Aneurysmal Bone Cyst (ABC)

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Definition: Benign tumor characterized by cystic cavities lined by septa containing spindle cells, inflammatory cells, giant cells, and histiocytes with hemosiderin deposition.

Production of variable amounts of reactive bone is frequently encountered. The cavities are filled with flowing blood. This lesion, when primary, is now considered to be tumoral, because in about 40–50% of cases, it has been demonstrated the presence of typical translocation t(16; 17) (q22; p13) and t(17; 17) (q22; p13) that produce the chimeric proteins CDH11-USP6 and COL1A1-USP6, respectively. When this lesion is described side by side with another bone tumor (e.g., giant cell tumor, chondroblastoma, or fibrous dysplasia), it is considered reactive and does not display any kind of genetic abnormality.

Importance: It is often mistaken for a malignant tumor because of its possible radiological and pathological aggressiveness.

Epidemiology: Primary lesions represent about 2% of primary bone tumors. No differences in gender. They are found mostly in the second decade of life (85% under 20 years of age; rare above 50).

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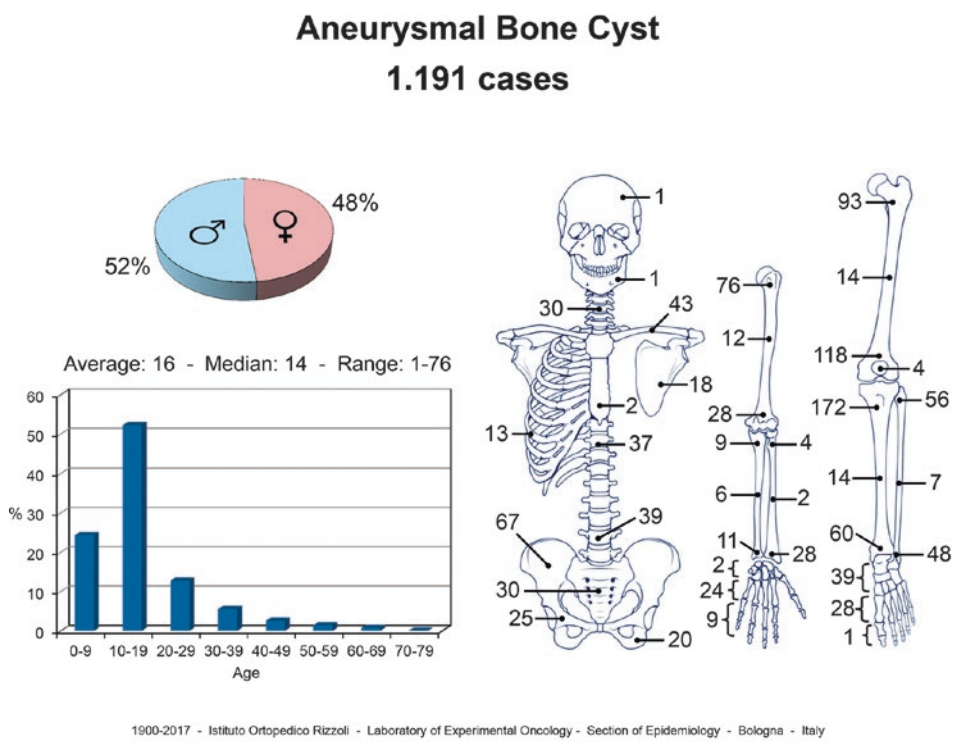


Localization: Any osseous segment may be affected, but the most common sites are the metadiaphyses of the long bones and the spine.

Clinical: Pain and swelling generally of less than 3 months duration. Frequent report of a previous trauma in the same skeletal site. Onset or progression of symptoms has been observed in some cases during pregnancy.

Imaging: The characteristic radiographic appearance of ABC is a subperiosteal, poorly defined osteolysis, elevating and inflating the periosteum, and progressively eroding the cortex. CT and MRI are often helpful in showing fluid levels within the cyst. Angiograms show intense and persistent accumulation of contrast media. Isotope scan commonly reveals an increased uptake with a cold central area.

