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# CYSTIC LESIONS OF BONE RESEMBLING GIANT CELL TUMORS

*By*

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Multinuclear cells are found in the stroma of a number of different types of cysts and cystlike processes within the skeleton. Because of the occurrence of these giant cells, tumor-forms embracing widely differing anatomical and clinical processes often were collected under the general term of giant cell tumors. During the past 20 years it has been possible by comparative histological, clinical and roentgenological studies, notably by American pathologists (*Jaffe, Lichtenstein*) to divide this previously heterogeneous group of giant cell tumors into distinct subgroups of tumors or tumorlike processes.

The following pathological conditions have been separated from the heterogeneous group of giant cell tumors:

true giant cell tumor;
solitary bone cyst;
aneurysmal bone cyst;
chondroblastoma;
non-ossifying fibroma;
(certain cystic changes in the jaw-bones;)

During recent years, an increasing number of bone tumors have come for treatment at the orthopaedic clinic of the Karolinska Institutet, and a closer collaboration has developed with Radiumhemmet and the institutions for tumor pathology and general pathology. A short description seems justified of some of the cyst-like changes which usually cause diagnostic difficulties. For more detailed descriptions the reader is referred to the monographs by *Dahlin* 1957 and *Jaffe* 1958.

*The true giant cell tumor* commonly occurs between 20 and 40 years

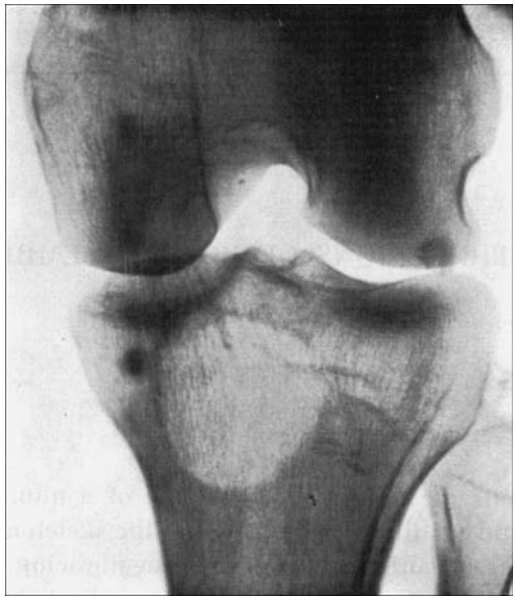

*Fig. 1.*
Roentgenogram of a giant cell tumor in the upper end of a tibia. Note the pure lytic lesion, eccentrically located. No periostal reaction. Female 55 of age.

of age. The sites of predilection are the inferior femoral and superior tibial epiphyses together with the inferior radial epiphysis. The clinical symptoms consist of the gradual onset of pain, swelling and eventually limitation of movement in the adjacent joint. X-ray shows a fairly large, well delineated osteolytic process excentrically placed in an epiphysis (see fig. 1.). One can often see that it has been a relatively fast progression. Periosteal reaction is lacking. Sclerosing new-bone formation occurs only in cases treated by radiation. The macroscopic appearance of the tumor varies. Sometimes one sees a greyishwhite, rather dense tumor tissue, at other times the colour is reddish or brown and the consistency soft.

Histologically there is a stroma of different forms of fibroblasts, a network of blood vessels, collagen fibrils and multinuclear cells. The histological picture often varies in different tumors and even in different parts of the same tumor, which can complicate the histological differential diagnosis.

From a prognostic and therapeutic point of view it is of the greatest importance in an individual case to be able to differentiate between the

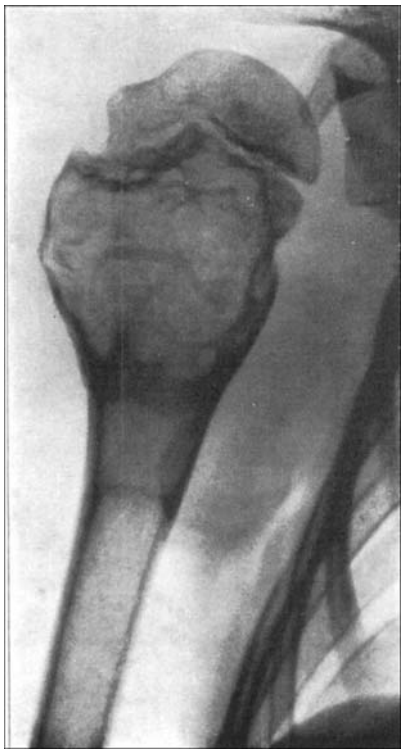

*Fig. 2.*
Roentgenogram of a solitary bone cyst.
Boy 7 years of age.

true giant cell tumor and the other cyst-like changes. A giant cell tumor, although benign at first, can in certain cases undergo a malignant transformation and metastasize to the lungs.

