Definition

- **Epidemiology**
  Rare highly malignant tumor (round-cell sarcoma) occurring in childhood and adolescence • Age 5–20 years.
- **Etiology, pathophysiology, pathogenesis**
  Closely related to PNET • Permeative osteolytic tumor arising from the medulla, usually with a large soft tissue component • The intervertebral disk is spared • Arises from the vertebral body (occasionally from two) • Only 5% of lesions are sclerotic • Extraosseous soft tissue component in over 50% of cases • Metastases (lung, bone, lymph nodes) arise in over 20% of cases.

Imaging Signs

- **Modality of choice**
  Conventional radiographs, CT (diagnosis) • MRI (extent of tumor).
- **General**
  Aggressive permeative (moth-eaten) osteolysis • Periosteal reaction • Soft tissue component (50% of lesions).
- **Radiographic findings**
  Permeative bony destruction with a broad transitional zone • Periosteal reaction ("onion peel" appearance, "hair on end," Codman triangle) on the vertebral bodies is often not distinct • Compression fracture.
- **CT findings**
  Bony destruction • Infiltration of paravertebral structures • Soft tissue component.
- **MRI findings**
  T1 hypointensity, significant enhancement after contrast administration • T2 and STIR hyperintensity • Evaluation of soft tissue component • Infiltration of paravertebral and intraspinal structures • *Caution:* Hemorrhage and necrosis can alter the appearance of the lesion.
- **Nuclear medicine**
  Pronounced radionuclide uptake (searching for metastases).

Clinical Aspects

- **Typical presentation**
  Pain • Fever and leukocytosis (mimics osteomyelitis) • Swelling • Developmental anomaly • Pathologic fractures present in over 10% of cases.
- **Therapeutic options**
  Preoperative chemotherapy (goal: 90% reduction in tumor size); combination modality therapy • Resection followed by chemotherapy.
- **Course and prognosis**
  In 50% of cases with localized disease, survival is good • Rate of tumor recurrence is over 15%.

Imhof, Direct Diagnosis in Radiology. Spinal Imaging
(ISBN 9783131440716), © 2008 Georg Thieme Verlag KG

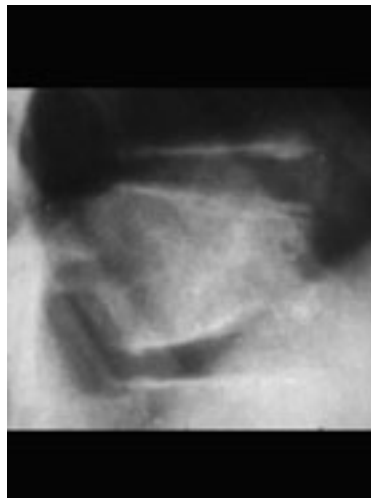

Fig. 5.41 Fatigue, slightly elevated temperature, leukocytosis. Conventional lateral radiograph of the thoracic spine (detail). Moderately sclerotic anteriorly compressed vertebral body. Tumor has penetrated the posterior cortex. The matrix is highly inhomogeneous, a sign of osteoblastic and osteolytic posterior bone remodeling.

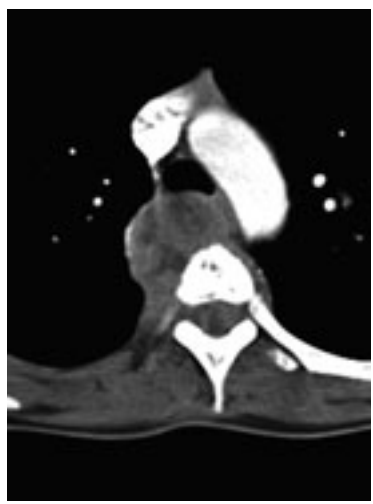

Fig. 5.42 CT of the thoracic spine (axial). Large right anterolateral soft tissue tumor with impression of the trachea. Lesion extends to the aortic arch.

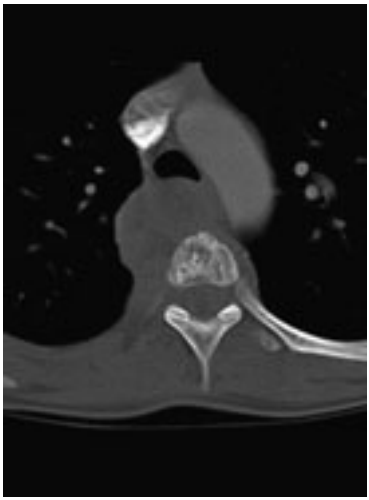

Fig. 5.43 CT of vertebra T8 (axial, bone window). Extensive osteolysis of the vertebral body with paravertebral tumor masses isodense to soft tissue.

Differential Diagnosis

Neuroblastoma	– PNET can only be distinguished by histologic examination
Langerhans cell histiocytosis	– More geographic lesions
Osteosarcoma	– 80% involve matrix calcification
Eosinophilic granuloma	– Histiocytosis X
Osteomyelitis	– Fever, leukocytosis, elevated erythrocyte sedimentation rate – Intervertebral disk also affected

Selected References

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