Aneurysmal bone cyst

Definition
A benign cystic lesion of bone that expands the cortex.
It is the only bone lesion that derives its name from its X-ray appearance rather than its histology.
Term first used by Jaffe and Lichtenstein in 1942[1].
ABCs have been divided into primary (arise de novo) and secondary (arise in pre-existing tumours, frequently GCTs). More than 50% of ABCs arise from preexisting lesions.

Incidence
2.5% of all primary bone tumours.
Half as frequent as giant cell tumours in the Mayo clinic data.
M=F (perhaps slight female preponderance).
76% in the first two decades of life (Mayo). (In contrast only 15% of patients with GCT are in the first two decades of life).

Aetiology
Trauma may be related.

Lichtenstein: intraosseous haemorrhage becoming an AV fistula or malformation.

Localisation
Some say most common around the knee, and the spine, but Dorfman's book states that they can be found throughout the entire skeleton with equal frequency, a distribution that is unique to this tumour.
Can involve any bone.
In long bones involves the metaphysis but can cross the growth plate.
In the spine involve the posterior elements (DDx here is osteoblastoma).
ABC is the commonest benign tumour of the clavicle.

Symptoms
Pain and swelling. Rapid increase in pain
Rarely, a pathologic fracture

Physical findings
May be a mass or neurological symptoms if the spinal cord is compressed.

Radiology
One way to think of the X-ray appearances of an ABC is to think of the lesion as an expansile, multilocular balloon disrupting the adjacent bone and elevating the periosteum.

Most commonly, an area of lucency situated eccentrically in the medullary cavity of a long bone.
Less commonly, may be situated centrally within the medulla. Much less commonly, may arise in the cortex or periosteum.
ABCs may cross joints and involve several bones, particularly in the spine where several adjacent vertebrae and ribs may be involved.
The lesion tends to involve the cortex and may destroy it completely, when it may bulge out into the soft tissue where it usually forms a thin rim of calcification.
Most ABCs are completely lytic.
The margins can be poorly or well defined; in half of cases the X-ray appearances suggest a benign process, and in a small number of cases they may suggest malignancy.
Periosteal new bone formation causing a buttress effect is characteristic.

[1] "The term aneurysmal relates to a sort of blowout distension of the contour of the bone and the term bone cyst relates to the fact that it represents mainly a blood filled cavity"

There is no matrix calcification.

CT and MRI may show internal septae and fluid levels with a layering effect. Fluid levels are best seen on T2 weighted scans.

Clyde Helms uses expansion and below the age of thirty as his discriminators.

Finger in balloon sign: preservation of a cortical cuff extending for a short distance into the expanded area of destructive blowout.

Gross pathology
Red brown granular material, often in the form of curettings.

The operating surgeon frequently encounters what appears to a hole containing blood (Dorfman). The blood in the lesion is not clotted, which some feel is evidence for an AVM, however there is no endothelial lining.

The pressure in an ABC may be elevated to arteriolar levels.

Histology
Essential feature is cavernomatous spaces, with walls that lack the normal features of blood vessels, such as muscle or elastic lamina.
Thin strands of bone are often present in the fibrous tissues of the walls.
The septae almost invariably contain giant cells; this helps to distinguish from SBC.
Solid areas contain spindle cells that are loosely arranged.
Chondroid like zones of calcification in solid portions of septae are commonly found and are relatively specific.

Behaviour
ABCs are usually aggressive lesions associated with major bone destruction, pathological fractures and local recurrence.
Spontaneous malignant transformation not recorded in the Mayo files but occurs very occasionally.

Differential diagnosis (histology)
1. Giant cell tumour
2. Giant cell reparative granuloma
3. Low-grade osteosarcoma
4. Telangiectatic osteosarcoma – probably most difficult DDx
    a. This disease is uncommon and rarely involves the vertebrae or small bones of the hands or feet
5. Renal cell carcinoma metastasis

Treatment
Intralesional curettage and bone grafting is the treatment of choice. In the Nov 2001 Mayo series a lower recurrence rate was noted with the use of phenol. Phenol is not used around nerve roots e.g. in sacral lesions.

Lesions in expendable bones are excised.

If the lesion has a large soft tissue component or is large (>5-6cm) should consider preoperative embolization.

Even incomplete resection may be followed by regression of the lesion.

There is a high recurrence rate. Recurrence tends to occur within 6-18 months. It is very rare after 2 years. The recurrence rate is higher in spinal tumours.

Radiotherapy has no role unless surgery is impossible. It used to be used adjuvantly but no longer is used to minimise the risk of sarcomatous transformation.

Prognosis
The underlying lesion determines the prognosis of secondary ABC.

May be an increased rate of recurrence in younger patients.