*The solitary bone-cyst* usually is discovered in childhood often by a fracture through the cyst in association with trauma. The site of predilection is the superior humeral diaphysis. Solitary bone-cysts occur even in the superior and inferior tibial and femoral diaphyses, and in the superior fibular diaphysis. There is a widening of the shaft and a thinning of the cortex. (See fig. 2.). A callus formation during healing of fractures through the original cyst can produce a multilocular appearance of the cyst on the X-ray film. At exploration the cysts are usually empty or contain only a little fibrinous slime or straw-coloured fluid. Fresh bleeding can appear, however, in association with fractures. The material obtained by curettage of the cyst-wall is usually of small amount and uncharacteristic appearance. If bleeding has occurred into the cyst an organized hematoma may be encountered. Histologically

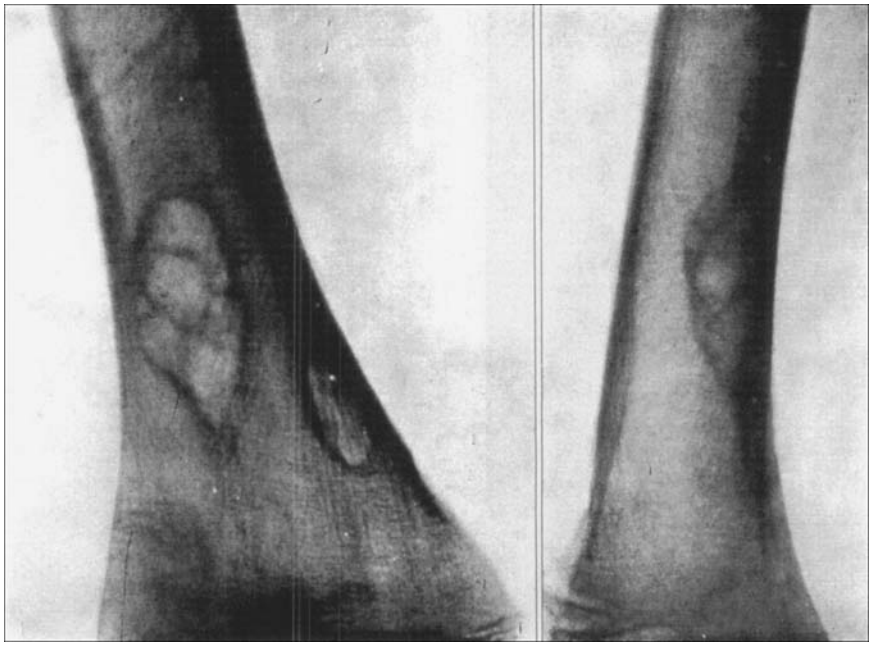

*Fig. 3.*
Roentgenograms of a non-ossifying fibroma in the femur.—Boy 7 years of age.

the cyst-walls consist of a very thin layer of fibroblasts, overlaid by fibrin and even blood pigment, phagocytes and sometimes multinuclear cells.

*The non-ossifying fibroma* usually occurs in adolescence up to the age of 20 years. The site of predilection is the long bones of the lower limbs with localization in the ends of the diaphysis. The clinical findings are unspecific. Local tenderness is often present. Usually the changes are discovered incidentally on X-ray examinations for other reasons. In some cases there seems to be a certain connection between trauma and the onset of symptoms. With enlargement of the lesion spontaneous fractures can occur.

Radiologically one observes a subcortically placed conglomeration of bubble-formed rarefaction which is surrounded by a sclerotic zone (see fig. 3). Small fibrous cortical bone-defects are common in children and are not considered as tumors. In certain cases these cortical bone defects can enlarge into the medullary cavity and thus assume the character of a non-ossifying fibroma. The fibroma can in addition grow towards the cortex which is thus thinned and pushed out into the surrounding

soft parts. Microscopically the fibroma is often brown or yellow-coloured with a rather dense consistency. The bone tissue is sclerotic around the fibroma. The histological picture has certain similarities to giant cell tumors. The stroma consists of spindle-shaped cells and often contains lipophages and multinuclear cells. The characteristic features are: 1) The patients are usually under 20 years of age; 2) The change is localized to the diaphysis not the epiphysis; 3) The condition is benign, there is a certain tendencey to spontaneous healing. Recurrence seldom occurs after a surgical curettage.

The following is a typical case: Case 1, B.K.: A boy of 7 years of age, treated elsewhere in 1954 for fracture through the lower part of the right femur-shaft, developed a cystic lesion in the fracture region. After curettage of the cyst and packing with autologous bone-chips the pathological report showed: Giant cell tumor.

