Epithelioid Hemangioma

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Abstract

Epithelioid hemangioma is a special type of hemangioma characterized by a benign although locally aggressive behavior, composed by proliferated small-caliber blood vessels lined by plump endothelial cells with an epithelioid morphology. It is rare in bone, where it grows in a more solid fashion than its soft tissue counterpart. It is seen more often between 20 and 60 years of age. Long bones of the extremities are preferably affected and may be multifocal. EH is usually painful. Regional lymph node metastases and soft tissue extension of the tumor do not mean altered aggressiveness. Besides endothelial markers, epithelial markers are positive. It has a 10 % recurrence risk.

Definition

- A special type of hemangioma characterized by a benign although locally aggressive behavior, composed by a proliferation of small-caliber blood vessels lined by plump endothelial cells with an epithelioid morphology. It is rare in bone, where it grows in a more solid fashion than its soft tissue counterpart.

Epidemiology

- It is a rare tumor in bone.

Age

- Can affect any age group but is seen more often between 20 and 60 years of age

Sex

- Sexes appear to be equally affected.

Sites of Involvement

- Long bones of the extremities are preferably affected. The feet, pelvis, ribs, vertebrae, and hands follow in frequency.
- May be multifocal (20–25 %), affecting different foci in the same bone or, more rarely, several bones of the same anatomical area as well as soft parts.

Signs and Symptoms

- Different from classical hemangiomas, EHs are usually painful and swelling may be present.
- May be asymptomatic.

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Image Diagnosis

Radiographic Features

- Radiographs show an expanded bone with cortical erosion.
- Internal trabeculations may be seen.

CT Features

- Expansile, septated, lytic lesion that may present soft tissue extension.
- Trabeculations are more readily seen with this image method.
- Lesions and their well-limited contour, inside and outside the bone, are enhanced by contrast injection.

MRI Features

- On MRI, EH has a usually low signal intensity on T1-weighted and high signal intensity on T2-weighted image.
- Its limits, inside and outside the bone, are clearly demarcated.

Imaging Differential Diagnosis

Angiosarcoma and Hemangioendothelioma

- Are radiographically less delimited lesions

Cystic Angiomatosis

- Presents multiple widespread skeletal and visceral lesions. Soft tissue lesions usually contain phleboliths.

Pathology

Gross Features

- Radiography of the specimen is helpful to localize multiple lesions and orient sectioning.
- The cut surface shows a well-delimited lesion, mostly hemorrhagic and soft, with spongy areas alternating with sometimes more solid areas.
- Cortical destruction and soft tissue tumor may be apparent but the limits are usually clear.

Microscopic Features

- EH is composed of lobules of neoplastic blood vessels presenting a kind of zonation phenomenon, where the vessels found at the periphery of the lobules are hypocellular, with flattened endothelium, that gradually turns more cellular towards the center of the lobules. The cells get plumper, with epithelioid features, abundant eosinophilic cytoplasm, sometimes resembling the so-called "tombstone-like" aspect; the foci of solid epithelioid cell proliferation, devoid of evident vascular lumina, may be seen in the center, leading to a differential diagnosis with EHE. The nuclei of these cells are typically round or oval, sometimes lobulated, and occasional nucleoli may be seen. Mitoses occur in an average of 1/10 high-power fields and atypical mitoses are not seen. The stroma is loose, with frequent eosinophils and a few other mononuclear inflammatory cells.
- Less frequent features are intracytoplasmic vacuoles, occasionally containing red cell fragments, osteoclast-like giant cells, necrosis, dense inflammatory foci, and new bone formation.
- Host bone trabeculae may be seen entrapped between the neoplastic lobules.
- The tumor expands the bone by growth pressure of its pushing borders and erodes but does not infiltrate the cortex.

