

Aneurysmal bone cyst

A review of 52 primary and 16 secondary cases

M. Szendrői, I. Cser, A. Kónya, and A. Rényi-Vámos

Orthopedic Department of the Semmelweis Medical School, Budapest, Hungary

Summary. Authors report on the results of treatment of 52 primary and 16 secondary aneurysmal bone cysts (ABC). ABC grow rapidly; 84% of them have already destroyed more than the half of the bone width at recognition. En bloc resection is preferred when the ABC is growing superficially and eccentrically and more than half of the bone width is intact. Careful curettage and bone grafting still remains the surgical method of choice in the majority of cases, when the ABC is more destructive and affects the subchondral bone of the joints. Segmental resection is only indicated when removal of the affected bone does not influence the function of the extremity. Superselective embolization of the cyst was performed in seven cases with excellent results. This method is suggested for ABC in certain locations inaccessible to surgical intervention, e.g., the pelvis, or to avoid excessive bleeding in hypervascularized tumors. In one case, however, an incomplete rebuilding of the ABC could only be achieved by the administration of calcitonin. The 16 cases of secondary ABC were observed mostly in association with osteoblastomas, giant-cell tumors, and osteosarcomas. The incidence of the secondary ABC was 23% in the whole ABC group but not more than 2–4% among the osteosarcomas and giant-cell tumors. Secondary ABC may confuse the histological and clinical diagnoses and that, especially in cases of osteosarcoma, may have fatal consequences.

The name “aneurysmal bone cyst” (ABC) was first used by Jaffe and Lichtenstein [7], separating this tumorlike lesion of the bone on the basis of its distinct clinicopathologic appearance from other bone cysts and from the giant-cell tumors. ABC is usually an eccentric lytic lesion blowing up the bone. It consists histologically of blood-filled spaces surrounded by connective tissue septa containing osteoclast giant cells and newly formed bone trabeculae [6, 10].

The ABC is rather rare. Its incidence is estimated at 6% of all bone tumors; it is four times more rare than osteosarcoma [19]. In consequence, only a few articles have been published to date reporting on the results of the treatment of more than 50 patients [1, 2, 9, 13, 18, 22].

By assessing the ABC cases collected from our Bone Tumor Registry, our aim was to summarize the clinico-

pathologic characteristics, to review the differential diagnostic problems, and above all to discuss the optimal possibilities for treatment in the light of our therapeutic results, and of the recurrences and complications.

Materials and methods

In the Bone Tumor Registry of the Semmelweis Medical School in Budapest 52 cases of primary and 16 cases of secondary ABC were recorded between 1970 and 1990. The data collected for each case included sex and age of the patient, location and size of the ABC, duration of symptoms, and the clinical laboratory data relating to the bone metabolism (serum calcium, phosphorus and alkaline phosphatase values). Radiographs were evaluated for whether, when the cyst was discovered, it measured more than 50% of the bone's width, had broken through the physis, reached the subchondral bone, and whether it had increased centrally or eccentrically.

The surgical interventions were classified into three types following Enneking [5]: (1) intralesional (curettage and bone grafting), (2) marginal (en bloc resection), and (3) wide resection (segmental resection). Forty-five patients with primary ABC could be followed up and their present condition evaluated. The follow-up time was on the average 7.5 years.

In seven patients the supplying arteries of the ABC were superselectively embolized. For the catheterization French 2–5 catheters were used, through which 0.8–1.5 ml Histoacryl-Lipiodol 1:1 mixture was injected into the vessels. In one case the embolization treatment was complemented by intracystic and intravenous calcitonin hormone injections (Myacalcic, Sandoz, 50 UI/ml).

Reviewing other bone tumors and tumorlike lesions in the Registry, 16 cases were found in which the typical histological and X-ray appearance of an ABC was associated with other bone tumors and tumorlike conditions.

Results

There was a distinctly higher incidence of ABC in male patients (31 male to 21 female). Their age in decades is shown in Fig. 1. Twenty-nine percent of the ABC occurred in the first and 48% in the second decade of life. Symptoms had existed on the average for 7 months before admission; pathological fracture was reported in seven cases.

