SHORT REPORT

Primary Leiomyosarcoma of the Great Saphenous Vein: Case Report

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Objective. To describe a case of primary leiomyosarcoma of the great saphenous vein.

Patient record. A 59-year-old Chinese lady presented with two painful lumps in the right thigh in the line of the great saphenous vein. At surgery, in September 2004, two tumors in the right great saphenous vein above the knee were excised with the intervening normal vein. Histopathological studies confirmed both masses as leiomyosarcoma. Radiotherapy was given postoperatively. The patient has been followed-up for 300 days after surgery with no evidence of local or distant metastasis.

Conclusion. Leiomyosarcoma of the great saphenous vein can exist at more than one site. Tumor resection and radiotherapy was associated with good patient prognosis.

Keywords: Leiomyosarcoma; Saphenous vein; Radiotherapy.

Introduction

Leiomyosarcoma usually occurs in the gastrointestinal tract or uterus, but rarely occurs in the wall of large veins and arteries. Leiomyosarcoma affecting venous system is rare, particularly in the greater saphenous vein. Since, van Ree described the first case in 1919, only 26 such cases have been reported in the world. Hence, we document the first case in China.

Case Report

A 59-year-old lady was hospitalized in September 2004 because of a mass on the inner aspect of the right thigh. The mass had been growing slowly for 9 months accompanied, with localized pain for the previous 2 months. The pain increased with walking or bearing weight on the affected limb. There were two solid, hard masses, located along the line of the great saphenous vein above the right knee (Fig. 1(A)) with slight tenderness but without transverse or longitudinal mobility. The proximal mass measured about 2 cm × 4 cm, and the distal 3 cm × 6 cm. Chest X-ray, ultrasound of the abdomen and routine examination of blood and urine were normal. A diagnosis of ‘thrombus’ of the saphenous vein was made by ultrasound examination of the two masses. Color Doppler studies showed patent deep and superficial veins of the limb. Two expanded segments were seen in the great saphenous vein, which emitted low echo signals without blood flow (Fig. 1(B)). The MRI scan showed two dilatations in the great saphenous vein with abnormal signals (Fig. 1(C)). A T2-fat-suppression MRI demonstrated that the lump had a higher intensity and a smooth margin. There was no abnormal soft tissue surrounding the masses. Under epidural anesthesia, the lumps were dissected and confirmed to be in the great saphenous vein (Fig. 2(A)). Both masses were removed.
carefully, along with the intervening vein. The excised masses (Fig. 2(B)) were free from perivenous infiltration. The cut surface of the specimen was greyish-white and had a fish-flesh appearance. Hematoxylin and eosin (H&E) staining of sections revealed varying size tumor cells. Most were fusiform and arranged as fasciculations with smooth margin with eosinophilic cytoplasm and heteromorphic nuclei (Fig. 3). These cells stained positively for smooth muscle \(\alpha\)-actin, Vimentin and S100 markers by immunochemistry: CD10 staining was negative (data not shown). Therefore, the diagnosis of primary leiomyosarcoma of the great saphenous vein was made. The saphenous vein between two masses was normal. In order to prevent recurrence, postoperative radiotherapy was given. No local or distant metastases were identified within the first 10 months of follow up.

Discussion

Primary vascular leiomyosarcoma originates from smooth muscle of the vessel wall and is extremely rare. Since, the first case of venous leiomyosarcoma reported by Perl in 1871, less than 300 cases have been reported. More than 50% of them were from the inferior vena cava, 15% from the iliac veins and the rest from other veins.\(^2\,^3\) Leiomyosarcoma of the great saphenous vein is very rare. We describe a case with a further unusual feature, there being two separate masses of leiomyosarcoma along the same segment of great saphenous vein; this has not been reported previously. Reviewing the information from our case and the 26 previously reported cases, we obtain the following information. The median age of presentation was 54 years (range 2–85 years), with a 3:2 female to male ratio. The median size of the tumors was 4.1 cm (range 2–12 cm) and metastases were described in seven cases. If any form of adjuvant therapy is used it is usually radiotherapy. Chemotherapy is reserved for cases where metastasis occurs. Currently, the best treatment seems to be wide excision of the tumor combined with adjuvant radiotherapy.

The neoplastic cells may grow along the lumen of the vessels and are usually limited within the lumen. The presence of two separate masses with normal vein in between appearing as an hourglass is of interest. They could be of dual origin or the smaller proximal mass may be due to endoluminal seeding or other unknown
causes. The tumor usually expands slowly and is difficult to diagnose before surgery because of its rarity. Symptoms of tumor are not specific. The lesion may be palpable and tender. Nonspecific findings such as phlebitis, occlusion of the deep venous system, painful walking or lower limb edema, may be present. Primary leiomyosarcoma may be misdiagnosed as thrombophlebitis and lymphedema. Color Doppler ultrasonography may serve as a screening procedure and demonstrate a mass with low echo-level signal. MRI can contribute to localize the lesion and detect perivenous changes.

Fig. 2. (A) Lesions (arrows) seen during surgery. (B) Specimen after resection.

Fig. 3. Low and high power images of H&E staining of the tumor sections reviewed varied size of tumor cells. Most were fusiform and arranged as fasciculations with smooth margin. The nucleus was heteromorphic.
The venous leiomyosarcoma cells, located mostly in the vein wall, are well differentiated. The nuclei are pleomorphic. Histologically, the tumor may be misdiagnosed to be a malignant fibrohistiocytoma. Specific stains and immunohistochemical analysis help in the final diagnosis. Desmin staining and electronmicroscopy also have been used for diagnosis.\(^4\)

Surgical resection en bloc is the treatment of choice. Postoperative radiotherapy is recommended. Chemotherapy is reserved for those with metastases and for palliation in advanced cases.

References


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