Histopathology: Grossly it appears as a blood-filled sponge limited by fibrous septa. The wall of the cystic cavities is not formed by endothelial cells but by mesenchymal tissue rich in fibroblasts, histiocytes, thin capillaries, and scattered multinucleated giant cells. Immature trabeculae of reactive bone may be seen. Blotchy purple calcifications (considered pathognomonic by some authors) can be seen. Brisk mitotic activity is frequent.

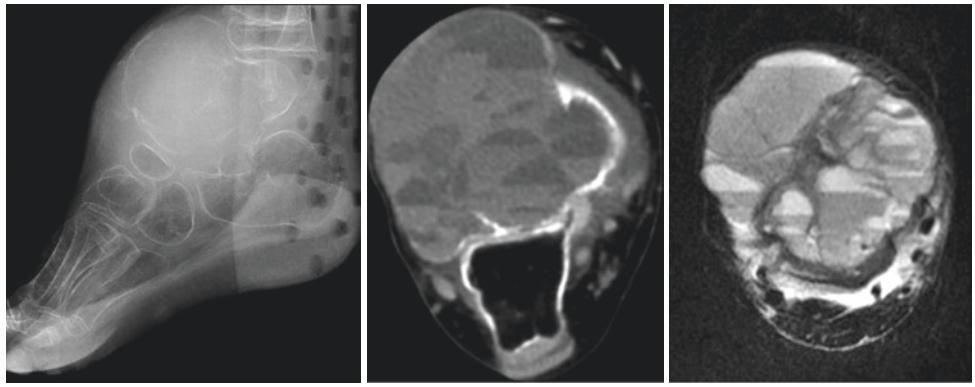
Course and Staging: The lesion may progress very rapidly, but in some cases it may heal spontaneously, after a fracture or a biopsy. Recent cytogenetic data have demonstrated the presence of fusion oncogenes in a subgroup of ABCs. Nevertheless, its origin is controversial, ABC is commonly staged as a benign tumor.

Treatment and Prognosis: After intralesional surgery, local recurrence occurs in a 20% of cases. Selective arterial embolization of the nutrient vessels represents the standard treatment in difficult locations (spine or pelvis) or big proximal lesions (proximal femur, proximal humerus). Various minimally invasive treatments have been used with satisfactory results, as curopsy, sclerotherapy, cryotherapy, injection with concentrated bone marrow, and demineralized bone matrix. Radiation therapy (30–40 Gy) has been proven effective in inducing cyst ossification, but it is charged by the risk of sarcoma induction, or growth plate damage in children.

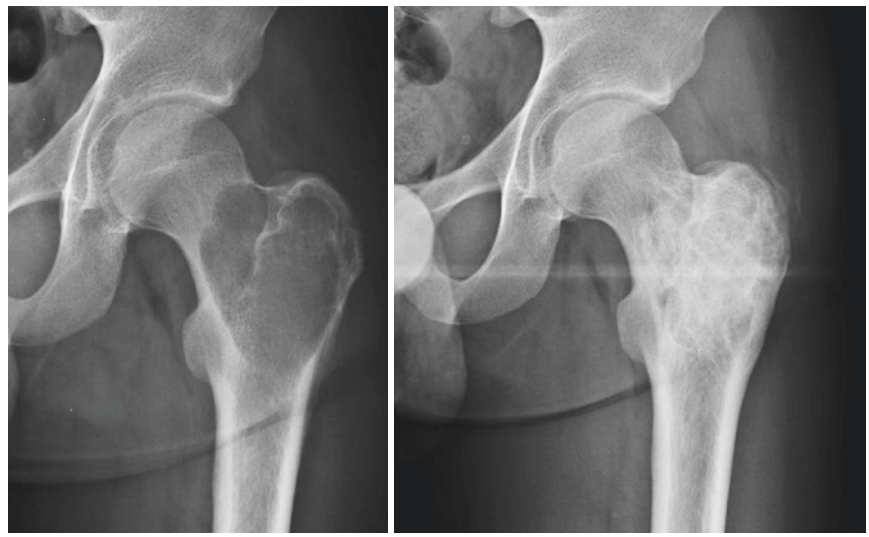
Key points	
• Clinical	Pain and swelling in young people
• Radiological	Eccentric, lytic lesion. Fluid/fluid levels
• Histological	Cystic cavities and septa not lined by endothelial cells
• Differential diagnosis	Telangiectatic osteosarcoma

