Soft Tissue Tumors

25

Recep Öztürk

Overview

The definition of soft tissue mass more commonly includes benign soft tissue tumors and reactive lesions, but also includes a rare and more serious group of soft tissue sarcomas.

Soft tissue tumors are rare and are a heterogeneous group of tumors consisting of many subgroups with different natural trends, biologies, and treatment responses. For this reason, despite the availability of immunohistochemistry markers and studies on genetic and biochemical new markers in recent years, defining histopathological subgroups may be difficult.

25.1 Definition

Soft tissue tumors include the group of mesenchymal tumors seen in non-epithelial tissues. These tumors are grouped mainly by their histogenesis, and cytogenetic and molecular genetic information is included in these examinations. The presence of more than 100 benign and malignant soft tissue tumors has been identified.

R. Öztürk (✉)
Department of Orthopedics and Traumatology, Dr. Abdurrahman Yurtaslan Ankara Oncology Training and Research Hospital, Ankara, Turkey
e-mail: recep.ozturk6@saglik.gov.tr

25.2 Epidemiology

Soft tissue sarcomas are known to cover about 0.5% of all cancers, and benign/malignant ratio of soft tissue tumors is about 100:1. About 13,000 cases are reported annually in the USA, and in Europe, approximately 23,000 new cases of soft tissue sarcoma are reported.

These tumors can be seen at any age; 15% of cases occur in childhood. Men are more affected compared to women [M:F: 1.4:1]. About 40% of these tumors are located in the lower extremities, 20% in the upper extremities, 30% in the trunk and retroperitoneum, and 10% in the head and neck.

25.3 Etiology/Pathogenesis

Benign and malignant soft tissue tumors often develop de novo without identifiable risk factors or predisposing conditions.

In some cases, radiation, chemical burns, thermal burns, trauma, phenoxyherbicide exposure, and HHV8 virus (Kaposi's sarcoma) have been identified.

In addition, some genetic diseases (neurofibroma, malignant peripheral nerve sheath tumor in neurofibromatosis type 1; fibromatosis in Gardner's syndrome, etc.) have an increased risk of developing soft tissue tumors.

UG. Longo, V. Denaro (eds.), *Textbook of Musculoskeletal Disorders*,
https://doi.org/10.1007/978-3-031-20987-1_25

25.4 Classifications

Soft tissue tumors are classified histologically by predominant cell type. There are hundreds of different soft tissue tumors, some of which are given in Table 25.1.

The most commonly used system for staging is the American Joint Committee on Cancer (AJCC) system (Table 25.2). The order of importance of prognostic factors is presence of metastasis (stage IV), grade (low grade—stage I, high grade—stage II), size (T1 ≤5 cm, T2 >5 cm), and location (superficial or deep) [3].

Table 25.1 Histological classification of soft tissue tumors

Histologic type	Benign	Malignant
Adipocytic	Lipoma	Well-differentiated liposarcoma
Fibrous	Nodular fasciitis	Fibrosarcoma
Nerve sheath	Schwannoma	MPNST
Skeletal muscle	Rhabdomyoma	Rhabdomyosarcoma
Smooth muscle	Leiomyoma	Leiomyosarcoma
Synovial	Focal PVNS	Malignant PVNS
Vascular	Hemangioma	Angiosarcoma

DFSP dermatofibrosarcoma protuberans, *MPSNT* malignant peripheral nerve sheath tumor, *PVNS* pigmented villonodular synovitis

Table 25.2 AJCC version 8 staging for soft tissue sarcomas

	Primary tumor (T)
Tx	Primary tumor cannot be assessed
T1	Tumor ≤5 cm
T2	5 cm < tumor ≤10 cm
T3	10 cm < tumor ≤15 cm
T4	Tumor >15 cm
	Regional lymph nodes (N)
N0	No regional lymph node metastasis or unknown
N1	Regional lymph node metastasis
	Distant metastasis (M)
M0	No distant metastasis
M1	Distant metastasis
	Staging
IA	T1; N0; M0; G1
Ib	T2, or T3, or T4; N0; M0; G1
II	T1; N0; M0; G2 or G3
IIIA	T2; N0; M0; G2 or G3
IIIB	T3, or T4; N0; M0; G2 or G3
IV	Any T; N1; M0; any G or any T; any N; M1; any G

25.5 Diagnosis

25.5.1 Story

The story includes current symptoms and related history/family history.