The distribution of the ABC over the skeleton is shown in Fig. 2. The most frequently affected bones are the humerus, tibia, pelvic bones, and femur. The cysts in the short tubular bones reached an average size of $3 \times 2 \times 2.5$ cm, growing with one exception centrally and

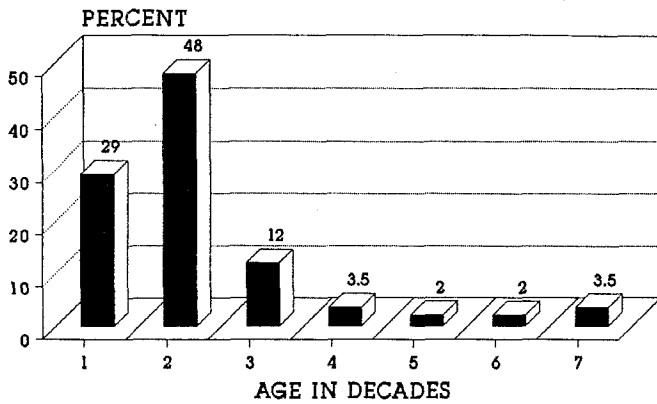


Fig. 1. Age distribution of patients with aneurysmal bone cysts (ABC; $n = 52$)

bulging out the bone (Fig. 3). By contrast, ABC in the long tubular bones and pelvis were on average $6 \times 4 \times 3.3$ cm in size, growing mainly eccentrically, expanding the bone (Fig. 4). Not rarely, the cysts increased centrally but did not cause the bone to bulge, so it did not show the characteristic X-ray picture of an ABC.

Sixty percent of the ABC were purely metaphyseal, 28% metaepiphyseal. In young patients they did not break through the physis: here we observed ABC without exception metaphyseally. In eight cases the ABC reached the subchondral bone, and in two of them it broke into the joint.

In 44 cases (85%) the size of the ABC when first seen was already more than 50% of the bone width.

The laboratory data relating to bone metabolism showed an augmentation of alkaline phosphatase in some cases only.

Four out of nine ABC investigated by angiography proved to be hypervascular. In two with mildly pathological structure of the vessels, malignancy of the process was assumed radiologically.

Table 1 shows the surgical interventions performed, and the complications and recurrences. Surgical intervention proved to be necessary in 48 of the primary ABC. The overall incidence of recurrence was 13%. In 26 cases (54%) curettage and bone grafting were performed. In 7 cases (27%) recurrences occurred, after an average of 14 months. In 3 patients the curettage was followed by irradiation (18 Gy); 11, 15, and 19 years respectively after the radiotherapy we have observed no malignant transformation of the ABC. Recurrences were treated either by curettage or resection followed by arthrodesis (2 cases).

En bloc resection, i.e., marginal resection of the ABC was performed in six ABC, mostly superficially located and growing eccentrically. No recurrence occurred (Fig. 5).

Segmental resection, according to Enneking's classification "wide resection" [5] was performed in 16 cases (34%). In the majority, substitution of the removed bone segment was not necessary (fibula, radius, ulna, etc.). In some cases, however, the wholly destroyed metatarsal metadiaphysis had to be replaced by a piece of iliac bone. Two humeral heads were replaced by auto-

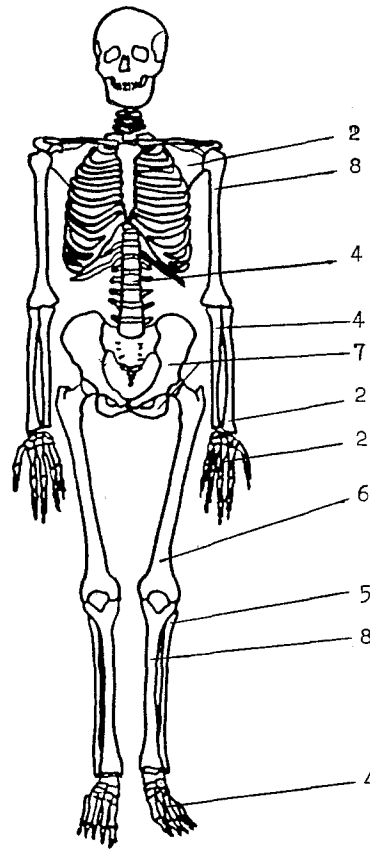


Fig. 2. Distribution of 52 ABC over the skeleton

logous fibula, and one destroyed and fractured femoral head by tumor prosthesis (Fig. 6). The recurrence rate after segmental resection was zero in our patients.

