Intravascular papillary endothelial hyperplasia of superior vena cava: a rare cause of the superior vena cava syndrome

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Abstract
The superior vena cava syndrome associated with an intraluminal vascular proliferative lesion is extremely rare. A case of intravascular papillary endothelial hyperplasia of the superior vena cava causing obstructive symptoms is reported that was successfully managed by surgical excision.

About two thirds of the cases of superior vena caval obstruction are caused by primary lung cancer, metastatic carcinoma, and lymphoma. Less common benign causes are mediastinal fibrosis associated with granulomatous disease or radiation, retrosternal thyroid, aortic aneurysm, and inflammatory lymphadenitis. It may also result from thrombosis after intravenous procedures such as Swan-Ganz catheterisation. Primary intravascular proliferative lesions should be included in the differential diagnosis of the superior vena cava syndrome, as illustrated by this case.

Case report
A 57 year old obese white woman presented with episodes of epistaxis and headache. Her family history and her own medical history were unremarkable except that she frequently experienced a "full sensation" in her head when she lay flat or attempted to bend over. There was no history of trauma or previous admission to hospital and she denied taking any medication, including hormonal replacement.

On admission she had a temperature of 36.8°C and a blood pressure of 160/110 mm Hg; the general physical examination showed nothing remarkable. The initial laboratory tests, including a full blood count, urine analysis, blood chemistry and coagulation measurements gave results within normal limits. A chest radiograph showed a right hilar mass suggestive of a lung cancer. Computed tomography, however, showed a soft tissue density within the lumen of the superior vena cava (confirmed by venography) and clear lung fields (fig 1). The mass did not obstruct the passage of the venogram catheter to any extent, but the dye appeared to be streaming around the mass.

The patient underwent a thoracotomy and exploration of the superior vena cava and a polypoidal intravascular mass was found attached to the medial wall of the superior vena cava, distal to the junction with the right atrium. The surrounding structures, including the lungs, appeared normal, and there were no mediastinal adhesions. An intra-operative biopsy disclosed a vascular lesion without histological evidence of malignancy. The lesion, along with the medial wall of the superior vena cava, was excised and the superior vena cava was reconstructed with a pericardial patch. The patient made a good recovery and two years after surgery remains well.

Figure 1  Computed tomogram of the chest showing a dilated superior vena cava due to an intraluminal mass of soft tissue density (indicated by arrow). The aorta and pulmonary arteries appear normal.

Figure 2  Photomicrograph of the intraluminal lesion in the superior vena cava: large thin walled vascular channels with papillary projections in the lower field. (Haematoxylin and eosin.)
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The resected specimen consisted of an intraluminal broad based purplish red fungating spongy mass, which measured 3·5 × 2·0 cm. The cut surface showed irregular spaces containing red-brown bloody material, alternating with patchy yellow, firm, and solid areas. Microscopically, the lesion was composed of vascular channels of varying size and shape, which were separated by thin fibrous walls (fig 2). No thrombus was found in multiple sections. Many of the large vascular spaces contained papillary ingrowths with small fibrous cores, lined by endothelial cells, simulating a haemangioendothelioma or angiosarcoma. No significant cellular pleomorphism of the endothelial cells or mitotic activity was found. Although the intimal surface of the superior vena cava was focally disturbed, with loss of internal elastica at the point of fibrous adhesion with the vascular lesion, the muscular wall and the adventitia were intact. No evidence of vasculitis or tissue necrosis was found. The histological features were consistent with “intravascular papillary endothelial hyperplasia.”

Discussion
Superior vena caval obstruction has been reported in association with lesions within the superior vena cava, such as congenital aneurysm, vasculitis, and thrombosis. Although idiopathic thrombosis of the superior vena cava may occur, thrombosis occurs more commonly as a complication of intravenous instrumentation. Primary tumours of the vena cava are extremely rare. Hallock et al. reported a case of leiomyosarcoma of the inferior vena cava and noted four previously reported primary tumours of the inferior vena cava. Reisinger in 1942 described a case of “endothelioma” affecting the inferior vena cava and heart with tumour extension to the adventitia of the aorta and pulmonary artery. Intravascular papillary endothelial hyperplasia in the superior vena cava has not, to our knowledge, been described.

Intravascular papillary endothelial hyperplasia most commonly occurs in fingers, head, neck, and trunk, though it may occur in virtually any location, such as synovium. The prognosis is excellent and cure is by simple excision. It is considered to be a peculiar organising process of a thrombus with an exuberant intravascular endothelial proliferation, sometimes mimicking haemangioendothelioma or angiosarcoma. Although no thrombus was found in our case, the possibility of a completely organised idiopathic thrombosis of the superior vena cava should be considered.

Correct diagnosis of superior vena caval obstruction is important so that appropriate therapeutic intervention can be carried out. In our patient superior vena caval obstruction was due to an unusual primary vascular lesion, intravascular papillary endothelial hyperplasia of the superior vena cava. The lesion developed in the absence of a history of trauma, intravenous instrumentation, or evidence of antecedent haemodynamic disturbances.