Endoscopic tumor resection of the inferior vena cava

H. Jeanmart, MD,*, P. Lecompte, MD,† F. Casselman, MD, PhD, FETCS,*
J. Coddens, MD,‡ G. Van Vaerenberg,* and H. Vanermen, MD,* Aalst, Belgium

Preoperative History

A 51-year-old woman was referred for evaluation of an asymptomatic systolic murmur. Transthoracic echocardiography showed a large right atrial (RA) mass with intermittent prolapse through the tricuspid valve and extending into the pulmonary artery. On the preoperative transesophageal echocardiographic examination, a tumor with multiple strands was seen. One strand intermittently obstructed the outflow of the hepatic vein (Figure 1). There was apparent obstruction of the RA inflow and right ventricular inflow and outflow tracts by another strand.

Operative and Postoperative Period

The resection was performed with a minimally invasive endoscopic approach through a small thoracotomy on the right side. After induction of deep hypothermia and circulatory arrest, the RA was opened, and the mass was immediately seen coming out of the inferior vena cava (IVC; Figure 2, A). The mass could easily be removed from the ventricle by means of simple traction. One large zone of insertion was seen inside the IVC and extending until the RA. After resection, a second mass was seen, originating even deeper in the IVC, below the origin of the subhepatic veins (Figure 2, B). The complete specimen was more than 15 cm long and seemed intact on macroscopic examination.

Small tears on the tricuspid valve were primarily repaired, but after coming off pump, a major tricuspid insufficiency was seen, and the valve had to be replaced. The final pathology of the tumor was a leiomyoma. The postoperative period was without complications, and the patient left the hospital after 8 days.

Discussion

Intravenous leiomyoma is a rare benign vascular tumor, most commonly found in the pelvic veins, of unclear cause that develops exclusively in women, many of whom, as in the case of our patient, have undergone a hysterectomy in the past for uterine leiomyoma. Some authors believe that the tumor results from the vascular invasion from a uterine leiomyoma, but it has also been proposed that the tumor is vascular in origin and is derived from the medial smooth muscle cells of a vessel wall.

The peripheral fibers of the tumor are compressed into a pseudocapsule with a white glistening appearance and a characteristic swirling pattern on the surface. A characteristic of benign leiomyomas is that they grow freely within the vessel lumen but do not invade the wall. Histologic analysis of resected specimens reveals benign smooth muscle architecture without hemorrhage or necrosis.

When right-heart involvement is present, the patient presents with typical right-heart failure symptoms. Management of an intravenous leiomyoma requires surgical excision of the tumor to relieve symptoms and prevent embolism and sudden death. Biopsy of the tumor is generally not indicated because resection of the tumor is indicated, regardless of histologic findings. Few data are available regarding the natural history or the risk of malignant transformation. Although benign in nature, local recurrence has been reported after surgical resection and lung metastasis also.
Mandelbaum and colleagues described, in 1974, the first successful resection of a leiomyoma in the right heart, which originated in the IVC just below the RA. Various surgical approaches have been used for the removal of the tumor. If the tumor is too extensive or adheres to the cardiac and vascular structures, requiring resection of the abdominopelvic and intrathoracic components, then a separate operation might be mandatory. Otherwise, 1-stage resection during circulatory arrest and hypothermia can be used with success. The case reported here is the first case of an IVC tumor removal with an endoscopic approach.

References