Leiomyosarcoma of the Inferior Vena Cava: An Unusual Cause of Pulmonary Embolism*

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A patient with leiomyosarcoma of the inferior vena cava is presented. The diagnosis was suggested by cavography and confirmed at necropsy. To the best of the authors' knowledge, this is the first reported case with the clinical picture of pulmonary embolism.

Leiomyosarcoma is a rare malignant tumor which often arises from soft tissue, but it can also arise from viscera, particularly from the media of arteries and veins.1

In 1871, Perl2 published the first description of a sarcoma of the inferior vena cava. So far, only 20 cases have been reported in the literature.3,4 This report is concerned with the description of a further case present-

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Figure 1. Inferior ilio-cavography (anteroposterior incidence). Note hypoplasia of inferior vena cava in its proximal portion, the distal dilatation with enormous endoluminal formation reaching the atrium and the collaterality through renal and perivertebral veins.

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performed; it disclosed an obstruction of the inferior vena cava with collateral enlarged vessels (Fig 1). A mass surrounded by vegetations was found in the right atrium. This mass seemed to grow towards the pulmonary arterial tree where obliterations of peripheral vessels were noticed.

In the following days, the patient developed circulatory shock with repeated bouts of ventricular tachycardia. She was admitted to the emergency surgical theater and a Trendelenburg operation was undertaken. The heart fibrillated during opening the chest and she died in spite of institution of electrical countershock.

Necropsy Findings

A greyish white tumor arising from the inferior vena cava was found 10 cm below the renal veins (Fig 2). The tumor infiltrated the vein wall on 2 cm, narrowing the lumen; it then ran freely along the whole length of the inferior vena cava and was attached only loosely to the endothelium. The tumor had a smooth surface.

In the right atrium, it showed a cauliflower shape with vegetations projecting into the right ventricle and infundibulum of the pulmonary artery.

Small tumor fragments were found in the peripheral branches of the pulmonary artery, but no metastases were detected. The necropsy failed to reveal other features of interest.

At microscopic examination, the tumor was diagnosed as a leiomyosarcoma. It was formed of spindle cells with elongated nuclei showing pleomorphism and mitotic activity. Mitotic abnormalities were numerous and some plurinuclear giant cells were noticed. Occasional longitudinal striation was seen (Fig 3).

![Figure 3. Microscopic aspect: spindle cells with elongated nuclei showing pleomorphism and mitotic activity. Numerous mitotic abnormalities with some plurinuclear giant cells (H and E × 250).](image)

COMMENTS

Leiomyosarcoma of the inferior vena cava is an extremely rare tumor: only 20 cases have been described so far.

The diagnosis is usually made at autopsy or during surgical laparotomy. The diagnosis was reached by venacavography in only two instances.

Symptoms of pulmonary embolism have never been described in similar cases. Intermittent closure of the pulmonary infundibulum by a tumor fringe or embolization of tumor vegetations separated from the tumor mass could account for such a complication.

Leiomyosarcoma of the inferior vena cava, like leiomyosarcoma arising from other tissues, occurs mainly in women. It has been described in patients from 23 to 83 years of age with the higher incidence in the middle-aged group.

In spite of the malignant histologic aspect of this tumor, leiomyosarcoma of the inferior vena cava rarely metastasizes. When present, metastases are located in the liver or in the abdominal lymph nodes. Only one case with metastasis in a costal rib was described. Due to the rarity of metastases, resection can still be considered even in the presence of a large tumor.
The correct diagnosis with a good definition of the tumor limits is thus mandatory. Careful study of the clinical picture may be of considerable help in this respect. The symptoms indeed depend upon the level and extension of the mass. The different clinical symptoms are generally related to progressive occlusion of the inferior vena cava followed by development of collateral circulation.

In previous reports, when the tumor was located in the upper third of the inferior vena cava, Chiari's syndrome was observed with hepatomegaly, ascites and jaundice. The origin of the hepatic vein was invaded by the tumor or occluded by antemortem thrombosis. Hepatic failure was sometimes more dramatic when the occlusion of the hepatic vein was abrupt. Tumors located in the middle third of the inferior vena cava give rise to a nephrotic syndrome and renal failure.

Leiomyosarcoma located in the lower third of the inferior vena cava are extremely difficult to diagnose. Very often, they extend to the peritoneum with no obstruction in the vena cava, but produce severe lumbar pain. It is only when the tumor grows inside the vessel that swelling of the legs may appear with a palpable abdominal mass.

Cavography seems to be the most suitable method to reach the diagnosis and to obtain information as regards the tumor extent and location. This technique was used in two previous instances by Roussak and Heppelston and Deutsch and colleagues. In our patient inferior cavography revealed a defect of filling with many anastomotic vessels. The superior cavography showed extension of the tumor into the right atrium. Filling defects were also observed in peripheral branches of the pulmonary artery. Propagation into the right atrium was found in four cases so far. In all of them, the tumor arose in the upper third of the inferior vena cava.

Resection of the tumor was reported in six cases. The first operation was carried out by Melchior in 1921. Three patients survived the surgical procedure and were still alive between nine months and one year after surgery.

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Rhythmic Shoulder Girdle Muscle Contractions as a Complication in Pacemaker Treatment

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A 69-year-old man had been treated for 5½ years with permanent transvenous pacemaker for Adams-Stokes syndrome. On the day following vigorous arm exercise he had muscle contractions in the right shoulder synchronous with the heart activity. Local damage to the electrode cable with current leakage to the upper brachial plexus was verified by neurophysiologic examination and at operation. A simple method of repairing the insulation defect is reported.

A pacemaker-treated patient presented some peculiar neurologic symptoms—rhythmic muscle contractions arising in the right shoulder girdle on abduction of the arm. Neurologic examination led to suspicion of damage to the electrode cable close to the upper brachial plexus. After operation, the patient made an uneventful recovery and became free of the muscle jerks. No similar case is known to have been reported previously.

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