Pathology Differential Diagnosis

Epithelioid Hemangioendothelioma

- These tumors usually present chords and small groups of cells in a myxohyaline or chondroid matrix, which is not seen in EH.
- Presents translocation t(1;3)(p36.3;q23-25) resulting in the WWTR1-CAMTA1 fusion, not found in EH.

Angiosarcoma

- Presents large irregularly anastomosing vascular channels, intense cellular atypia, and high mitotic activity

Ancillary Methods

Immunohistochemistry

- Endothelial markers (CD34, CD31, Fli1, vW, and ERG) are expressed by the tumor cells.
- Epithelial markers are frequently positive in the neoplastic epithelioid cells.

Ultrastructure

- Weibel-Palade bodies may be seen, characterizing endothelial cells.

Prognosis

- Presents a 10 % recurrence risk.
- EH is a benign tumor and wide metastatic spread and death from disease does not occur.
- Regional lymph node metastases and soft tissue extension of the tumor do not mean altered aggressiveness.

Treatment

- Thorough curettage or en bloc surgical resection is the preferred choice of treatment.
- Preoperative embolization has been used.
- Radiation therapy has also been used in incompletely resected tumors, usually as a complement to surgery. The risk of secondary radiation effects has to be evaluated against the risk of an observation attitude.

Images

See Figs. 33.1, 33.2, 33.3, 33.4, 33.5, 33.6, and 33.7 for illustrations of epithelioid hemangioma.

