Partial anomalous pulmonary venous connection to the superior vena cava

Clinical History:

A 64-year-old male with small-cell lung carcinoma was admitted to our hospital with complaints of dyspnoea, facial oedema and swelling of neck and arms.

Imaging Findings:

We present the case of a 64-year-old male already diagnosed with small-cell lung carcinoma who was admitted to our hospital due to dyspnoea, facial oedema and swelling of neck and arms. A contrast CT was performed to evaluate the symptoms and the progression of the underlying disease. CT revealed enlarged, necrotic, anterior mediastinal lymph nodes which caused extrinsic compression to the Superior Vena Cava (SVC) causing SVC syndrome (Fig. 1). The patient was then transferred to the angiosuite for palliative endovascular treatment with stent implantation. SVC venogram showed a rather unusual finding, the direct opacification of the right superior pulmonary vein (Fig. 2). The patient was diagnosed with partial anomalous pulmonary venous connection (PAPVC) to the superior vena cava. PAPVC creates a left-to-right shunt. SVC syndrome creates high systemic vein pressure in the upper body and in the upper lobe anomalous pulmonary vein. This causes reflux of the contrast from the SVC to the anomalous pulmonary vein. After stent implantation the SVC pressure dropped and the anomalous vein was no longer opacified (Fig. 3). The patient reported no symptoms apart from a slight limitation of physical exercise prior to the accession of the lung carcinoma.

Discussion:

Partial anomalous pulmonary venous connection (PAPVC) is present when one or more, but not all, of the pulmonary veins connect to a systemic vein, the right atrium, or the coronary sinus. The prevalence reported by ElBardissi is 0.4% to 0.7% of postmortem examinations. The veins of the right lung have two to ten times the number of anomalous connections as those from the left lung. When an anomalous vein exists, it usually connects to the nearest adjacent systemic vein. The left lung veins connect to the left innominate vein, the coronary sinus, the hemiazygous vein, or to an anomalous vertical vein that drains into the innominate vein. In the right lung, the right superior pulmonary vein may connect to the right superior vena cava or azygous vein. The right inferior vein may connect with either the inferior or superior vena cava, or hepatic vein, or the azygous vein. The right pulmonary veins may also connect directly to the right atrium. Less common variations include absence of the coronary sinus with pulmonary veins from either side connecting to multiple systemic sites or to the left atrium. Rarely, an anomalous pulmonary vein will connect to the portal vein. Many variations exist in the number, size, and connections of the four pulmonary veins. If an atrial septal defect is present, about 10% of patients will have a pulmonary venous abnormality. The chest radiograph is usually normal because the pulmonary-to-systemic flow ratio is generally less than two to one. Pulmonary venous anomalies may be identified with cross sectional imaging.
and obviate the need for further studies. CT will demonstrate the course and termination of the anomalous vein, the bronchial distribution, abnormal lobation, and mediastinal shift and confirm the distinction from other venous structures, such as a left superior vena cava, pulmonary varices, and pulmonary arteriovenous fistulas. Since the majority of patients with PAPVC are asymptomatic, further evaluation after positive identification with CT is usually unwarranted. Complications of PAPVC result from pulmonary infection, atrial septal defect or dilation of the right atrial septal defect, or dilation of the right atrium and ventricle. Other imaging modalities may be required in these circumstances. Pulmonary angiography remains the standard examination for locating all the pulmonary veins. Selective injection into the right and left pulmonary arteries will visualize all pulmonary veins during a prolonged filming sequence. Alternative methods of locating pulmonary veins at catheterization include exploring the right atrium and adjacent systemic veins and documenting the anomaly by the entrance of the catheter into the vein or by a small hand injection of contrast material. While most patients with PAPVC are asymptomatic, the natural history dictates that if a significant left-to-right shunt exists, patients may develop irreversible pulmonary hypertension, pulmonary vascular obstructive disease, or right ventricular failure. Definitive treatment for PAPVC is surgical repair either using internal patch or the Warden technique. Radiologic intervention has been used in limited occasions for treating stenosis in the anastomotic sites after surgical treatment.

**Differential Diagnosis List:** Partial anomalous pulmonary venous connection to the superior vena cava

**Final Diagnosis:** Partial anomalous pulmonary venous connection to the superior vena cava

**References:**


Pennes DR, Ellis JH (1986) Anomalous pulmonary venous drainage of the left upper lobe shown by CT scans. Radiology 159:23-4

Description: Direct opacification of the right upper lobe pulmonary vein. Origin:
Figure 2

Description: Large, necrotic, anterior mediastinal lymph nodes which cause extrinsic compression to the SVC.

Origin:
**Description:** Large, necrotic, anterior mediastinal lymph nodes which cause extrinsic compression to the SVC. **Origin:**

**Description:** Large, necrotic, anterior mediastinal lymph nodes which cause extrinsic compression to the SVC. **Origin:**
Figure 3

**Description:** After stent implantation the pressure in the SVC is relieved and the anomalous pulmonary vein is no longer opacified. **Origin:**