

Leiomyosarcoma of somatic soft tissue origin in the leg - a case report

Jawahar Krishnaswamy^{1*}, Deivanayagam Shanmugam², Vijay Kalimuthu³, Vinod Balaji Baskar⁴

¹Associate Professor, ²Professor, ³Assistant Professor, ⁴P.G. Student, Department of Surgery, Saveetha Medical College and Hospital, Saveethanagar, Thandalam, Tamil Nadu.- 602105, INDIA.

Email: drkjawahar@gmail.com, dr_deiva@yahoo.com, viji_00765@yahoo.co.in, drvinodbalaji@yahoo.com

Abstract

Leiomyosarcoma of soft tissue is thought to arise from the smooth muscle cells lining small blood vessels. Most common site of involvement is retroperitoneum, accounting for approximately 50% of occurrences. Leiomyosarcoma of somatic soft tissues presents as an enlarging, painless mass. Although these tumours are generally associated with small blood vessels they usually do not present with signs or symptoms of vascular compression. However, when leiomyosarcoma arises from a major blood vessel, symptoms of vascular compromise or leg edema may be present, as well as neurologic symptoms such as numbness from compression of an adjacent nerve. We report a case of leiomyosarcoma of somatic soft tissue of the leg of a young male presented to us with swelling in the upper one-third of left leg which was associated with severe pain on extension of the joint. Patient was diagnosed as leiomyosarcoma of the somatic soft tissue origin by clinical, pathological and immunohistochemistry methods.

Keywords: Leiomyosarcoma, somatic soft tissue.

*Address for Correspondence:

Dr. Jawahar Krishnaswamy, Associate Professor, Department of Surgery, Saveetha Medical College and Hospital, Saveethanagar, Thandalam, Tamil Nadu.- 602105, INDIA.

Email: drkjawahar@gmail.com

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INTRODUCTION

Leiomyosarcoma is aggressive soft tissue sarcoma derived from smooth muscle cells typically of uterine, gastrointestinal or soft tissue origin. Sarcomas are malignant tumours arising from mesenchymal cell lines. Soft tissue sarcomas account for 0.7% of malignancies. Of all soft tissue sarcomas, approximately 5-10% are leiomyosarcomas. Leiomyosarcoma of soft tissue is thought to arise from the smooth muscle cells lining small blood vessels. The prognosis is poor, with survival rates among the lowest of all soft tissue sarcomas.

CASE PRESENTATION

A 23 year male patient, an Industrial worker by occupation, presented with swelling in the Upper 1/3 rd of left leg for 1yr. Swelling was progressively increasing in size with sudden increase in size since 2 months. Patient had severe pain on extension of the joint. he had no other symptoms and no history of loss of weight or appetite. On examination he had a Diffuse swelling of size 8x7 cm on the lateral aspect of the left leg, extending 4 cm from the knee joint below and laterally, 4cm below the popliteal fossa posteriorly. Surface was nodular, skin over the swelling was hyperpigmented. Dilated veins were present over the swelling. Swelling was warm and tender. Resting position of limb was in semi flexed- genu valgus position. Knee extension, dorsiflexion and eversion were Painful and decreased. There was no muscle wasting. peripheral pulsations were present with no foot drop and sensations over the limb was intact, No palpable lymph nodes in the popliteal fossa and no significant inguinal lymphadenopathy. Ultrasonogram of the left leg revealed a heterogenous mass in the intramuscular plane with features suggestive of a soft tissue sarcoma. A MRI of the leg was done which showed a well defined lobulated mass lesion partially encasing the neck and upper shaft of fibula with complete

encasement of the common peroneal nerve. A trucut biopsy of the swelling showed features suggestive of a malignant spindle cell sarcoma. A multidisciplinary team including orthopedician and plastic surgeon was set up and planned for wide excision of the tumour .Pt was clearly explained about the possibility of foot drop in the post operative period and after obtaining consent for the same he was taken up for surgery. Tumour was found adherent to the bone but not fixed, wide local excision including skin, underlying muscle and fascia with 2 cm margins was done. Since common peroneal nerve was found totally encased by the tumour, decision was taken to sacrifice the nerve. Tumour along with fibula and the skin with markings of the edges were sent for HPE.

