serial chest films, which as expected showed complete resolution in the next 24 hours.

Finally, we were interested in a continued follow up of our patient since some of the respiratory complications of this type of shunt procedure may occur in the delayed post-operative period. That this patient had a right - sided pleural effusion of moderate size (symptomatic 2 weeks post-procedure) meant that the shunt was functioning adequately. Further confirmation of this was also seen on imaging the brain that showed resolution of the ventriculomegaly that was seen pre-operatively. (Figure 5) Shunt related complications could be generalized or specific based on the anatomical location of the shunt. General complications are usually mechanical and or functional as such occlusion or migration of the catheter tip. However, the ventriculopleural catheter has been associated with some specific complications such as respiratory insufficiency from pneumothorax, tension hydrothorax, symptomatic pleural effusions, empyema and pneumocephalus. There is also an isolated report of cardiac tamponade and heart failure from a malfunctioning shunt [1, 6–10]. Therefore, the immediate post-shunt period should be one where the patient is monitored for a prolonged period of time in the PACU and later on in the high-dependency unit (HDU) or a step-down unit.

4. Conclusions

Ventriculopleural shunt procedures are a challenge for the entire surgical and perioperative team. These patients need good pre-operative optimization, accurate lung isolation and collapse during surgery and vigilant post-operative monitoring. Our case report highlights the special challenges in this patient based on the need for lung collapse in a severely short statured individual with a dysmorphic severely kyphoscoliotic thoracic cavity along with underlying restrictive lung disease and background neurodevelopmental midline defects.

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