As current symptoms, pain, palpable mass presence, and growth rate of the mass should be evaluated. A soft tissue sarcoma may not cause any symptoms or signs in the early stages. As the tumor grows, it can cause a noticeable lump or swelling. Pain may occur when pressing on muscles or neural structures. Rapid growth may be an indicator of malignancy, but malignancy may also be considered in a long-standing and slow-growing soft tissue mass.

Related story/family story includes the presence of cancer or syndrome (like neurofibromatosis) in the patient or in the family, exposure to carcinogens, infection, or trauma (myositis ossificans).

25.5.2 Physical Examination

Mass presence (compressible lesions suggest benign tumors, and firm masses suggest sarcomas or desmoid tumors), range of motion (tumor-associated joint range of motion may decrease), muscle atrophy (painful lesion), and lymphadenopathy (lymph nodes may also grow due to infection or metastasis) are examined.

25.5.3 Imaging

Plain radiographs: Scattered calcifications (at 30%) in synovial sarcomas, peripheral mineralization in myositis ossificans, phleboliths in hemangiomas, and radiolucent lesions in lipomas can be seen on radiographs.

MRI: It is the gold standard imaging method in soft tissue imaging. The nature of some lesions can be determined by MRI (in lesions such as lipoma, hemangioma, ganglion, muscle damage), and definitive treatment can be given in these lesions. In masses where the nature of the lesion cannot be determined with MRI, biopsy should be performed first. In the majority of sarcomas, the diagnostic image is uncertain in terms of

malignancy, and biopsy is required. Even if a lesion is small and painless, when a specific diagnosis cannot be made, potential malignancy should be considered and not merely observed.

CT: Soft tissue sarcomas most often metastasize to the lung. Lung CT is used to detect lung metastases.

25.5.4 Biopsy

It is the most important step in the diagnosis of many soft tissue lesions. Biopsy should be done by a team that will perform the final surgery at a center specialized in tumors. Inappropriate biopsy can lead to misdiagnosis due to the intake of tissue that does not represent the mass, or complications such as infection and neurovascular damage. In addition, the unplanned biopsy location can harm the final surgery. Contamination or inappropriate incision line during biopsy can cause more complicated surgeries that require a flap/graft or lead to unnecessary amputations.

Needle biopsy (fine needle or true-cut), open incisional biopsy, and excisional biopsy options are available. The most common method is needle biopsy, which requires experienced pathologist. In open incisional biopsy, the rules of biopsy should be followed at the highest level, and very good hemostasis should be performed. Excisional biopsy can be performed if the surgeon is sure that the lesion is benign.

25.6 Treatment

Treatment in soft tissue tumors is planned according to the biological activity of the lesion. In benign tumors, inactive-latent lesions (superficial lipoma, etc.) are followed, inactive-symptomatic or active lesions (intramuscular myxoma, etc.) are treated with simple excision, and aggressive lesions (extra-abdominal fibromatosis, etc.) are treated with wide resection.

In the treatment of soft tissue sarcoma, wide resection or combination of resection with external beam radiotherapy is applied. Preoperative or postoperative radiotherapy can be administered. Amputation is applied to the tumor in cases where wide resection cannot be performed or a functional limb cannot be obtained after wide resection. Today, about 90% of patients can be operated with limb-sparing surgery. Surgical margins are shown in Fig. 25.1.

