CASE REPORT

**Superior vena cava syndrome with retropharyngeal edema as a complication of ventriculoatrial shunt**

Mohammed S. Al-Natour, Pouya Entezami, Munier M. S. Nazzal, Andrew B. Casabianca, Ragheb Assaly, Kalen Riley & Daniel Gaudin

1Department of Radiology, University of Toledo Medical Center, Toledo, Ohio
2Department of Surgery, Division of Neurosurgery, University of Toledo Medical Center, Toledo, Ohio
3Department of Surgery, Division of Vascular Surgery, University of Toledo Medical Center, Toledo, Ohio
4Department of Anesthesiology, University of Toledo Medical Center, Toledo, Ohio
5Department of Internal Medicine, University of Toledo Medical Center, Toledo, Ohio

**Correspondence**
Daniel Gaudin, Department of Surgery, Division of Neurosurgery, University of Toledo Medical Center, 3000 Arlington Avenue, Toledo, OH 43614. Tel: (419) 383 3547; Fax: (419) 383 6570; E-mail: daniel.gaudin@utoledo.edu

**Funding Information**
No sources of funding were declared for this study.

Consent to publish has been obtained.

Received: 3 May 2015; Revised: 11 June 2015; Accepted: 24 June 2015

**Clinical Case Reports 2015; 3(10): 777–780**
doi: 10.1002/ccr3.331

**Key Clinical Message**
Thirty-seven-year old female with hydrocephalus managed by a ventriculoatrial (VA) shunt presented with upper body edema, dysphagia, and headache. Imaging demonstrated thrombosis of the superior vena cava (SVC). Direct catheter thrombolysis led to resolution of thrombus burden. Superior vena cava thrombosis is a rare consequence of VA shunting and must be managed emergently.

**Keywords**
Retropharyngeal edema, superior vena cava syndrome, ventriculoatrial shunt.

**Introduction**
Despite having several potential complications, ventricular shunting for the treatment of hydrocephalus remains one of the most common neurosurgical procedures. The risk of shunt failure due to obstruction or disconnection is well characterized [1]. For ventriculoatrial (VA) catheterization in particular, the incidence of infection and cardiovascular compromise (including perforation, thrombophlebitis, and pulmonary embolus causing pulmonary hypertension) is of significant concern [1]. However, superior vena cava (SVC) syndrome secondary to VA shunting is a devastating complication which can rarely occur [2, 3].

Acute or subacute occlusion of the SVC results in facial and upper extremity edema, dysphagia, pain, and dyspnea, collectively referred to as SVC syndrome [4]. Common etiologies for this process include tumor, mediastinal fibrosis, indwelling catheters, and pacemaker wires [1, 4]. The incidence of this obstructive process due to VA shunting is rare, with only a limited number of cases reported in the literature [2, 5].

**Case Report**
We present the case of a 37-year-old African American female with medical history significant for obesity (BMI of 39), Dandy-Walker malformation, and congenital hydrocephalus managed by two shunts, including one VA shunt and one ventriculopleural shunt. Her surgical history was significant for multiple shunt revisions since initial shunt placement at 6 months of age; her most recent VA catheter had been in place for 20 years. She presented to our Emergency Department with progressive facial swelling for the past 3 weeks, neck and bilateral upper extremity edema, dysphagia with solid foods, and headache (Fig. 1A). Of note, she had recently begun taking combined oral contraceptive pills (OCPs) for menometr-
orrhagia. Physical examination revealed mild hypertension, absence of fever, and significant engorgement of the face and neck. Besides her presenting dysphagia she was unable to lay supine. There was no evidence of Horner’s syndrome or paralysis of the vocal cords (notably no hoarseness or stridor). Physical examination and laboratory findings were otherwise unremarkable.

