Soft Tissue Sarcoma of Neural Sheath Origin

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A one-year-old female presented with right thigh soft tissue fungating tumor, which was diagnosed previously as soft tissue sarcoma. Amputation was recommended several times, but her father refused. The case was delayed with extended tumor including the whole thigh and became infected. Wide resection of the tumor was performed and the very small remaining part of quadriceps was sutured to cover the bone. Primary closure of the wound without skin graft was performed. The patient was discharged in a good condition. Upon follow-up a few weeks later, she was walking independently with healed surgical site.

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Soft tissue sarcoma is a group of diseases that originate from different aspects of connective tissues[1]. According to World Health Organization (WHO) International Classification of Diseases for Oncology, Third Edition (ICD-O-3), there are more than 50 histologic subtypes of STS according to cell type of origin and other histologic and molecular features[2]. At an early stage, the soft tissue tumor starts off as painless swelling. As it increases in size, it becomes symptomatic due to compression of neurovascular bundle or stretching of fascia[3-5].

The aim of this report to present a case of soft tissue sarcoma which was treated successively with amputation and reconstruction.

THE CASE

A one-year-old female presented with right thigh soft tissue, fungating tumor, which was diagnosed previously as soft tissue sarcoma. The biopsy revealed infantile fibrosarcoma. The father did not consent to amputation. The patient received two cycles of chemotherapy. The family tried traditional medicine with the injection of honey into the tumor. After which, the patient presented with infected deep gapped wound. The infection improved after supportive treatment. Chest CT scan revealed no evidence of dissemination. The MRI was performed to assess surgical resection. However, limb sparing surgery (LSS) was not possible, therefore, amputation was recommended, but her father still refused. Two months later, the mass involved the circumstance of the thigh, together with lung nodules and pelvic lymph nodes. The patient received 3 cycles of chemotherapy, but she had limited clinical response. Tumor board advised palliative treatment and chemotherapy because of the family refusal for amputation. Her father also refused chemotherapy due to its complications. The case was referred to a higher center, but was not accepted.

The patient presented later with a fungating tumor with foul smell. X-ray of femur showed a fungating soft tissue tumor. Chest CT was unremarkable. MRI of the thigh showed a large soft tissue mass (7.2x8.3x9 cm) involving the quadriceps muscle and displacing the neurovascular bundle medially with no invasion. Open biopsy revealed soft tissue sarcoma of neural sheath origin. Culture swab showed Klebsiella and pseudomonas aeruginosa species growth. The patient received ceftazidime and floxacrine. A team composed of a vascular surgeon, plastic surgeon, medical oncologist, and radiation oncologist was consulted and recommended amputation as the best option. The option was discussed with the family, emphasizing the risk of metastasis to the lung, invasion of neurovascular bundle, and development of septicemia, however, the family refused amputation again. A limb sparing procedure with wide resection of the tumor was performed after explaining the high risk of recurrence and other complications such as neurovascular injury, see figure 1.

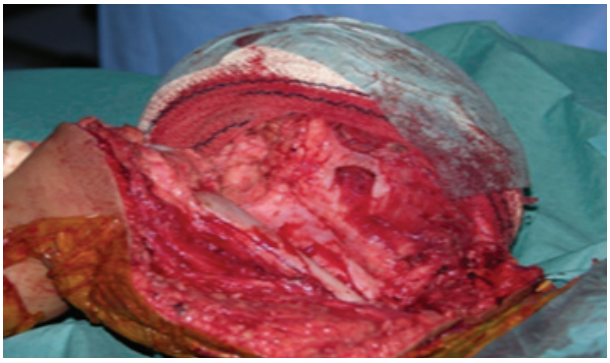

Figure 1: Wide Resection of the Tumor

Wide resection of the tumor was performed and the small remaining part of the quadriceps was sutured to cover the bone, see figures 2 and 3. Primary closure of the wound without skin graft was performed.

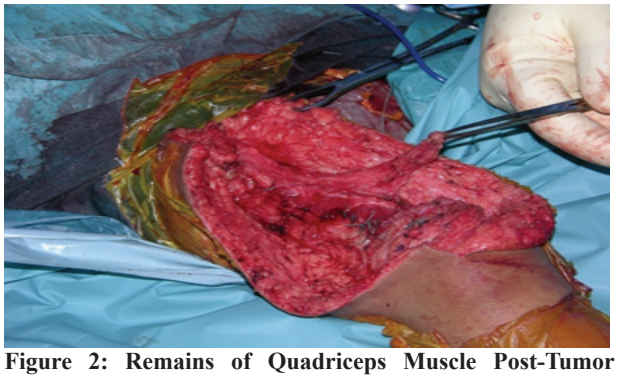

Figure 2: Remains of Quadriceps Muscle Post-Tumor Excision

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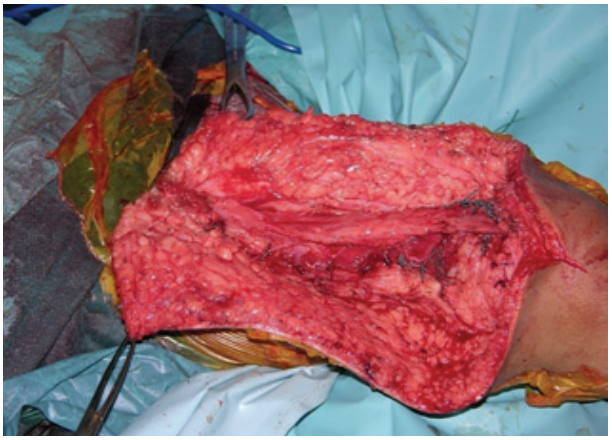

Figure 3: Small Remaining Part of Quadriceps was Sutured to Cover the Bone

The resected tumor was sent to histopathology and all tumor margins were negative. The patient was seen few weeks later, walking independently with healed surgical site.

DISCUSSION

Soft tissue sarcomas are rare; proper evaluation is critical in management. Unplanned excision of a soft-tissue sarcoma may negatively impact patient outcome. Delay in diagnosis can adversely affect the ultimate outcome; it may be confused with more common pathology. The lung is the most common site of metastasis[6,7]. A mass greater than 5cm growing deep into the superficial fascia should be presumed to be a soft tissue sarcoma until proven otherwise, and assessed with three-dimensional imaging first. MRI is the best imaging modality for defining the anatomy and helping characterize the lesion. CT scan of the chest is required to evaluate metastasis. CT scan of the abdomen and pelvis is obtained for liposarcoma was performed because of synchronous retroperitoneal liposarcoma. Radiation therapy is an important adjunct to surgery in the treatment of soft tissue sarcomas. Excisional biopsy should not be performed when the origin of a soft tissue tumor is unknown.

CONCLUSION

Soft tissue sarcomas are not easy to manage especially if presented at a young age due to its complications which may include disability. Availability of other treatment option is not common but should not be neglected in young patients to save their functional ability.

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