SHORT REPORT

Leiomyosarcoma of the Popliteal Vein

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Primary vascular leiomyosarcomas are rare tumors arising from smooth muscle cells of the vessel walls and they involve the extremities in about one third of reported cases. Leiomyosarcomas of the popliteal vein are even more seldom. We report a case of a 68-year-old woman who has been treated for a popliteal vein leiomyosarcoma with en bloc resection and bovine mesenteric vein interposition grafts to restore continuity.

Keywords: Leiomyosarcoma; Popliteal vein; Diagnostic methods; Surgery.

Introduction

The first vascular leiomyosarcoma, which originated from the inferior vena cava, was described by Perl in 1871. Reports of leiomyosarcomas arising from popliteal vein are very rare and only five cases have been reported.1 We report, a case of leiomyosarcoma of the right popliteal vein in a 68-year-old woman.

Case Report

A 68-year-old woman presented with pain in the right leg. Physical examination showed right leg oedema, tenderness, motor dysfunction of the knee and a palpable mass in the popliteal fossa.

Duplex ultrasound (Fig. 1) showed a popliteal fossa mass of mixed echogenicity and a reduction of the blood flow in the popliteal vein at the distal edge of the mass. MRI (Fig. 2) showed a mass with normal borders and pathological vessels inside the mass.

Because of these findings, we decided an operative removal of the tumor. We exposed the tumor through a posterior S-type skin incision. The tibial nerve was carefully preserved (Fig. 3). The popliteal vein was encased by the tumor. The popliteal artery was only compressed by the tumor without being encased. The tumor was completely resected, with a 15 cm length of popliteal vein, from the femoral till the bifurcation of the popliteal vein into the tibial veins. Continuity was restored by interposition of two 6 mm bovine mesenteric vein grafts to each tibial vein. (Fig. 4). The patient was discharged after 3 days. She was anticoagulated with low molecular weight heparin and then vitamin k antagonist.

The surgical resection specimen consisted of lobulated tissue with clear margins, measuring $7 \times 4 \times 3$ cm, and containing the popliteal vein. Sections showed a fleshy white tumor surrounded by a capsule. Microscopic examination (Fig. 5) revealed a low-grade spindle cell sarcoma arising from the wall of the popliteal vein with nuclear pleomorphism, atypia and medium mitotic rate. Immunohistochemical stains showed positive immunoreactivity for smooth muscle actin (SMA) and focally for vimentin and CD34, but negative for S-100 protein. The tumor was a leiomyosarcoma (Grade 1 according to Guillou-Coindre). The margins of the specimen were free of tumor.

A post operative venography after 1 month showed normal blood flow. A CT and MRI examination of the thorax and the abdomen showed no metastatic spread. We decided, there was no indication for chemotherapy, since she had a totally incapsulated leiomyosarcoma, and negative CT and MRI findings. At 18 months
Fig. 1. Preoperative ultrasound examination of the popliteal fossa (popliteal vein mass).

Fig. 2. Preoperative MRI of the popliteal fossa (popliteal vein mass).

Vein Leiomyosarcoma

Fig. 3. Popliteal fossa mass and tibialis nerve.

Fig. 4. The restoration of popliteal vein continuity, after excision of popliteal vein leiomyosarcoma, using bovine mesenteric vein grafts.

Fig. 5. Histological examination of popliteal vein leiomyosarcoma.
follow up the patient had no signs of metastatic disease. However, the venography (Fig. 6) showed thrombosis of grafts with satisfactory collateral vein circulation.

Discussion

Leiomyosarcomas that arise from venous wall are rare. They have a poor prognosis with 5-year survival rate of 32%. Although they grow slowly, they have a poor prognosis because liver and lung metastases are often present at the time of diagnosis. Half of the cases of venous leiomyosarcomas arise from the inferior vena cava and only one third of them involve the veins of lower extremities. Veins of the lower extremities, which are affected, are in decreasing frequency, femoral, external iliac and popliteal vein. In the literature of the last 20 years we found five other cases of leiomyosarcomas from the popliteal vein out of a total of 80 cases of venous leiomyosarcomas of the extremities.

The clinical presentation of a venous leiomyosarcoma in the lower extremities is variable. It can present as deep pain, edema, a palpable mass, or it can mimic symptoms of deep venous thrombosis. Especially in healthy persons with no surgical history, pain and edema of the extremities must raise the suspicion of malignancy although it is very rare.

MRI is considered to be the method of choice for soft tissue tumors. Venography and ultrasound, can assist with making the diagnosis. CT is helpful for surgical planning by demonstrating the anatomic relationships with the surrounding structures.

The treatment is surgical. The resection of the tumor follows the principles of surgical oncology whilst not harming the surrounding arteries and nerves. The surgical reconstruction of veins and, if it is necessary, the arteries of the lower extremities can be achieved with vein grafts or biological grafts. A more radical operation, such as above knee amputation, was not indicated in our case, because the tumor was fully encapsulated, there was no thrombus in the popliteal or femoral vein, and the tumor had not spread to surrounding structures. The diagnosis of leiomyosarcoma is made by a biopsy and immunohistochemical studies. If the diagnosis is made after surgical removal of the tumor, the patient must be examined with CT or MRI in order to exclude metastatic spread.

References


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