Primary suturing of the wound by the plastic surgeons were done by undermining the edges. Patient was put on a POP slab. Post op drains were removed subsequently. Regular wound dressing were done. Patient subsequently developed foot drop and a foot drop splint was advised .Regular physiotherapy was given and he was able to walk without support. Histopathology showed leiomyosarcoma with no evidence of infiltration of muscle, bone or skin. All margins are free of the tumour infiltration. Pathological staging pT2B-pNx. Immunohistochemistry was done which showed the tumour cells were positive for CK, SMA, and focally for S100. EMA was negative.



Figure 1: Swelling lateral aspect of left leg

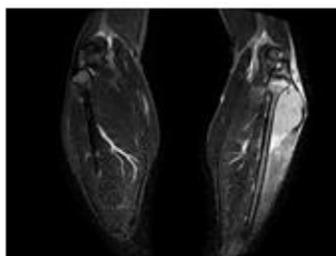


Figure 2: MRI leg



Figure 3: Per-operative picture showing the tumour

DISCUSSION

Sarcomas are malignant tumours arising from mesenchymal cell lines. Leiomyosarcoma of soft tissue is thought to arise from the smooth muscle cells lining small blood vessels. They are aggressive tumors that are often difficult to treat. The prognosis is poor, with survival rates among the lowest of all soft tissue sarcomas. Women are affected more than men (2:1), occurring in the 5th and 6th decades of life. Histologically, soft tissue leiomyosarcomas that arise in different anatomic locations are similar. However, based on the location of the tumor, prognosis and possible treatments differ. They are classified as Leiomyosarcoma of Soft Tissue – Retroperitoneum, Somatic soft tissue, Cutaneous Origin, Vascular Origin (large vessel), in the Immunocompromised Host and Bone. Leiomyosarcoma of somatic soft tissue involves the deep tissues of the extremities. They were once believed to arise from leiomyomas, extremely rare occurrence. Somatic soft

tissue leiomyosarcomas arise directly from the smooth muscle cells lining small blood vessels. Retroperitoneal leiomyosarcoma is an aggressive disease, not amenable to complete surgical resection. Gastro-intestinal leiomyosarcoma can arise from the smooth muscle in the GI tract. Initial imaging should include plain radiographs of the affected area, an MRI of the lesion, and a chest CT scan. MRI is the study of choice for evaluation of the anatomic extent of the tumor. Important considerations are the involvement of adjacent structures such as bone, nerves or compression of vascular structures. Angiography may be a useful in cases involving a major blood vessel. CT chest is useful to evaluate metastatic disease in the lungs. PET and PET-CT for evaluating patients for local disease recurrence, or in the search for metastatic lesions. Biopsy is necessary to establish a specific diagnosis of leiomyosarcoma, and is often accomplished using a CT guided core needle biopsy. Immunohistochemistry helps support the diagnosis by

demonstrating the presence of muscle specific markers including: desmin, muscle specific antigen (HHF35), cytokeratin (CK) and epithelial membrane antigen (EMA). Local control of soft tissue sarcomas is usually achieved with surgical resection. Pre-operative planning based upon radiographic and pathologic information is important to ensure adequate surgical margins. Achieving wide surgical margins is important in preventing local recurrence. Achieving a wide surgical margin is impossible when tumors involve or are directly adjacent to vital structures. Radiation therapy helps improving rates of local control when surgical margins are close, especially in high-grade sarcomas. Primary role of chemotherapy is in the treatment of metastatic disease. While not curative, it may slow the progression of systemic disease. Agents that are used in some sarcoma centers include: doxorubicin and ifosfamide, gemcitabine and taxotere (docetaxel), dacarbazine, and ecteinascidin.

CONCLUSION

Leiomyosarcoma is an aggressive sarcoma- remains one of the more difficult soft-tissue sarcomas to treat. Accurate diagnosis, classification, and multi-modality treatment by physicians who are familiar with these tumors are essential to favorable outcome. The rarity of these tumors makes definitive studies difficult to perform. Carefully designed prospective randomized clinical trials, may help further define the best treatment of these tumors in the future. Currently, in general, local control is obtained with wide surgical excision. Neoadjuvant or adjuvant radiation therapy is appropriate in some circumstances where local control is an issue. Chemotherapy is employed for the treatment of systemic disease.

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