Superselective embolization of the ABC was performed in seven cases, in some of them as a preventive measure to avoid the heavy bleeding that occurred at the biopsy. In four patients the location of the ABC (pelvic bones) made surgical treatment very difficult, and for this reason embolization of the cysts was preferred. In all cases but one the cysts showed no further growth and progressive ossification started after 2–3 months, being finished 10–18 months later. Almost perfect bony rebuilding was seen in the ABC of the pubis (Fig. 7) treated by embolization of the obturator arteries, while ABC affecting the ischial and acetabular bones have shown only incomplete rebuilding. There were no complications except transitional ischaemic muscle pains.

In one patient we have seen progression even after embolization treatment. Follow-up angiography did not show any revascularization, so calcitonin was administered intravenously and by direct puncture of the cyst. After 3.5 months bony rebuilding, in some places incomplete, could be detected.

To turn now to the 16 secondary ABC found retrospectively in our Bone Tumor Registry records, in 4 out of the 124 recorded osteosarcomas regular ABC wall details were seen in addition to the classical histological picture of the tumor (2 telangiectatic, 2 osteoblastic subtypes). In one case, the histological diagnosis from the biopsy was erroneously ABC. Besides these, 4 out of 18 osteoblastomas (Fig. 8), 2 out of 21 chondroblastomas, 2



Fig. 3. Eccentrically growing ABC causing the posterior part of the tibia to bulge. Lateral view

Fig. 4. Centrally growing ABC, “ballooned-out” metacarpal bone. Anteroposterior

Fig. 5. ABC in the distal femur, localized eccentrically and superficially. Optimal conditions for en bloc resection

Fig. 6. a ABC blowing up and destroying the left femoral neck leading to pathological fracture. Anteroposterior. **b** Wide segmental resection followed by prosthesis implantation. Anteroposterior view

Table 1. Surgical interventions, complications, and recurrences

Surgical intervention	Number of cases	Complications	Recurrences
Curettage + spongiosa grafting	26 (54%)	Suppuration (2)	7 (27%)
En bloc resection	6 (13%)	0	0
Segment resection	16 (33%)	Suppuration (2), scoliosis (1), transient peroneal nerve palsy (1)	0

out of 89 fibrous dysplasias, 1 out of 113 nonossifying fibromas, and 3 out of 82 giant-cell tumors were found in which the basic disease was associated with ABC. The patient age and the localization of the lesion corresponded well with those of the basic disease, but the duration of the symptoms and radiographic appearance were remarkably modified by the secondary ABC.

The present status of 45 patients who could be examined after an average follow-up of 7.5 years can be summarized as follows. In 14 out of 21 patients the cysts healed without complications after curettage and bone grafting, 7 patients had recurrences, and 2 patients had to undergo resection followed by arthrodesis of the ankle and knee for recurrent ABC. After the knee-arthrodesis suppuration and fistula formation occurred as complication.

Every patient with en bloc resection of the ABC was free of complaints and full movement of the joint had been retained.

Among the 14 patients with segmental resection included in the follow-up study, only 9 were free of complaints. We observed scoliosis after partial resection of a vertebra, reduced pronation and supination after partial resection of the ulna and radius, and increased movement of the shoulder after resection of the humeral head and fibular autotransposition.

Three patients were free of complaints after embolization of the ABC; one of them has moderately increased movement in the hip joint.

Discussion

The sex distribution of our 52 primary ABC patients differs somewhat from that reported in the literature [19], as among our patients males were in the majority (3:2). The age distribution agrees well with that reported elsewhere: 77% of the patients were under 20 and 29% under 10 years old.

The clinical data usually show rapid growth of the cyst, which can, especially in the case of hypervascularized cysts, be misinterpreted as a sign of malignancy. The duration of the symptoms was 7 months on average, twice as long as for cases of osteosarcoma [21]. During that time the ABC can reach considerable size and can involve more than half of the width of the bone. Pathological fractures are frequently the first sign of the ABC. Pain and swelling are less expressive than in malignant bone tumors, while laboratory values are normal with exception of the moderately elevation of the alkaline phosphatase levels, which varies from case to case.

ABC may affect almost any bone, but there is a predilection for the long tubular bones. It usually arises in the metaphysis and then grows into the epi- or diaphysis. In our patients no ABC destroyed the growing plate.