The fracture healed. He returned to the hospital in his home province for regular half-yearly control. He began to have aching above the right knee at the age of 13. In October 1960 a new cyst-formation was observed in the right femur, somewhat distal to the previous pathological region. Uncertainty of the diagnosis, the highly varying opinions which the parents received, together with uncertainty regarding the plan of continued treatment led to his referral to the orthopaedic clinic.

*Preliminary assessment:* The age of the patient, the localization and the roentgen appearance indicated "non osteogenic fibroma". (Fig. 3).

Bloodcalcium normal.

Because of the present symptoms and the previous histological interpretation of giant cell tumor a second curettage was performed. The contents of the cystic lesion were sparse and of unspecific macroscopic appearance. The preliminary histological diagnosis was: Fibromatous giant cell tumor. Because of the clinical and roentgenological picture the histological interpretation was reviewed and the final diagnosis accordingly was revised to a "non-ossifying fibroma".

*Benign chondroblastoma.* This has previously been described as calcifying giant cell tumor or chondromatous giant cell tumor. It occurs in elder children and young adults, rarely over the age of 20 years. The usual sites are the femoral and tibial epiphyses around the knee-joint, and the upper humeral epiphysis. The symptoms are insidious, with slight pain on movement, swelling and gradually a reduced range of movement in the adjacent joint. X-ray shows a rounded osteolytic process which can reach a diameter up to 5–6 cm. Larger or smaller calcifications are generally found within the osteolytic region and this gives the lesion a mottled appearance (fig. 4). There is the same tendency to an eccentric position in the epiphysis as with the giant cell tumor but not so outstanding, and furthermore the latter is very rarely seen under 20 years of age. The differential diagnosis usually includes giant cell tumor or a malignant tumor such as chondrosarcoma or osteogenic

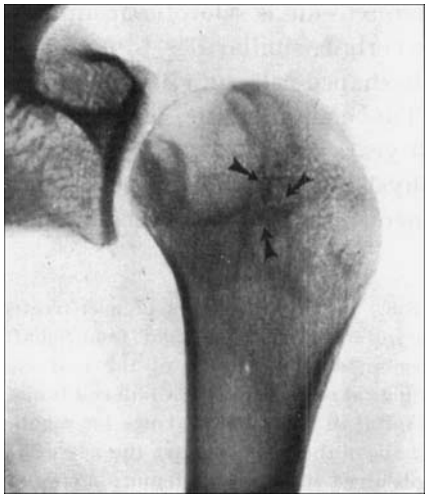

*Fig. 4.*
Roentgenograms of a chondroblastoma in the epiphysis of a humerus. Note small calcifications, indicated by arrows.

sarcoma, sometimes the lesion is taken for an infectious process. Macroscopically the tumor is bluish grey, yellow or brownish-red and the consistency moderately firm. The histological picture is characterized by chondroblasts, the appearance of calcification and numerous areas of necrosis. These tumors are often misinterpreted because of the appearance of multinuclear cells. The prognosis is, however, good and malignant change is unknown.

Case 2. L.S.: A girl of 15 years with pain and reduction of movement in the right humeral scapular joint, autumn 1961. The X-ray showed a localized cyst formation in the head of the humerus. The patient was treated in another hospital with short-wave. X-ray control Febr. 1962 showed progression. Needle-biopsy: Unspecific cells; uncertain origin. Radiation therapy was started.—The pain, however, increased. This led to the patient being referred to the orthopaedic clinic in March 1962.

Preoperative assessment: Benign chondroblastoma.—Biopsy and curettage. The large cavity in the head of the humerus contained a bluish-grey firm tissue. Microscopic pathology: Benign chondroblastoma (Codman-tumor). After this operation healing was uneventful. The prognosis was judged as favourable. The mobility of the humeroscapular joint improved.

*Aneurysmal bone-cyst:* This peculiar skeletal change occurs in children and adolescents, seldom in patients over 20 years of age. The commonest localizations are the ends of the diaphyses of the long bones and vertebral column. The symptoms depend on the site. In the back stiffness, swelling, eventually root-pain may occur. In the long bones there will be local swelling, tenderness, minor ache and limitation of move-

