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Modern concepts of primary aneurysmal bone cyst

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Abstract
Introduction Despite the long experience of radiologists, pathologists and orthopaedists with aneurysmal bone cysts (ABC), there is limited knowledge regarding the cause of the lesion and the optimal treatment. The pathogenesis of ABC remains unclear with theories ranging from a post-traumatic, reactive vascular malformation to genetically predisposed bone tumours. Recent genetic and immunohistochemical studies proposed that primary ABC is a tumour and not a reactive tumour-simulating lesion. The chromosomal analyses and some reported familial cases of this osteolytic bone lesion propose a hereditary factor in a presumably multifactorial pathogenesis.
Materials and methods The imaging studies, even CT scan and MRI sometimes do not provide clearly diagnostic criteria for the diagnosis of ABC. The radiographically differential diagnosis between ABC and unicameral bone cyst (UBC) is sometimes not clear. Double density fluid level, septation, low signal on T1 images and high intensity on T2 images strongly suggest the bone cyst is an ABC, rather than a UBC.
Conclusion Common methods of treatment vary considerably in the literature. The usual methods of treatment are curettage, resection, intracystic injections and embolization. Biopsy is imperative before any treatment. Ethibloc® treatment remains highly controversial. For some authors Ethibloc® injection can be recommended as the first-choice treatment excluding spinal lesions. A minimally invasive method by introduction of demineralized bone and autogenous bone marrow is able to promote the self-healing of a primary ABC.

Keywords Aneurysmal bone cyst · Bone tumour · Treatment

The experiments comply with the current laws of the country in which they were performed.

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Introduction

Aneurysmal bone cyst (ABC) is a cystic and expanding vascular bone lesion that remains a diagnostic and therapeutic challenge [68]. ABC can exist either as a primary bone lesion (in about 70% of the cases) when it arises de novo (no precursor bone lesion is identified), or as a secondary lesion when a pre-existing osseous lesion can be identified (in about 30% of the cases) [22]. This tumour occurs most frequently in patients under 20 years of age and represents approximately 1% of all bone tumours [22]. Primary ABC is a rare lesion with an incidence of 0.14 per 10^5 individuals [45].

A growing number of studies have described the clinical, radiological and pathological features of ABC. However, many problems remain unanswered. What is its pathogenesis, is it possible to only use radiographs and MRI for definitive diagnosis and, how is it best treated?

The pathogenesis of ABC remains unclear with theories ranging from a post-traumatic [24, 37, 66],

reactive vascular malformation [46, 47] to a genetically predisposed bone tumours [2, 44, 46, 58, 62, 63, 78, 79]. The most widely accepted pathogenetic mechanism of ABC's involves a local circulatory disturbance leading to markedly increased venous pressure and the development of a dilated and enlarged vascular bed within the affected bone area [5, 10]. However, the recent identification of recurrent chromosome abnormalities has challenged this historical perception [2, 4, 29, 37, 39, 58, 60, 62, 79].

The lesion usually has the radiographic appearance of a well-defined central radiolucency in the metaphyseal region of long bones [72]. The radiographically differential diagnosis between ABC and UBC is sometimes not clear.

The usual methods of treatment are curettage [5, 9, 11, 34, 43, 57, 67, 71, 73, 75, 77], resection [5, 9, 67, 73, 77], intracystic injections [1, 27, 31, 33, 35, 38, 41, 61, 64, 74, 76] and embolization [13, 16, 26, 28, 30, 40, 54, 56]. Opinion varies considerably, as to which method should be used, particularly in children.

In three previous reports [18, 20, 21], we performed a multi-hospital retrospective study of 156 primary ABC's in children and adolescents. In the first one, we reviewed the demographic data [18]. A literature review was included regarding gender, location of the lesion and, age at diagnosis. In this demographic data on 411 children with primary ABC's, the femur (22%), tibia (17%), spine (15%), humerus (10%), pelvis (9%) and fibula (9%) were the most common locations [18]. The second study concerned 15 pelvic ABC's with a review of the literature. We found that spontaneous healing in a few cases (even in active or aggressive lesions) argued for clinical and radiologic observation after biopsy when possible instead of a primary surgery [20]. In the third study, we found that ABC in children, 5 years of age or younger, did not seem more aggressive than in elder children [21].

Despite a descriptive history of more than 60 years, the nature, character and optimal treatment of ABC remain obscure [51]. During these last years, the knowledge about ABC strongly evolved, especially about pathogenesis, modern imaging and treatments.

In the light of our experience [6, 17, 18–23] and a review of the literature, we illustrate the new concepts about pathogenesis, imaging features and treatment of these lesions.