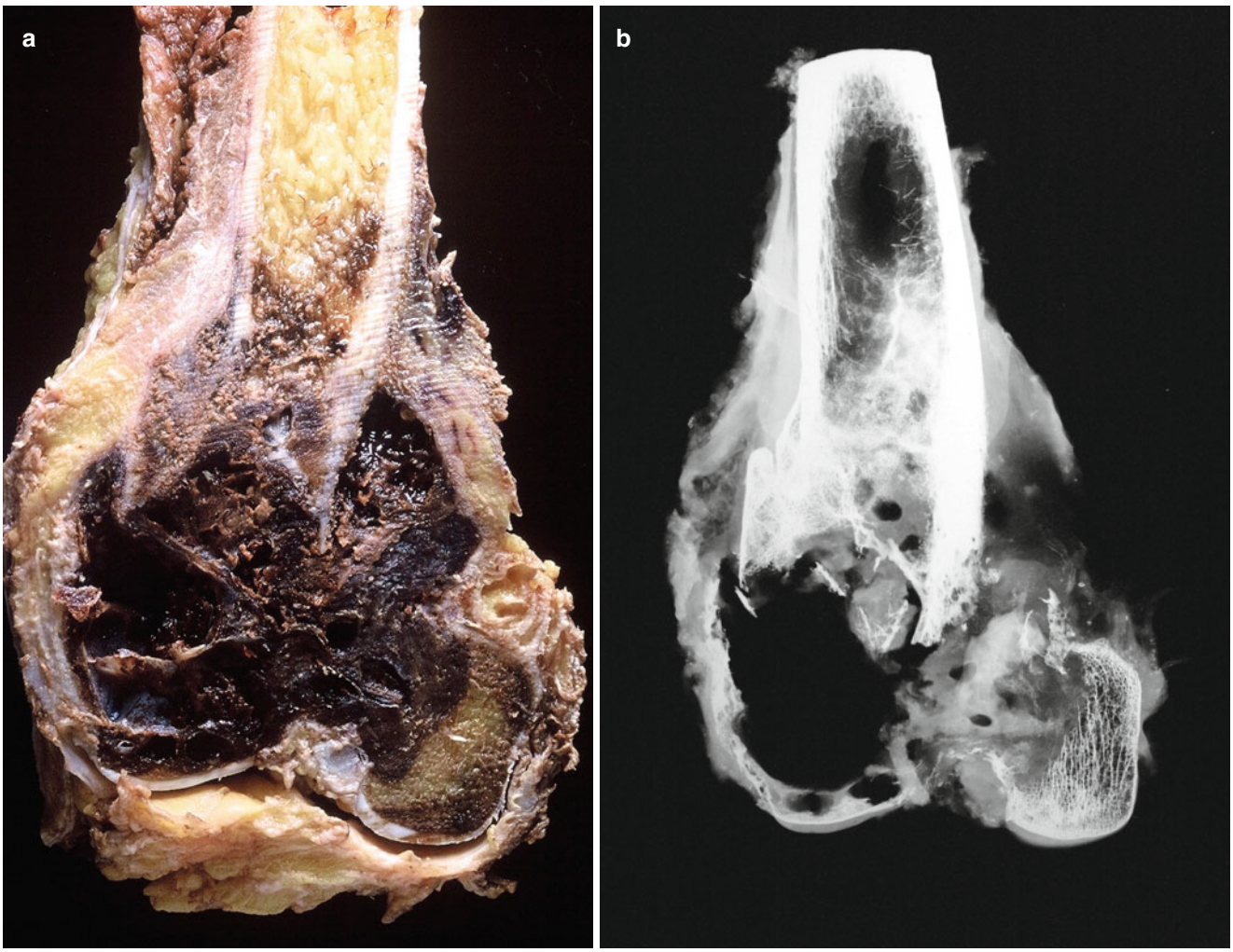

Fig. 33.1 (**a**) Macrophotography and (**b**) specimen radiograph of an epithelioid hemangioma of the distal femur with pathological fracture. The lesion has a hemorrhagic pattern and expands to soft tissue