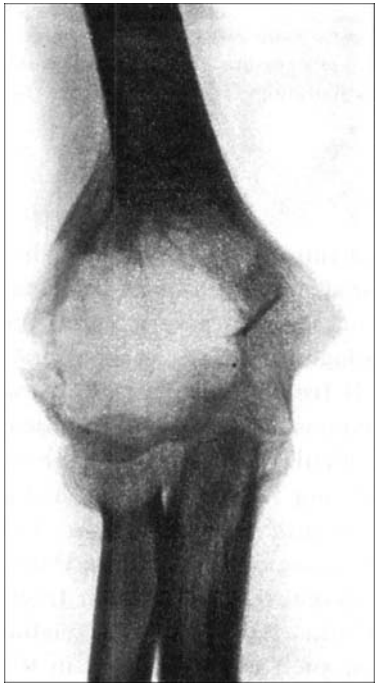

*Fig. 5.*
Roentgenogram of an aneurysmal bone cyst in a humerus. Note the ballooned out area, laterally. Girl 17 years of age.

ment in the adjacent joint. In most cases the roentgenographic findings are characteristic. The cyst-formation grows eccentrically and is surrounded by an eggshell-like periosteal calcification. The similarity to an aneurysm has given rise to the name. The above mentioned eccentric growth serves to differentiate the aneurysmal bone cyst from a solitary bone cyst which grows concentrically. The differential diagnosis is not difficult in the typical cases, with the eccentric protrusion of the cyst formation and the periosteal calcification.

A typical case is described below:

Case 3. P.B.: Previously healthy girl of 17 years of age. 1959 minor contusion of the right elbow region followed by local tenderness and limitation of movement. X-ray examination: Walnutseized cystic rarefaction laterally in the distal part of the humerus.—Biopsy. Sparse contents. Pathological diagnosis: "A picture resembling a giant cell tumor. Probably congenital bone cyst".—Immobilization in plaster one month. A following roentgenogram showed progression. Operation with curettage revealed a cyst with profuse bleeding.—Pathology report: Aneurysmal bone cyst.

Due to the differing opinions of the histological diagnosis, the patient was referred to the Orthopaedic clinic 7/1 1960. Roentgen (Fig. 5) showed a large ballooned out cystic lesion of the lower end of the humerus.—Operation: *Curettage of bone cyst right humerus + packing with autogenous cancellous bone-chips from the iliac crest.*

The walls of the cyst were paper thin. Perforation into the soft tissues was present in several places. There was found blood under slight pressure within the cyst and brownish-red soft slime covering the walls. The pathology report confirmed the diagnosis of an aneurysmal bone cyst.

## DISCUSSION

From a practical view-point it is of great importance that the orthopaedist and the roentgenologist who care for skeletal tumors and skeletal diseases also have a knowledge of the pathologic characteristics of the conditions. It is also clear that the histological picture alone is not always sufficient for the exact diagnosis. It frequently occurs that the pathologist, in order to reach a definite diagnosis, is in need of clinical and roentgenological information in the particular case. Certain of these cyst-like lesions, for example the non-ossifying fibroma, have a histological picture which closely resembles the genuin giant cell tumor. The combined clinical, roentgenological and histological analysis is therefore of greatest value in determining the diagnosis, deciding on treatment and assessing the prognosis. The cyst-like changes in the skeleton which have been described are examples of such lesions, which in the older textbooks were gathered together in a large heterogenous group, the so-called giant cell tumors, but where modern diagnosis allows more specific differentiation. These conditions are not in general sufficiently well recognized and because skeletal tumors are relatively seldom seen, a certain centralization is desirable to institutions where the time, interest and combined resources are available.

## SUMMARY

For the diagnosis of cystic lesions of bone resembling giant cell tumors a clinical, roentgenological and pathological cooperation is emphasized.—A short description is given of the true giant cell tumor, the solitary bone cyst, the aneurysmal bone cyst, the benign chondroblastoma and the non-ossifying fibroma, which all may be confused with giant cell tumor.

## RESUME

Il est recommandé une collaboration clinique, radiologique et pathologique pour établir le diagnostic de lésions kystiques osseuses ressemblant à des tumeurs de cellules géantes. – Une courte description est donnée de la véritable tumeur de cellules géantes, du kyste osseux soli-

taire, du kyste osseux aneurysmal, du chondroblastome bénin et du fibrome non ossifié qui peuvent tous être confondus avec les tumeurs de cellules géantes.

ZUSAMMENFASSUNG

Zur Diagnose von cystischen Lesionen des Knochens, die Riesenzellengeschwülsten ähneln, wird die Notwendigkeit der klinischen, röntgenologischen und pathologischen Zusammenarbeit hervorgehoben. – Eine kurze Beschreibung der echten Riesenzellengeschwulst, der solitären Knochencyste, der Aneurysma-Knochencyste, des gutartigen Chondroblastoms und des nicht knochenbildenden Fibroms, die alle mit Riesenzellengeschwülsten verwechselt werden können, wird gegeben.

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