Pathogenesis

Several hypotheses have been formulated for the pathogenesis of this bony lesion, but the cause of primary ABC remains unclear. Histologically, this lesion has no similarity to either a cyst or an aneurysm [7, 37].

Lichtenstein [47, 48] proposed that ABC be related to a circulatory disturbance causing increased venous pressure due to thrombosis of a sizeable vein, or formation of arteriovenous fistulas [7, 47]. The evidence supporting the hemodynamic hypothesis includes: (a) the often marked and occasionally rapid extension of some of these cysts; (b) the common operative finding of a blood filled cyst, the blood often welling up, occasionally to the extent of giving rise to some concern and (c) angiography may sometimes show changes suggesting a vascular lesion and the presence of an arteriovenous shunt [14].

Tillman [75] believes that the lesion probably arises de novo.

Biesecker et al. [5] supported the theory that ABC may originate in pre-existing lesions, e.g. benign bone tumours. Biesecker et al. [5] performed a manometric study of six cysts. Cystic pressures were elevated in three of six cases measured. The elevated readings were equal to normal vascular pressures at arteriolar (58 mm Hg), capillary (31 mm Hg) and venous (7.4 mm Hg) levels. The pressure in one recurrent cyst was identical to the pressure in the primary occurrence. Three other cases, which were measured, had essentially no elevation in pressure. Three cases, which were not measured manometrically, were thought to have elevated pressures because of a "spurt of blood" at initial needle aspiration. For Biesecker et al. [5], a primary lesion of bone initiates an osseous, arteriovenous malformation and thereby creates, via its hemodynamic forces, a secondary reactive lesion of bone, which we know as an ABC. For Biesecker et al. [5], the incidence of associated lesions in ABC is too high for chance alone, and the nonneoplastic, reactive pathology of ABC mitigates against their being but a separate manifestation of the associated lesion. This hypothesis supposes that ABC, by it own extension could destroy in two-third of the cases the pre-existing tumoural lesion, looking like a primary lesion. This theory remains controversial.

Essadki et al. [32] hypothesised that ABC is due to the opposed direction of periosteal and medullary blood circulation. This could explain the relative rarity of diaphyseal localisations compared with metaphyseal localisations [32].

For some authors, ABC can be caused by trauma [24, 37, 66]. Trauma may be the triggers that lead to the development of the ABC [66]. The development of an ABC after a fracture in a previously normal bone supports the view that the lesion may occur secondary to a local abnormality within the bone [66]. It is likely that the initial stimulus for the formation of the ABC is an

alteration of the intraosseous blood flow, as a result of a fracture, and subsequent hyperaemia during the healing phase [66]. The exact mechanism by which the normal changes in bone vascularity caused by a fracture are converted into a pathological process resulting in an ABC is unknown, and why ABC so rarely arises after a fracture is a mystery [66].

For Campanacci et al. [10], ABC is not a neoplasm. Its histological features and course rather indicate a reactive and repair tissue consequent to a particular type of haemorrhage [10]. The "primum movens" of this haemorrhage remains obscure. Hematic cavity images similar to those of ABC may be observed in some areas of a giant cell tumour, of a brown tumour in hyperparathyroidism, of a fibrous dysplasia, of a chondroblastoma, of an osteoblastoma, of a hemangioendothelioma [10]. This does not mean that ABC constitutes the haemorrhagic transformation of some other pre-existing tumoural lesion [10]. In Campanacci's opinion, in genuine ABC there should be no traces of any other lesion; if this were so, it would not be an ABC, but rather another lesion with occasional cystic-haemorrhagic features [10]. Furthermore, the anatomo-clinical features of ABC are such that it is an entity in itself [10]. All that may be said based on the facts at hand is that ABC probably begins due to some changes in circulation and local haemorrhage [10]. This produces a repair and osteolytic tissue, which in turn nourishes the haemorrhage. Thus, a vicious circle is produced, explaining the continuous, and at times monstrous, expansion of ABC [10]. In some cases, instead, the phenomenon tends to slow down and become exhausted, and the ABC "matures", stabilises and even spontaneously repairs [10]. The facts, which agree with this hypothesis, are as follows. The cavities of the ABC are not vessels. They do not communicate with either an arteriole or a venula, rarely with minute capillaries [10]. The cavities originate as "small lakes" of red blood cells in the specific tissue of the ABC and progressively dilate [10]. It seems that the red blood cells ooze from the thin capillaries of the tissue itself, where perhaps there exists an open circulation.