Imaging shunt series revealed disruption in the ventriculopleural shunt in the neck, widening of the prevertebral soft tissue stripe, prominent azygous arch, and an intact VA shunt with its distal tip in the SVC. CT scans of the head and neck with contrast were obtained to better characterize the retropharyngeal swelling and assess ventriculopleural shunt disruption. These demonstrated a retropharyngeal fluid collection compromising the airway, extensive venous collaterals in the upper mediastinum and paraspinal regions, a prominent azygous arch, and confirmed the ventriculopleural catheter disruption (Fig. 2). SVC thrombosis was considered as a possible etiology, and a CT of the chest with contrast confirmed thrombosis of the catheter-bearing SVC and the azygous vein. Imaging was negative for thoracic malignancy. She was taken promptly to the angiogram suite where she was fiberoptically intubated awake using a reinforced endotracheal tube before induction of general anesthesia due to her high risk airway.

Angiogram revealed acute occlusive thrombus of the SVC at the level of the distal tip of the VA catheter and thrombosis of the azygous vein (Fig. 3A). Extensive venous collaterals were visible again in the mediastinum and paraspinal region. Direct catheter thrombolysis using tissue plasminogen activator (tPA) was initiated using a multi-side hole catheter with simultaneous intravenous heparin administration. After 24 h there was resolution of thrombus burden in both vessels, with remnant severe SVC stenosis at the catheter tip (Fig. 3B and C). The stenosis was crossed with difficulty and balloon angioplasty was performed (10 mm × 4 cm and 12 mm × 4 cm balloons) relieving the catheter, which was removed surgically and revised at a later date to the pleura. Immediately after, the SVC was stented (18 mm × 4 cm bare metal balloon expandable stent), (Fig. 3D).

The patient’s symptoms improved significantly following the procedures (Fig. 4). However, her hospitalization was complicated by hemoperitoneum and alveolar hemorrhage, which resolved after laparoscopic evacuation and multiple blood transfusions. Anticoagulation was maintained to assure stent patency and prevent rethrombosis. She was extubated and discharged shortly thereafter in stable condition (Fig. 1B).

**Discussion**

Ventriculoatrial shunting of CSF for the treatment of hydrocephalus is a relatively common neurosurgical procedure that can be performed safely. Nevertheless, there is a significantly high risk of associated complications. Though rare, SVC syndrome may occur – as it did with our patient – in patients who have a VA shunt in place. The incidence of this is unknown since it is seldom reported in the literature. Due to the smaller size of the VA catheter, the risk of developing SVC syndrome is considerably lower than when using larger indwelling catheters. The etiology of our patient’s SVC thrombosis was multifactorial, including SVC stenosis secondary to long-
term VA catheter implantation, and recent OCP initiation in an obese patient over the age of 35. Hereditary hypercoagulability workup was negative.

Superior vena cava syndrome results from disruption of blood flow through the SVC to the right atrium [4, 6]. Historically, infections were deemed the primary cause; however, significant improvements in antibiotics and infection control led to a gradual decrease in infections as the primary etiology. Over the second half of the last century thoracic malignancy – specifically small cell and non-small cell carcinomas of the lung – predominated as the primary cause, accounting for up to 80% of cases through either invasion or external compression [6–9]. Benign causes of SVC syndrome (mediastinal fibrosis being the most common) were thought to account for only about 20% of the cases, though there has been a significant increase in benign causes (now accounting for up to 40% of cases) over the last two decades [10–12]. This is thought to be due to the increase in the use of indwelling central venous catheters and cardiac pacemakers [13].

Although the diagnosis of SVC syndrome is normally achieved by proper history taking and physical examination, there has been an increased reliance on imaging as a primary diagnostic modality in emergency rooms. Improved knowledge of possible extrathoracic imaging...
findings might aid in reaching the diagnosis in the appropriate clinical settings. Retropharyngeal fluid from venous hypertension secondary to SVC occlusion is a rare finding and has been described only once in a case secondary to small cell lung cancer [14]. We are uncertain of the reason for the rarity of this finding, though we postulate that this can be attributed to the prior infrequent imaging of the neck in such cases.