Chromosomal translocations		
• t(16;17)(q22;p13)	CDH11-USP6	30–50%
• t(17;17)(q12;p13)	COL1A1-USP6	5%

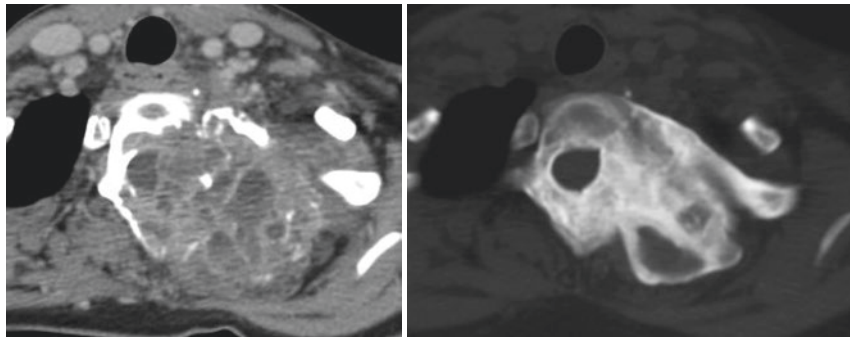
Chromosomal translocations		
• t(1;17)(p34,3;p13)	THRAP3 (TRAP150)-USP6	Rare
• t(3;17)(q21;p13)	CNBP(ZNF9)-USP6	Rare
• t(9;17)(q22;p13)	OMD-USP6	Rare
• t(17;17)(q13.3;p13)	PAFAH1B1-USP6	Rare
• t(6;17)(q21.1;p13)	RUNX2-USP6	Rare
• t(3;17)(p22.1;p13)	CTNNB1-USP6	Rare
• t(4;17)(q21.22;p13)	SEC31A-USP6	Rare
• t(17;17)(q21;p13)	EIF1-USP6	Rare
• t(2;17)(p23.2;p13)	FOSL2-USP6	Rare



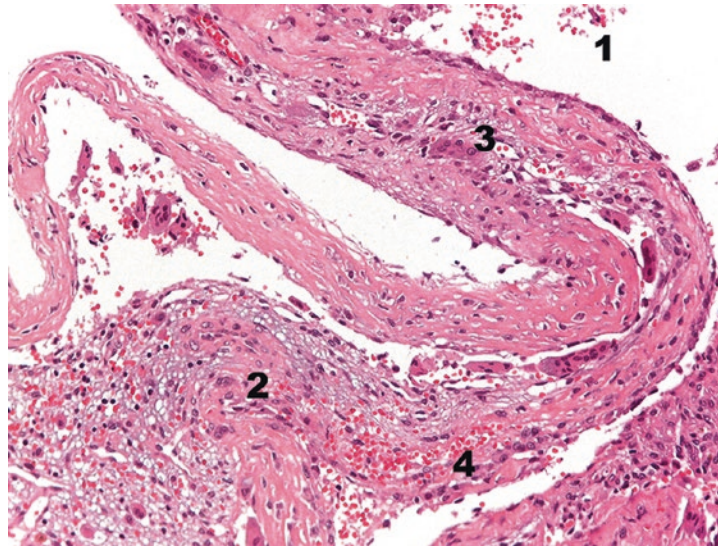
Radiograph, CT, and MR T2 axial image. The lesion is limited by a thin calcified periosteal reaction, indicating, despite a huge mass, slow growth. Fluid-fluid levels are detected on CT, but better seen on MR



Radiograph of ABC in a boy 16 years old, before and 9 months after curettage and bone grafts



CT scan of aggressive, stage 3, ABC in the first thoracic vertebra in a girl 15 years old. Two years after selective arterial embolization, the lesion was completed ossified and healed



The typical spongy structure is observed: multiloculated cystic spaces of varying size filled with blood are interspaced with solid areas of benign appearing fibrous tissue. (1) Blood-filled lacunae, (2) The walls of the cavities are formed by histio-fibroblastic tissue, (3) Multinucleated giant cells, (4) Thin blood capillaries

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