Furthermore, some phenomena have been reported that are not easily explainable using the theory of ABC as a reactive bone lesion [46]. First, five familial cases of ABC have been reported, including monozygotic twins [63], brothers [39], brother and sister [78], father and son [44], affecting both the same location at nearly the same age, and father and daughter [29]. Second, three congenital cases, two in human new-borns and one in a foal underline the possibility of a hereditary defect [37, 46]. In none of the cases has a trauma during pregnancy been noted [46]. Third, some proven hereditary diseases like Goltz syndrome, or Van Buchem disease have been reported to be sometimes combined with ABC.

Recent genetic and immunohistochemical studies proposed that primary ABC is a tumour and not a reactive tumour-simulating lesion [46]. For Leithner [46], a predisposing genetic defect could be part of a multifactorial pathogenesis in the development of some ABC's [46].

Cytogenetic findings suggest that at least some ABC's are true neoplasms [46]. Nearly all cases showed an involvement of 17p11–13 or 16q22 [2, 4, 60, 79]. Surprisingly, these rearrangements seem to be always present, irrespective of subtype (classic, solid, primary or secondary) and location (osseous or extraosseous). Recently, a neoplastic basis in primary ABC was evidenced by demonstration of clonal chromosome band 17p13 translocations that place the USP6 oncogene under the regulatory influence of the highly active CDH11 promoter [58]. Oliveira et al. [58] reported CDH11 and/or USP6 rearrangements in 36 of 52 primary ABC's. USP6 and CDH11 rearrangements were restricted to spindle cells in the ABC and were not found in multinucleated giant cells, inflammatory cells, endothelial cells, or osteoblasts. CDH11 and USP6 rearrangements were not found in any of 17 secondary ABC associated with giant cell tumour, chondroblastoma, osteoblastoma and fibrous dysplasia. These findings demonstrate that primary ABC is mesenchymal neoplasm exhibiting USP6 and/or CDH11 oncogenic rearrangements [58]. By contrast, secondary ABC lack CDH11 and USP6 rearrangements, and although morphological mimics of primary ABC, appear to represent a non-specific morphological pattern of a diverse group of non-ABC neoplasms [58].

Imaging

The early radiographic appearance of the ABC in the long bones is a subperiosteal, metaphyseal eccentric lesion, elevating and inflating the periosteum and progressively eroding the cortex [16, 22]. Concentric involvement is often seen in ABC of the fibula, radius and humerus [16] (Fig. 1). The osteolytic area often contains septa and ridges, producing a honeycomb pattern. Sometimes the lesion shows calcification in the central area [51]. A thin shell of cortical bone is often seen peripherally. In some cases, the lesions are separated from adjacent soft tissues by only a rim of periosteum that had not yet produced sufficient mineralised osteoid to be visible radiographically [16]. There is rarely a soft tissue mass [51]. Laminated periosteal reaction is common in the long bones [16]. ABC can penetrate into the epiphysis through the growth cartilage [17, 22, 52, 77].

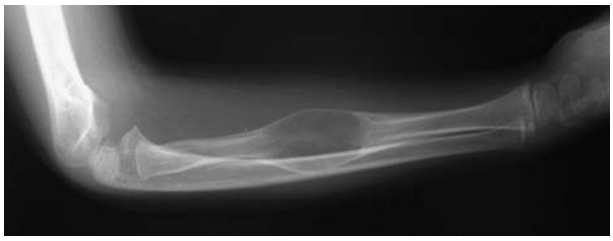

Fig. 1 Well-defined central radiolucency, elevating and inflating the periosteum, in the diaphysis of the radius (collection Pr. G. Lefort)

The imaging studies, even CT scan and MRI sometimes do not provide clear diagnostic criteria for the diagnosis of ABC and ABC is sometimes added on to a list of diagnoses including eosinophilic granuloma, giant cell tumour, unicameral bone cyst (UBC) [51]. MRI is an essential step in the investigation, particularly for pelvic and spine localisations. The radiographically differential diagnosis between ABC and UBC is sometimes not clear. Sullivan et al. [72] reviewed, in a blinded fashion, the preoperative magnetic resonance images of 14 patients with diagnostically difficult bone cysts (eight children with UBC's and six children with ABC's) and correlated these findings with diagnosis after biopsy or cyst aspiration and contrast injection. The presence of a double density fluid level within the lesion (because of layering of solid blood components) is highly suggestive for an ABC, rather than a UBC (Fig. 2). Although fluid levels are often seen, they are not pathognomic of ABC, also occurring in telangiectatic osteosarcoma, giant cell tumour and simple cysts with fracture [16, 55, 72]. Other criteria that suggested the lesion was an ABC were the presence of septations within the lesion, with soap-bubble features (particularly in T2 weighted images) with contrast enhancement of the septa and signal characteristics of low intensity on T1 images and high intensity on T2 images [22, 55, 72].