On the basis of the typical clinical and radiological appearance, we succeeded in making the right diagnosis in 80% of cases before biopsy. The remaining 20% show well the differential diagnostic problems of ABC. If an ABC is centrally localized and does not expand the bone, it is difficult to tell it from a juvenile bone cyst, eosinophilic granuloma, osteoblastoma, etc. “Ballooned-out”

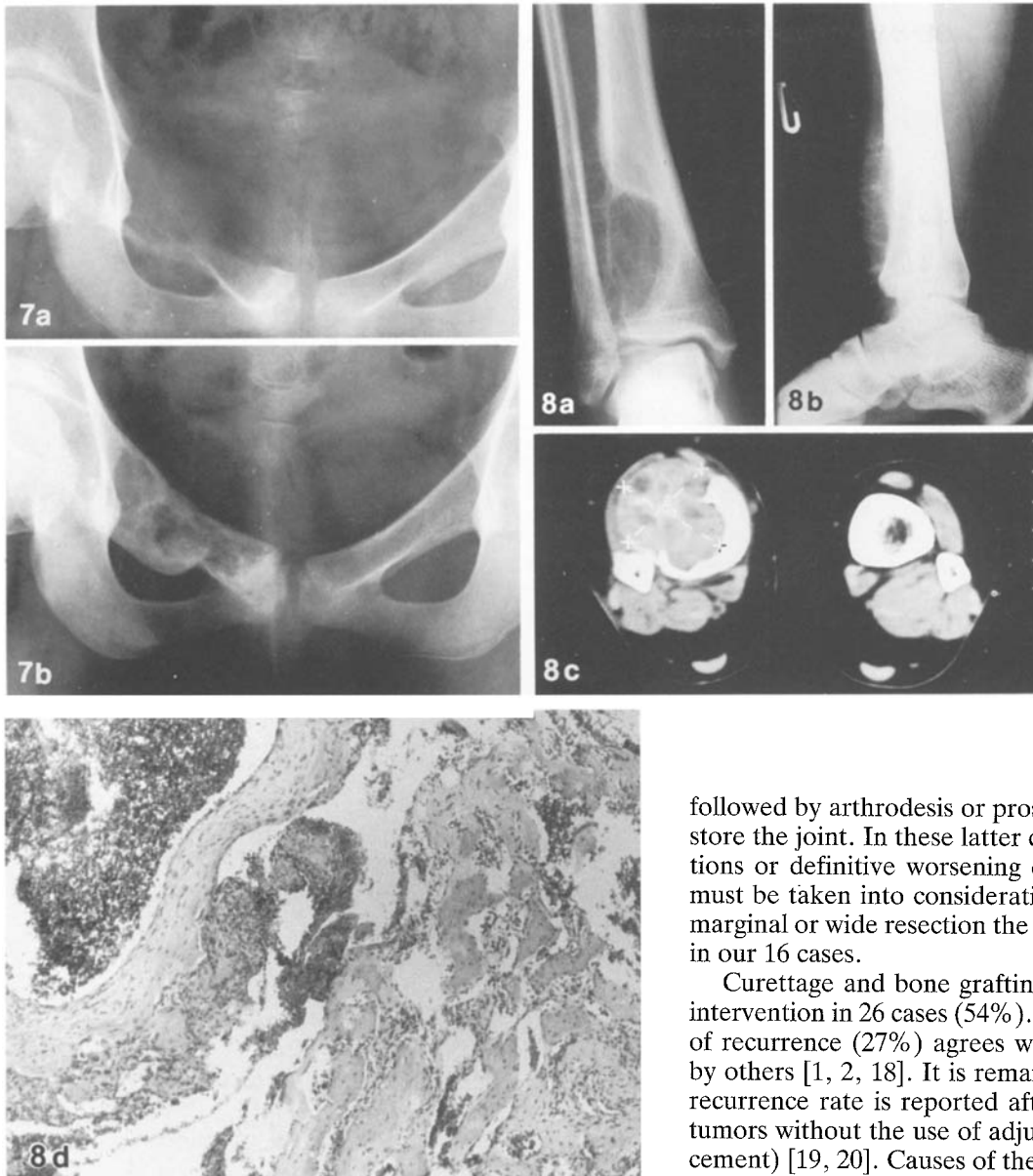


Fig. 7. **a** ABC in the right superior pubic ramus. Anteroposterior. **b** 8 months after selective embolization, ossification and rebuilding of the cysts is visible. Anteroposterior view

Fig. 8a-d. Eccentric lytic and cystic lesion imposing an ABC in the distal tibia. **a, b** Anteroposterior and lateral radiographic views. **c** CT scan shows the cyst containing tissue of varying density and small cavities. **d** Histology proved an osteoblastoma with associated secondary ABC. HA & E, $\times 160$

followed by arthrodesis or prosthesis implantation to restore the joint. In these latter cases, however, complications or definitive worsening of the patients' condition must be taken into consideration. In cases of successful marginal or wide resection the recurrence rate is zero, as in our 16 cases.