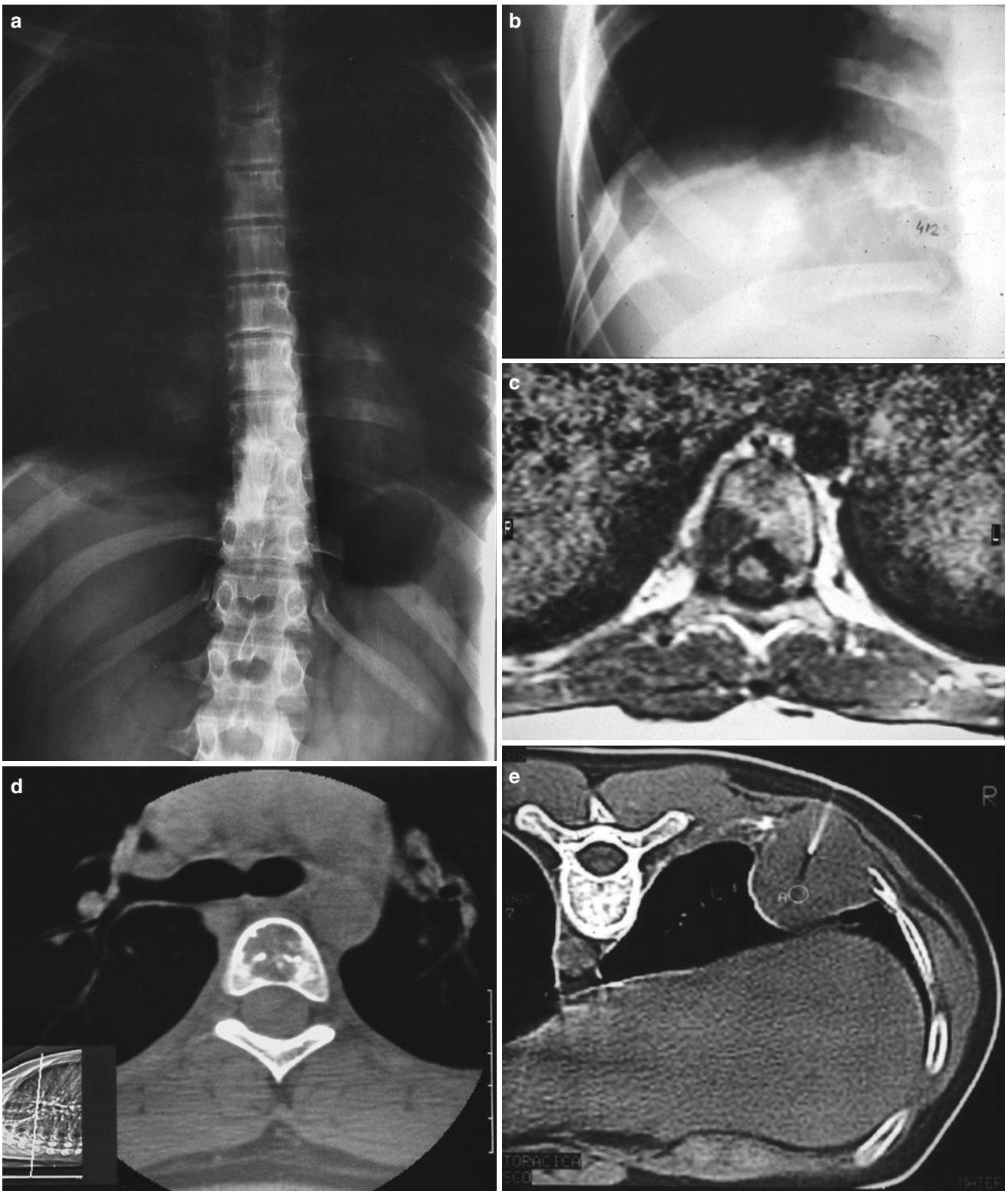

Fig. 33.2 Multifocal EH. (**a** and **b**) Radiographs showing lesions in vertebrae and a rib. (**c** and **d**) MRI and CT of a vertebral lesion. (**e**) CT-guided needle biopsy of the rib lesion

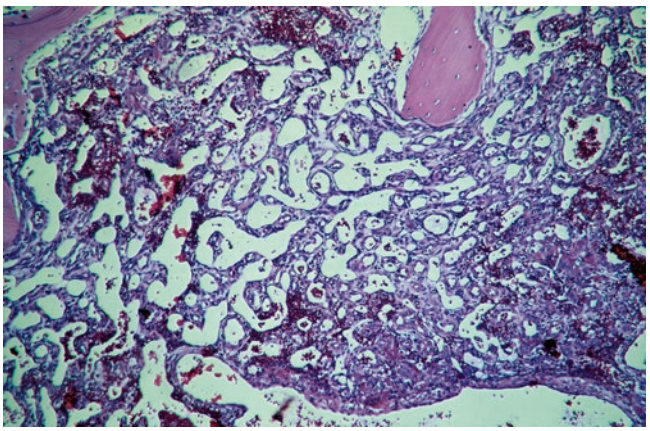

Fig. 33.3 Low-power microscopic view of EH. Endothelium is more flattened at the periphery, *left*, and more epithelioid towards the center of the lesion

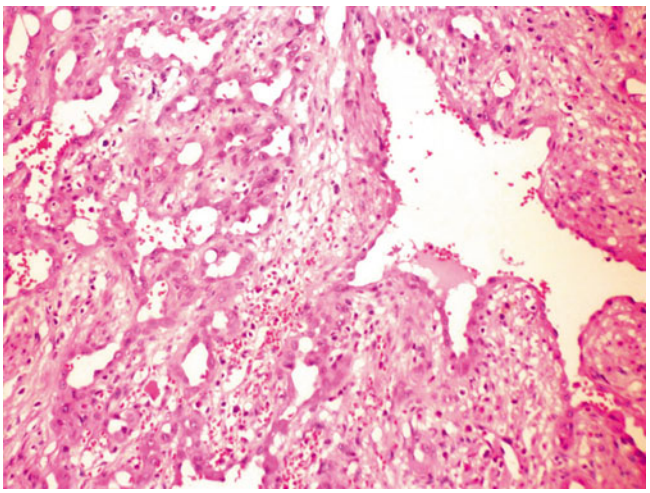

Fig. 33.4 Medium-power view: irregular blood vessels lined by epithelioid endothelial cells

