rapid resolution of right-side heart failure and subsequent amelioration of LV function.

Extreme cases of Ebstein’s anomaly like the one described in this case report are rare in developed countries, whereas they may more often present to a surgical team when working in a humanitarian mission in Third World countries. The choice to perform very high risk surgery within a setting of limited resources (and material) against a decision to leave such cases untreated is challenging, not to mention the ethical set of problems behind it. To operate on this case was clearly a gamble, but to wager and battle for the sake of a sick patient with no other options is a worthwhile and sometimes rewarding task.

We thank the Associazione Un Cuore Un Mondo (www.uncuoreunmondo.org) for the technical and financial support to the mission.

References

Leiomyosarcoma of the Superior Vena Cava
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Leiomyosarcoma of the superior vena cava is a very rare tumor and only a few cases have been reported, with various techniques of vascular reconstruction. We describe a new case of leiomyosarcoma of the superior vena cava in a 61-year-old woman with extension to the brachiocephalic arterial trunk. Resection and vascular reconstruction were performed using, respectively, polytetrafluoroethylene and polyethylene terephthalate vascular grafts.


Vascular leiomyosarcoma is a very rare tumor and the inferior vena cava is the most common site (50%) [1]. The superior vena cava (SVC) is exceptionally involved and only a dozen cases have been previously reported [2]. The tumor is commonly revealed by physical signs of a SVC obstruction syndrome, less frequently by metastasis or unexpected computed tomography scan finding. Leiomyosarcoma is considered a very locally aggressive tumor with rare distant dissemination and an
unfavorable prognosis. An aggressive therapeutic approach, including chemoradiotherapy and large surgical resection, is usually considered the treatment of choice [1]. Replacement of the SVC has been described using various techniques of vein reconstruction, on an arrested heart or not [3-6].

In January 2010, a 61-year-old woman was admitted to the hospital with facial swelling, collateral venous circulation, and tinnitus. A chest roentgenogram showed a right paravertebral mass with superior mediastinal widening, and computed tomography revealed a mediastinal tumor, 4 cm in diameter, involving the SVC and brachiocephalic artery. A positron emission tomography scan confirmed the malignant nature of the tumor, without locoregional or distant significant abnormalities. A cavography showed a very tight stenosis with thrombus, and a stent was placed to treat the SVC syndrome. Biopsy was performed through a mediastinoscopy by cervicotomy and revealed a spindle cell neoplasm. The immunohistochemical study showed the tumor was clearly positive for actin smooth muscle antigen and negative for S100 protein, CD 68, CD 34, cytokeratin AE1, AE3, H-caldesmon, and CD 117. This was in favor of an epithelioid leiomyosarcoma.

In February 2010, the patient was operated on. Through a median sternotomy and after opening the pericardium, inspection showed that the tumor infiltrated the SVC 2 cm before the right atrium and also the lateral face of the brachiocephalic arterial trunk 1 cm after its origin. A cardiopulmonary bypass was placed with a cannula between the aorta and the brachiocephalic artery just before its bifurcation, on the one hand, and a cannula between the right atrium and the internal jugular vein on the other hand (Fig 1). It was used in normothermia as a circulatory support. The SVC was resected with safety margins, and the brachiocephalic artery was dissected from its origin to its bifurcation. The right phrenic nerve was infiltrated. The tumor was completely removed, requiring the section of theazygos vein and the right phrenic nerve. The SVC was reconstructed using a polytetrafluoroethylene (PTFE) ringed vascular graft, and the brachiocephalic arterial trunk was replaced with a polyethylene terephtalate vascular graft anastomosed on the aorta upstream (Fig 2).

The patient’s postoperative course was complicated by a dysphonia due to recurrent laryngeal nerve paralysis, which was successfully treated by autologous fat injection. Pathology analysis confirmed the diagnosis of high-grade leiomyosarcoma of the SVC with margins of both the SVC and brachiocephalic artery free of tumor. From April to June 2010, four courses of chemotherapy with Adriamycin and Deticine were administered. In March 2012, the patient was restaged and no signs of mediastinal recurrences; neither were distant metastases found. Vascular grafts showed perfect patency.

Comment

Primary vascular leiomyosarcomas are very rare malignant tumors, approximately 2% of all leiomyosarcomas [3]. The tumor originates from proliferation of smooth muscle cells of the media and may grow intravascularly or extravascularly or both [7]. Distant dissemination is rarely described. It involves large veins almost five times more than arteries, and the most common site is the inferior vena cava and its branches [3]. It occurs in women in a large majority. The SVC localization is extremely rare, and only 15 cases have been previously reported [1, 6]. It usually results in SVC syndrome of gradual onset. Computed tomography and magnetic resonance imaging may suggest the original tumor, but the diagnosis is histologic and requires mediastinoscopy, thoracotomy, or percutaneous endovascular biopsy.

Therapeutic management should be aggressive and include preoperative radiotherapy to reduce the tumor volume and adjuvant chemoradiotherapy, but is mostly based on a large surgical resection when possible [3]. Several techniques for vascular reconstruction have been described. Autologous vein may be the best graft in a low-pressure venous system. The spiral vein graft offers a sufficient graft caliber but it extends the operating time [4, 5]. The use of custom-made autologous pericardial graft or bovine pericardial graft has also been proposed [6] but not reproduced. A PTFE prosthesis is most
commonly used because of its rapid endothelialization, and it produces less thrombogenicity than polyethylene terephthalate. Furthermore, PTFE has the advantage of being reinforced by rings, reducing the risk of collapse when the venous pressure drops [2]. However, in addition to the increased risk of infection, the long-term patency of synthetic implants after reconstruction of large vessels remains unclear. There is actually no evidence to confirm the superiority of one technique over the others but according to Dartevelle and associates [8], reporting a 5-year patency rate of 86%, we believe that vascular reconstruction of the SVC using PTFE prosthesis is a valuable option. In our case, 2 years of follow-up showed graft patency and no infection.

We have reported the first case of primary leiomyosarcoma involving both superior vena cava and brachiocephalic arterial trunk. Whereas large surgical resection is commonly recommended, several techniques for vascular reconstruction have been described. Our preference was for PTFE ringed vascular graft, and we obtained the outcomes of a simple postoperative course and no long-term complications.

References