Leiomyosarcoma of the superior vena cava: Resection with vascular reconstruction – case report and review of the literature

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The present report describes a case of pedunculated intraluminal leiomyosarcoma of the superior vena cava, extending to the right

atrium, that was successfully resected surgically. Superior vena cava reconstruction was performed using bovine pericardial graft, saving the sinus node. The pathological variants of this neoplasm according to the anatomical site of the tumour are described.

Key Words: Bovine pericardial graft; Leiomyosarcoma; Sinus node; Superior vena cava; Tumour resection; Vascular reconstruction

Leiomyosarcoma of the superior vena cava (SVC) is a rare mesenchymal tumour that originates from the smooth muscle cells of the vascular wall. The survival rate in unresected cases is generally measured in months. The clinical signs of SVC leiomyosarcoma and the possibility of its surgical resectability are dependent on its location. However, pathoanatomical variations of the tumour's localization have not been described until now. The present report describes a case of unusual intraluminal leiomyosarcoma of the SVC extending to the right atrium that was managed with complete surgical resection and SVC reconstruction, saving the sinus node. In addition, the anatomical variants of this malignancy are discussed.

CASE PRESENTATION

A 79-year-old woman was referred to the E Wolfson Medical Center (Holon, Israel) with effort-induced dyspnea. Physical examination was unremarkable. Two-dimensional transthoracic echocardiography showed a 3 cm × 5 cm nonseptal origin, mobile, irregular mass within the right atrium with mild to moderate restriction of SVC inflow. A computed tomography scan of the chest revealed a filling defect in the SVC (Figure 1). Other observations were within normal limits and the patient was referred for surgery.

Operative procedure

After median sternotomy, palpation of the SVC above the tumour was performed. The azygos vein was tumour-free. The patient was placed on cardiopulmonary bypass (CPB). Myocardial protection was achieved with antegrade and retrograde cold blood intermittent cardioplegia and moderate hypothermia (32°C). The right atrium was opened and the protruding tumour was seen. Because the tumour was adherent to the SVC wall, the SVC was opened as well. This was opened as medially as possible to avoid the sinus node. The

Figure 1) Computed tomography scan showing the superior vena cava partially occluded by a tumoral mass (arrow)

tumour was peeled from the SVC wall up to its origin in the posterior wall of the SVC, 2 cm above the junction of the SVC and the right atrium, proximal to the azygos vein. The SVC was resected to achieve free margins and was reconstructed with a Shelhigh bovine pericardial patch (Shelhigh Inc, USA). A lymph node removed from the area of the SVC was negative for malignancy.

At gross examination, the mass was whitish, ovoid and pedunculate. It measured 4.5 cm × 4 cm × 5 cm and weighed 62 g. The surface was irregular and multinodular. The tumour was entirely intraluminal. Histological examination revealed a typical pattern of high-grade primary SVC leiomyosarcoma. Tumour cells displayed vimentin, desmin and alpha-smooth muscle actin, and were positive for K1-67 on immunostatin.

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The postoperative course was uneventful and the patient regained normal sinus rhythm immediately after surgery. Postoperative chemotherapy was instituted. Follow-up over one year showed no recurrence of the tumour and good patency of the SVC (Figure 2).

DISCUSSION

Vascular origin leiomyosarcoma is a slow-growing neoplasm, usually presenting late; fast diagnosis with early treatment may improve patient survival (1). Leiomyosarcoma of the SVC was first described by Ehrenberg in 1911 (2), and the first repair was performed in 1971 by Stefanini et al (3). So far, 12 cases (including the present case) have been reported.

In most described cases of SVC leiomyosarcoma, a diagnosis was made preoperatively by using computed tomography and/or venography (4-7). In three cases (7-9), nuclear magnetic resonance imaging was used, and in two cases (8,9), percutaneous endovascular biopsy was used.

In the majority of reported cases, patients were admitted to the hospital with symptoms of acute SVC syndrome caused by massive thrombosis or infiltration of a tumoral mass (2,5-7,9,10). To prepare patients for surgery, the use of local fibrinolytic therapy (6), SVC metallic stent placement (9) or preoperative chemotherapy (7) has been required.

Special attention should be paid to the level of origin of the SVC leiomyosarcoma. In our opinion, it is an important question that influences a choice of surgical tactics and the subsequent treatment. Some of these described tumours arose from the confluence of the internal jugular and the subclavian veins (4,11), some originated in the area of the innominate veins (7,8,12), and others from the area of the azygos vein (5,13). Taking this into consideration, we believe that SVC leiomyosarcoma can be classified into three segments depending on the anatomical site of origin:

- Segment 1 (lower) – extending from the right atrium to the azygos vein;
- Segment 2 (middle) – extending from the azygos vein to the confluence of the right and left brachiocephalic veins; and
- Segment 3 (upper) – extending from the confluence of the brachiocephalic veins to the subclavian vein and right internal jugular vein.

In some cases, the tumour extended into two or three segments.

The best treatment of leiomyosarcoma of the great veins is complete surgical resection with free margins (1). It is interesting to note that for removal of a tumour in one case, authors used a Gott aneurysm shunt (Sherwood Medical, USA) (11), and in another, the authors used a CPB with circulatory arrest (7). Aggressive combined chemotherapy, radiotherapy and surgical treatment may permit a relatively long survival (7). One patient was still alive 7.5 years after the first operation (14). Mortality 30 days postoperatively was, in two cases, due to intracranial hemorrhaging (9) and sepsis (4). In two cases, patients died before the beginning of treatment (2,10). Recurrence was discovered in two patients (11,14).

After resection of a tumour, the next problems facing the surgeon are the options in vena cava management. One caval defect was closed primarily (1), another was closed with a saphenous vein patch (11), and a third, after a circumferential

Figure 2) Postoperative computed tomography venography of the superior vena cava showing good patency of the reconstructed vein

caval resection, with a saphenous spiral vein graft (8). Prosthetic grafts were mainly used for SVC reconstruction – polytetrafluoroethylene tube grafts (9,13), Dacron tubular Dacron graft (Invista Inc, Canada) (3) and Dacron regular prostheses (Invista Inc, Canada) (5). In the case described by Spaggiari et al (7), the right confluence between the subclavian and internal jugular vein was connected to the right atrium with a polytetrafluoroethylene graft, and the left innominate vein was anastomosed to this graft using a second graft. In the present patient, we used a bovine pericardial graft for reconstruction of the SVC.

The patient described herein had the unusual findings of an SVC leiomyosarcoma arising from the supra-atrial segment of the SVC below the azygos vein, with tumour extension to the right atrium. We believe that the location of the malignancy, as in the present case, demands the use of a specific surgical technique. First, using CPB with double caval cannulation allowed us to localize and precisely remove the tumour with free margins, and perform reconstruction of the SVC without technical problems. We also placed the SVC cannula in a very high position to prevent fragmentation and pulmonary embolization. For the same reason, manipulation of the heart was avoided before establishing cross-clamping and cardioplegic arrest. Second, during tumour excision and SVC reconstruction, we saved the area of the sinus node, which undoubtedly has a positive value in postoperative recovery.

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