Intimal sarcoma of the superior vena cava

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Summary: A case of superior vena cava syndrome caused by a primary intimal sarcoma of the superior vena cava is described. The known causes of superior vena caval obstruction are discussed, together with the difficulties in identifying the underlying lesion. The possibility of a primary superior vena caval neoplasm as a cause of superior vena caval obstruction should be considered in patients presenting with superior vena caval syndrome.

Introduction

Superior vena caval obstruction (SVCO) is characterized by symptoms and signs which result from a raised superior vena caval pressure. Common presenting features are headache, oedema of the face and arms and dilated veins on the upper chest. Severe cases may develop respiratory distress or cerebral haemorrhage and the prognosis is poor. The vast majority of cases of SVCO are caused by malignant disease, the commonest being bronchogenic carcinoma. However, on reviewing the established classifications of causes of SVCO, primary tumours of the superior vena cava are not included. We describe a case of intimal sarcoma arising in the superior vena cava which caused SVCO and resulted in fatal cerebellar haemorrhage.

Case report

A 63 year old female patient was admitted with a 2 week history of facial and upper arm swelling, prominent veins on her chest, headaches and hoarseness.

On examination, there was also a diffuse swelling in the lower anterior neck resembling a thyroid goitre and a lymph node was palpable in the right axilla. Indirect laryngoscopy showed oedema of the vocal cords. Her chest radiographs and the ultrasound scans of her thyroid gland and abdomen were all normal. A diagnosis of SVCO was made and because of its acute onset, a venogram was performed. This showed brachiocephalic vein obstruction by thrombus and the patient was anticoagulated. Computerized tomography (CT) of her thorax and mammography were arranged. Unfortunately, before these could be done, and 8

days after admission, the patient had a respiratory arrest and died despite ventilatory support.

At postmortem examination, the brain was found to be diffusely oedematous with flattening of the gyri and the cerebellum contained a haematoma measuring 4 cm in diameter. The lungs were oedematous and a pericardial effusion was noted. On opening the superior vena cava, it was almost completely occluded by a soft, partly nodular neoplasm 4.5 cm in diameter. Thrombus was overlying the tumour and was extending to the junction of the brachiocephalic and innominate veins. On slicing into the tumour it could be seen to be invading the vein wall. Despite an extensive search, no other primary site could be identified and there was no evidence of metastases to the local lymph nodes, lungs or liver.

The tumour consisted of epithelial-like cells arranged in groups with intervening thin fibrous trabeculae. The cells had open nuclei with prominent nucleoli. Mitoses were easily found but nuclear pleomorphism was only moderate. The cytoplasm was fairly distinct and eosinophilic. Tumour was invading the smooth muscle of the vein but adjacent lymph nodes were not involved. Centrally, the tumour was partly necrotic and was covered by thrombus. The cells expressed Cam 5.2, an antibody recognizing low molecular weight cytokeratin and usually found in epithelial tumours; Factor VIII was also weakly positive indicating that the cells were intimal in origin. The morphology and immunophenotype correlates with that described in intimal sarcoma (Figure 1).

Discussion

Intimal sarcoma describes a malignant neoplasm that arises from the intima of a blood vessel and has a characteristic immunophenotypic staining pattern.^{1–3} Arterial intimal sarcomas have been des-

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Figure 1 Pleomorphic tumour cells with intervening thin fibrous trabeculae (haematoxylin and eosin, $\times 300$).

cribed and are usually characterized by wide dissemination.⁴ Primary sarcomas originating from venous walls are rare. The majority are leiomyosarcomas and the most common vessel involved is the inferior vena cava.⁵⁻⁷ Identification and differentiation from other primary vascular neoplasms followed immunophenotypic staining. Analysis using a standard ABC immunoperoxidase technique showed the cells to be desmin, neurone-

specific enolase and leucocyte common antigen negative so excluding a leiomyosarcoma, neuroendocrine carcinoma or large cell lymphoma. Electron microscopy was not performed due to the degree of postmortem autolysis.

SVCO is usually diagnosed from the clinical presentation. The aetiology of SVCO has been described in several studies since it was first recorded in 1757 by William Hunter.⁸⁻¹⁰ Such studies have demonstrated a decline in the number of cases due to aortic aneurysm and an increase in the number of cases due to malignancies. None of the studies listing aetiologies that were examined included primary superior vena caval neoplasm as a cause of obstruction.

In retrospect, radiotherapy might have been a suitable early treatment¹¹ but the response of intimal sarcomas to radiotherapy has not been described in the literature reviewed. In conclusion, this case highlights the need to consider primary superior vena caval neoplasms as a possible cause of superior vena caval obstruction.

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