

Superior vena cava syndrome caused by bronchogenic cyst: Report of a case

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Citation

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Abstract

Superior vena cava (SVC) syndrome may be caused by intrathoracic benign and malignant processes. Although bronchogenic cyst is a rare cause of the superior vena cava syndrome (SVCS), it should be considered because it is a potentially treatable lesion. In this report, we present clinical and radiological features of a case of symptomatic mediastinal bronchogenic cyst causing SVCS.

INTRODUCTION

SVCS is characterized by gradual, insidious compression or obstruction of the SVC. Although the syndrome can be life threatening, its presentation is often associated with a gradual increase in symptomatology (1). This syndrome may be caused by intrathoracic benign and malignant processes. The commonest cause of benign SVC obstruction is fibrosing mediastinitis, which accounts for 10% of overall cases (2). Bronchogenic cyst is a rare cause of the SVCS. Here, we report an unusual presentation of right upper mediastinal bronchogenic cyst presenting as a SVCS.

CASE REPORT

A 20-year-old male was referred to our hospital with signs and symptoms of the SVCS. Symptoms were gradual onset and had a progressive course. At the time of admission, he was dyspnoeic but hemodynamically stable. There was no history of trauma, or thoracic malignant process in the past. Physical examination, revealed slightly, but diffuse swelling of the face, neck and upper limbs, and bilateral jugular venous distention. Dilated superficial veins over the chest or abdomen were not determined. Chest radiograph showed rounded radioopaque shadow at right superior paramediastinal region (Fig. 1).

{image:1}

Multi-detector computed tomography angiography (MDCTA) was performed for further evaluation of the patient. MDCTA demonstrated a homogeneous-hypodens (attenuation measurements consistent with fluid) mass with

dimensions of 10 x 6 x 5 cm in the right upper mediastinum. It was compressing the SVC and trachea (Fig.2).

{image:2}

The patient was referred to cardiothoracic surgeons for further management. The mass was surgically removed. Final histopathological report revealed it, as a bronchogenic cyst. After surgery, there was a significant improvement in all symptoms, and the patient is symptom free in the 6 months of follow up.

DISCUSSION

SVCS is not a common clinical presentation. The clinical features of this syndrome have been discussed by various authors. Clinical symptoms may develop over several weeks or longer. Increased venous pressure leads to edema of the head, neck, and arms and may turn the patient a shade of blue with cyanosis. Other common symptoms include distention in the neck and chest veins and flushing. Edema may impede the function of the larynx or pharynx resulting in cough, dyspnea, dysphagia, and stridor. Cerebral edema may lead to mental status changes such as confusion, coma, or migraines. Cerebral edema is rare, but if it occurs it may be fatal (3,4). Although, the symptoms are insidious onset and slowly progressive in benign nature of this syndrome, sometimes the symptoms may appear rapidly, even in benign type, due to superadded infection and rapid enlargement of the lesion. These cases are require urgent intervention (5).

Most of the reported cases of SVC syndrome have been described in patients with malignant disease, although

benign conditions such as aortic aneurysm, mediastinitis, mediastinal tumors, cardiac pathology, pulmonary lesions, traumatic and miscellaneous causes have also been mentioned. The most common cause of malignancy-related SVCS is bronchogenic carcinoma, which accounts for nearly 80% of cases (1). Mediastinitis caused by various granulomatous processes is most common cause of the benign SVCS (5). Several report of bronchogenic cysts causing SVCS have also been described in the literature (5,6,7).

Bronchogenic cysts are a rare cause of mediastinal mass (7,8). The embryologic foregut differentiates into oesophagus and trachea during embryogenesis. Bronchogenic cysts occur along the differentiating pathway of the trachea and bronchial tree, and are thought to represent abnormal budding of foregut tissue. They are most often found in the mediastinum or lung, and sometimes in the neck (7). The location of the cyst depends on the embryological stage of development at which the anomaly occurs. When this abnormal buddings occurs during early development, the cyst tends to be located along the tracheobronchial tree. Cysts that arise later are more peripheral and may be located within the lung parenchyma (9,10).

Bronchogenic cysts are typically asymptomatic, with many detected incidentally (6). The diagnosis may be made with chest radiograph, CT and MRI, but definitive diagnosis is usually only achieved by surgical excision. CT provides the optimal demonstration of cyst location, morphology, and contents. Generally, bronchogenic cysts have homogeneous CT attenuation and water density. However, because of their protein and calcium contents and presence of infection, the cysts may have attenuation values as high as 120 HU (11,12).

The treatment of SVC syndrome depends on cause and prognosis. Supportive measures such as elevation of the head, supplemental oxygen, diuretics and steroids may improve symptoms. Strategies for relief of malignant obstruction may involve radiation therapy, chemotherapy or

surgery. Surgical excision usually is the best treatment with a chance of a cure in benign obstructive causes such as bronchogenic cysts (5).

In conclusion, although bronchogenic cyst is usually asymptomatic, it may ultimately become symptomatic, and in time may develop life-threatening complications such as SVCS. CT has been considered to be very useful imaging technique for detection and observation of SVCS and bronchogenic cysts. MDCTA has a wide range of applications, and enables both surgeon and radiologist to produce vascular mapping that clearly show the relationship of mediastinal vessels to mass lesions.

References

1. Beeson MS. Superior Vena Cava Syndrome [emedicine.medscape Web site]. Dec 31, 2008. Available at: <http://emedicine.medscape.com/article/760301-overview>.
2. Dinis DE, Payne WS, Bernatz PE et al. Mediastinal granulomas and fibrosing mediastinitis. *Chest* 1975; 75: 320.
3. Parker, Robert; Catherine Thomas, Lesley Bennett. *Emergencies in Respiratory Medicine*. Oxford. 2007; pp. 96–7.
4. Longmore, Murray; Ian Wilkinson, Tom Turmezei, Chee Kay Cheung. *Oxford Handbook of Clinical Medicine*. Oxford. 2007; p. 162.
5. Joshi ML, Singh SK, Siddiqui MS, et al. Bronchogenic cyst presenting as superior vena caval syndrome: A case report. *IJTCVS* 2004; 20: 106–107.
6. Marshall G, Cheah C, Lenzo NP. Bronchogenic cyst with multiple complications, *Biomed Imaging Interv J* 2007; 3(4):e42
7. St-Georges R, Deslauriers J, Duranceau A, et al. Clinical spectrum of bronchogenic cysts of the mediastinum and lung in the adult. *Ann Thorac Surg* 1991; 52(1):6-13.
8. Ribet ME, Copin MC, Gosselin B. Bronchogenic cysts of the mediastinum. *J Thorac Cardiovasc Surg* 1995; 109(5):1003-10
9. DiLorenzo M, Collin P, Vaillancourt R, et al. Bronchogenic cysts. *J Pediatr Surg* 1989; 24(10): 988–991.
10. St. Georges R, Deslauriers J, Duranceau A, et al. Clinical spectrum of bronchogenic cysts of the mediastinum and lung in the adult. *Ann Thorac Surg* 1991; 52: 6–13.
11. Kuhlman JE, Fishman EK, Wang KP, et al. Mediastinal cysts: diagnosis by CT and needle aspiration. *AJR* 1988; 150: 75–78.
12. Mendelson DS, Rose JS, Efremidis SC, et al. Bronchogenic cysts with high CT numbers. *AJR* 1983; 140: 463–465.

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