Various forms of therapy

Even if plain radiographs and MRI often support the diagnosis of ABC, accurate histologic evaluation with correlation of radiographic and MRI findings is imperative for definitive diagnosis [23]. We believe a suspected ABC should not be treated without biopsy [21–23]. Puncture of the cyst is not sufficient to make the diagnosis. Clear liquid is often observed in solitary bone cysts but it can also be found in an inactive ABC [3, 10, 17, 67, 75]. Conversely, bloody liquid can be observed in an ABC but also in fractured solitary bone cysts [3, 17]. As ABC's can arise in a malignancy (namely telangiectasic osteogenic sarcoma) or mimic it,

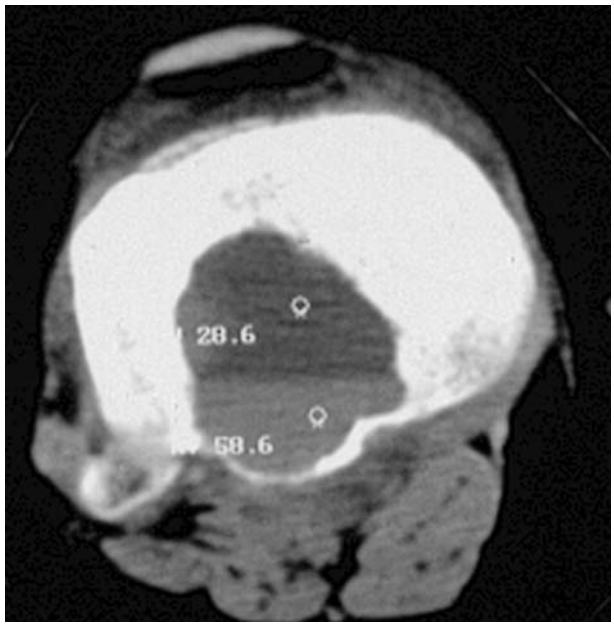

Fig. 2 Presence of a double density fluid level within the lesion. Even if fluid levels are often seen in ABC's, they are not pathognomic of ABC

the biopsy is imperative. Imaging diagnosis remains a diagnosis of probability and should always be confirmed by a biopsy [3, 17, 71]. Needle biopsies are sometimes a problem because the material obtained may consist of mostly blood elements [51]. Often, an open biopsy is necessary to establish the diagnosis [51].

Radiation therapy proved to be effective in inducing cyst ossification but it carries the risk of growth arrest, subsequent limb-length inequality and, radiation-induced sarcoma [10, 14, 49, 52, 57]. This procedure must now be avoided due to such side effects.

Treatment by perforation of the bone wall with cannulated needles introducing methylprednisolone acetate into the cyst was reported with negative results [3, 21, 69]. The lesion can show signs of further development after corticosteroid injections [3, 8, 21, 69]. Steroid injection is not indicated in patients with ABC [21].

The role of the calcitonin in the treatment of ABC is still hypothetical; it may block the activity of the osteoclasts and/or promote the formation of new bony trabecula in the fibrous septa of the ABC.

Recent methods of treatment

Percutaneous embolization with an alcoholic solution of zein

Alcoholic zein (Ethibloc®, Ethnor Laboratories/Ethicon) is a radiopaque alcoholic solution of corn protein.

Ethibloc® is a viscose sclerosing and embolizing substance able to occlude at the venous side the multiple parietal arteriolar afferents of ABC [33]. It is very important to completely fill the cavity because the therapeutic effect of Ethibloc® is due to its contact with the wall of the cyst [33]. In the largest and communicating cysts the therapy requires multiple repeated injections at intervals of 1–2 months [33]. The ensuing reparative process, which results in bone reconstruction and bone volume normalisation, lasts several months and even over 1 year after percutaneous injection (Fig. 3). This treatment remains highly controversial. For Garg et al. [35], Falappa et al. [33] and Sales de Gauzy et al. [68], direct percutaneous Ethibloc® injection can be recommended as the first-choice treatment excluding spinal lesions. Transitory local inflammatory reactions with fever and local pain limited to the injection site are common [1]. It is more a side effect than a complication. It is important to protect the skin so as to prevent a leakage of alcoholic zein into soft tissue resulting in local cellulitis. The complications and results of major series are summarised in Table 1. Falappa et al. [33] believe that careful asepsis, preliminary injection of hydrosoluble contrast medium, simultaneous monitoring during Ethibloc® injection, 10 min of manual pressure on the injection site and postprocedural antibiotic therapy could prevent major complications.