Direct catheter thrombolysis followed by endovascular angioplasty and stenting are emerging modalities of treatment in cases of thrombosis and SVC stenosis secondary to a benign etiology (such as an indwelling intravenous catheter). These options appear to be very promising and safe alternatives to open surgical bypass [15].

A principal used in other indwelling catheters during the time of implantation is to ensure that the tip be placed distally enough that it remains freely mobile, preventing stagnation of flow and thereby preventing thrombosis. Over time, as younger patients mature and grow into adulthood the tip of the catheter may move out of the atrium and into the great vessels, increasing the risk of thrombosis. Ensuring that the tip of VA catheters is implanted into the vessel at an adequate length – despite its relatively narrow diameter – and monitoring it during a patient’s continued growth will help ensure it remains mobile distally, decreasing the risk of this complication.

### Conclusion

Though rare, SVC thrombosis as a consequence of VA shunting is a distressing complication which must be managed emergently. The rarity of this outcome may delay proper diagnosis which may further postpone appropriate treatment, resulting in increased morbidity for the patient. Therefore, it is important to consider this complication when treating patients with symptoms of SVC syndrome who have received a VA shunt in order to maximize their outcomes.

### Conflict of Interest

The authors declare that they have no conflicts of interest concerning this article.

### References

1. Greenberg, M. S. 2010. Handbook of neurosurgery, 7th ed. Greenberg Graphics, Inc., Tampa, FL.
2. Vik, A., S. Kumar, K. Singh, and H. JB. 2003. Local thrombolysis with stent implantation in a patient with vena cava superior syndrome. Tidsskr. Nor. Laegeforen. 123:2049–2050.
3. O’Shea, P. A. 1978. Inferior vena cava and hepatic vein thrombosis as a rare complication of ventriculoatrial shunt. Case report. J. Neurosurg. 48:143–145.
4. Funaki, B. 2006. Superior vena cava syndrome. Semin. Intervent. Radiol. 23:361–365.
5. Forrest, D., and D. Cooper. 1968. Complications of ventriculo-atrial shunts. J. Neurosurg. 29:506–512.
6. Schechter, M. M. 1954. The superior vena cava syndrome. Am. J. Med. Sci. 227:46–56.
7. Cardiopulmonary Syndromes. National Cancer Institute, 2005 (accessed 11 March 3 2015).
8. Flounders, J. A. 2003. Oncology emergency modules: superior vena cava syndrome. Oncol. Nurs. Forum 30:E84–E90.
9. Ahmann, F. R. 1984. A reassessment of the clinical implications of the superior vena caval syndrome. J. Clin. Oncol. 2:961–969.
10. Klassen, K. P., N. C. Andrews, and G. M. Curtis. 1951. Diagnosis and treatment of superior-vena-cava obstruction. AMA Arch. Surg. 63:311–325.
11. Nieto, A. F., and D. B. Doty. 1986. Superior vena cava obstruction: clinical syndrome, etiology, and treatment. Curr. Probl. Cancer 10:441–484.
12. Parish, J. M., R. F. Jr Marschke, D. E. Dines, and R. E. Lee. 1981. Etiologic considerations in superior vena cava syndrome. Mayo Clin. Proc. 56:407–413.
13. Rice, T. W., R. M. Rodriguez, and R. W. Light. 2006. The superior vena cava syndrome: clinical characteristics and evolving etiology. Medicine 85:37–42.
14. Chang, C. Y., Y. C. Lai, and S. C. Chang. 2012. Superior vena cava syndrome related fluid collection in retropharyngeal space. QJM 105:907–909.
15. Rizvi, A. Z., M. Kalra, H. Bjarnason, T. C. Bower, C. Schleck, and P. Gloviczki. 2008. Benign superior vena cava syndrome: stenting is now the first line of treatment. J. Vasc. Surg. 47:372–380.