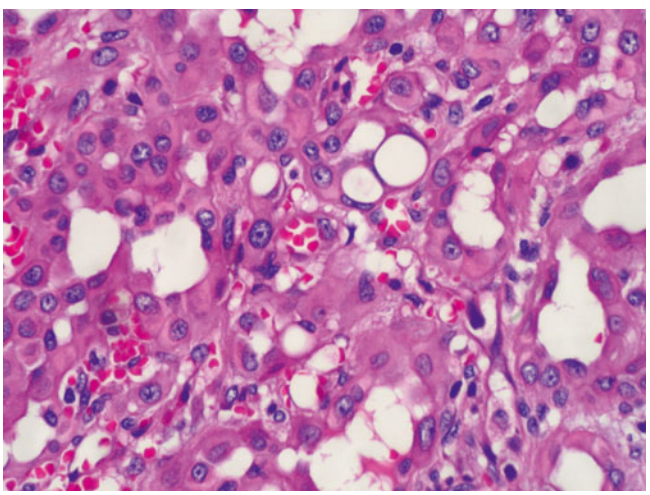

Fig. 33.5 Higher power of a more compacted cellular area

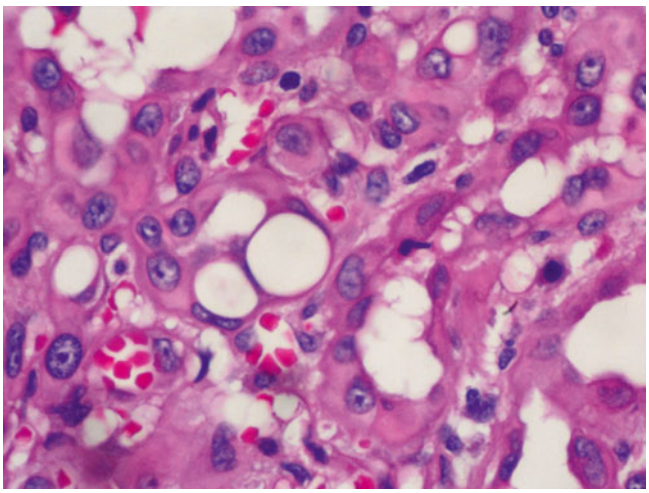

Fig. 33.6 High-power microscopic image. Large, epithelioid cells, with eosinophilic cytoplasm with occasional intracytoplasmic vacuoles and round to oval, sometimes lobulated, nuclei

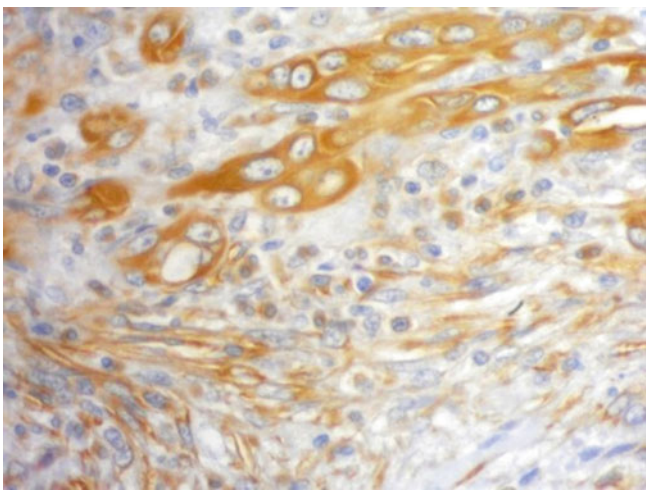

Fig. 33.7 Immunohistochemistry. CD34-positive epithelioid cells

Recommended Reading

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