Curettage and bone grafting were performed as first intervention in 26 cases (54%). The rather high incidence of recurrence (27%) agrees well with figures published by others [1, 2, 18]. It is remarkable that the same high recurrence rate is reported after curettage of giant cell tumors without the use of adjuvants (e.g., phenol, bone cement) [19, 20]. Causes of the frequent recurrence may be the small window on the cortex made by the surgeon or involvement of the subchondral bone: both make careful and radical curettage impossible.

To avoid recurrence of the ABC is extremely important, because if a recurrence destroys the articular surface, only resection combined with arthrodesis or prosthesis implantation is of any use. Therefore, the curettage should be complemented with phenol treatment, i.e., with an adjuvant without complications and accepted more and more in the treatment of giant cell tumors.

In our experience, curettage and bone grafting is still the recommended treatment to save the limb and function of the joints in cases of ABC growing centrally or eccentrically destroying the bone near to the joint.

Like some other authors [4, 15, 17], we have had very good experience with superselective embolization of ABC [8]. Embolization is indicated, we think, (1) to avoid excessive bleeding in hypervascularized cysts, (2) for cysts in certain anatomical locations (pelvis, sacrum, vertebra) which are difficult to treat surgically, and (3) when the subchondral bone is destroyed, in or-

eccentric lytic lesion may be produced by a giant-cell tumor or, in rare cases, by atypical myeloma (Fig. 9).

In planning the surgical intervention, the localization and growth pattern of the ABC are decisive. It is most advantageous if the ABC grows superficially and involves no more than one-third of the bone width. Such cases are best suited for *en bloc resection* (marginal resection), removing the cyst in normal bony tissue. This is a radical intervention for ABC and results recurrence-free healing with normal joint function. Unfortunately, only a small minority of our cases (6 out of 52) were suitable for *en bloc resection*.

Segmental resection is recommended for ABC located in bones where this treatment does not interfere with function (fibula, ulna, vertebral processes, etc.). It can also be performed where the ABC has destroyed the short tubular bones of the hand or foot, followed by reconstruction with pieces of iliac bone. Rarely, when the joint is involved, as a last solution resection is necessary,



Fig. 9. "Ballooned-out" lytic lesion of the clavicle mimicking ABC. Histology showed a myeloma. Anteroposterior view

der to save articular function. The results of embolization in our seven cases show that pain was reduced in every case if the cysts were excluded from the arterial circulation, and revascularization did not develop even months later as controlled by angiography. Detectable ossification occurred after 1–3 months, and definitive bony rebuilding happened 1–1.5 years after the onset of the treatment. The bony rebuilding was most complete after embolization of the obturator artery in cases of pubic ABC, while ischial cysts affecting the medial wall of the acetabulum showed incomplete bony rebuilding only.

Reports are often published of nonsurgical treatment of ABC [12, 16]. In three of our patients radiotherapy was given and no malignant transformation was observed after several years, however, irradiation is not recommended for ABC because of the high incidence of postirradiation sarcoma reported earlier [11].

Dubousset (personal communication) proposed calcitonin hormone treatment of ABC. The basic idea is that calcitonin suppresses the osteoclast activity and promotes the new bone formation in the connective tissue septa of the cyst wall. The hormone administration is combined with filling up the cyst with biopolymers. In one case where progression of the growth of the ABC was observed after successful embolization, progressive ossification and partial rebuilding were achieved by 3 months administration of calcitonin.

The proportion of secondary cysts among the ABC is relatively high [3, 13]: it was almost one-quarter of our patient group (23%). The incidence of secondary ABC among the bone tumors and tumorlike lesions is, however, insignificant: approximately 3% of the 124 osteosarcomas, 3.5% of the 82 giant cell tumors and 89 fibrous dysplasias showed areas of a secondary ABC. ABC were relatively often observed in association with osteoblastomas in our material (4 out of 18 cases), but considering the small number of cases, conclusions could not be made.

The significance of secondary ABC is: (1) the fact that a secondary ABC may develop in a number of bone tumors and tumorlike lesions with very different etiology confirms the hypothesis that an ABC is a secondary change in a preexisting lesion due perhaps to local hemo-

dynamic disturbances; (2) if the biopsy material contains a small amount of tissue representative of only the basic lesion and the secondary ABC rules the histological type, an erroneous diagnosis which may be fatal for the patient may be obtained.

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