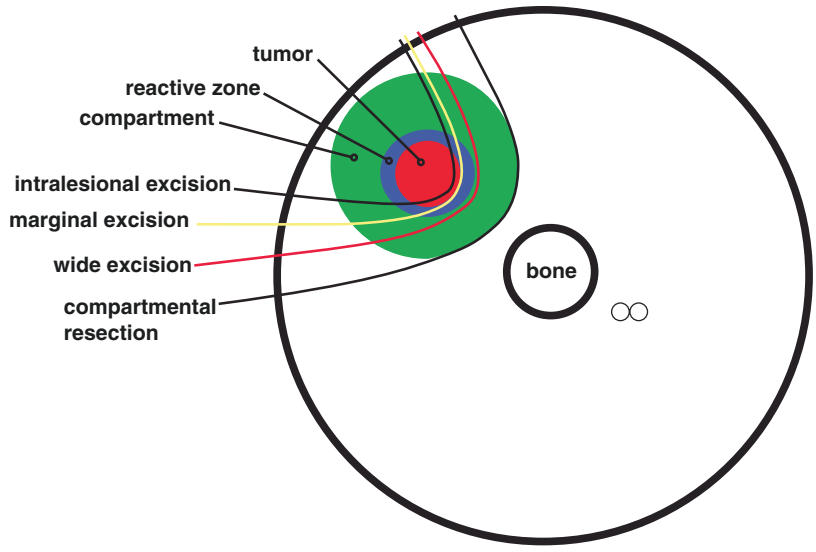

Fig. 25.1 Surgical margins: Intralesional excision is the method in which the surgical incision passes through the tumor and the macroscopic residual tumor is left behind. Marginal excision is the method where the surgical incision passes through the reactive region between the tumor and the pseudocapsule. Wide excision is the method where the surgical incision is outside the reactive region and pseudocapsule. The incision passes through normal tissue of all sizes. Compartmental excision is the excision of a tumor with an anatomical chamber containing the tumor

Take-Home Message

- Since soft tissue tumors are particularly rare and the majority of them are benign, there may be important problems in the diagnosis and treatment of malignant tumors.
- Soft tissue sarcomas are often painless masses, unlike bone sarcomas.
- Soft tissue sarcomas usually have well-defined margins, so it is difficult to distinguish them from benign soft tissue lesions in MRI.
- For lesions with malignant tumor characteristics (large and deep localization), STS should be kept in mind and biopsy should be performed.
- Unplanned biopsies and intralesional interventions can make extensive resection impossible or cause amputation.

Summary

Soft tissue tumors are divided into two main groups as benign and malignant. The most common treatment in benign STTs is simple excision. In soft tissue sarcomas, the basis of treatment is resection of the tumor with wide borders. In addition, RT is added to the treatment in appropriate cases.

The importance order of prognostic factors in soft tissue sarcomas is the presence of metastasis, grade, size, and localization. Soft tissue sarcomas most often metastasize to the lung. It is mainly detected by lung CT.

Questions

Multiple correct answers are possible. Answers available in the book back matter.

1. Which of the following is wrong with soft tissue tumor surgery?
   (a) Macroscopic tumor is left behind in intralesional excision
   (b) Marginal excision is the removal of the entire tumor
   (c) The preferred surgical treatment in soft tissue sarcomas is marginal excision
   (d) In wide resection, the surgical incision line passes through normal tissue
2. A 26-year-old male patient has a soft tissue mass on his left elbow. Figure A shows plain radiograph, and Fig. B and Fig. C show MRI images. What is the next most appropriate procedure for this patient?
   (a) Observation
   (b) Needle biopsy
   (c) Marginal excision
   (d) Wide excision
3. In which of the following, phleboliths can be seen on direct radiography?
   (a) Synovitis
   (b) Lipoma
   (c) Fibromatosis
   (d) Hemangioma
4. A 44-year-old woman has a painless mass on the posterior of the left knee. Figure A shows plain radiograph, and Fig. B and Fig. C show MRI images. What is the next most appropriate procedure for this patient?
   (a) Observation
   (b) Medical treatment
   (c) Needle biopsy
   (d) Marginal excision
5. After a simple excision of a 4 cm superficial mass on the right thigh of a 42-year-old male patient, the result of pathology was synovial sarcoma. No information was given about the surgical margins. What is the next most appropriate procedure for this patient?
   (a) Observation
   (b) Chemotherapy
   (c) Radiotherapy (to the tumor bed)
   (d) Wide resection of the tumor bed

Further Reading

Rochwerger A, Mattei JC. Management of soft tissue tumors of the musculoskeletal system. Orthop Traumatol Surg Res. 2018;104(1S):S9–S17.

Weber K, Rose P. Section 11: oncology. In: Lieberman JR, editor. AAOS Comprehensive Orthopaedic review. Rosemont, IL: American Academy of Orthopaedic Surgeons; 2020.

Papapietro N, Longo UG, Palumbo A, Bianchi A, Maffuli N, Denaro V. Synovial sarcoma of the anterior tibialis tendon. J Am Podiatr Med Assoc. 2012;102(2):157–60.