In Topouchian series [76], four patients experienced cutaneous fistulization with discharge of fibrosing agent; all four patients underwent repeated surgery with abscess and fibrosing agent excision. These complications led the authors to definitively abandon this technique in the treatment of ABC's. Mascard and Adamsbaum [53] was surprised by this article because of the high rate of local and general complications, these data contrasting with previous reports. Mascard and Adamsbaum [53] noticed that this article raised some questions about the technical conditions of Ethibloc® injection. The published images of Ethibloc® infiltration in soft tissues surrounding bone seem to indicate that excessive amounts of Ethibloc® were used, leading to the reported complications. Pulmonary embolism is a serious complication described in three papers [19, 41, 76]. Another major complication has been described in the literature. An Ethibloc® injection in an ABC of C2 in a 4-year-old boy was performed [61]. Subsequent CT and MR images demonstrated embolization material in the vertebrobasilar system. The child died of brain stem and cerebellar infarction 23 h after the intervention. Venous drainage being the main risk of Ethibloc® injection, the injection must not be used in dangerous locations. The complications encountered after percutaneous embolization of ABC with Ethibloc® encouraged some authors to abandon or to use Ethibloc® injection not as an initial treatment but an alternative to surgery. However, Ethibloc® injection should only be performed by medical team with an important experience of this treatment.

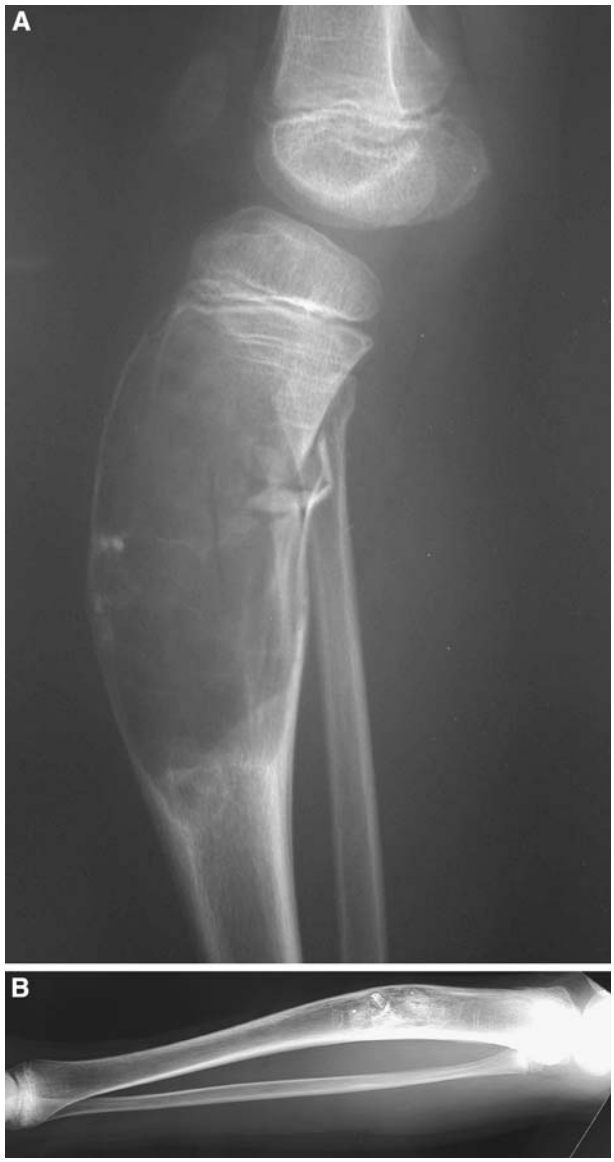

Fig. 3 **a** Initial plain radiograph shows an expansile, lucent lesion in contact with the growth plate with a pathological fracture in the upper end of the tibia in a young child. **b** Plain radiograph after percutaneous embolization with alcoholic zein without recurrence at more than 5 years follow-up

Demineralized bone particles

Delloye et al. [27, 31] reported 13 cases of induced healing of ABC's following intralesional implantation

Table 1 Complications and results of Ethibloc® treatment

Authors	Number of cases	Transitory local inflammatory reactions with local pain	Aseptic abscess, osteitis or cutaneous fistulae	Several injections[a]	Total improvement	Partial improvement	No bone reconstruction or secondary surgery
Adamsbaum [1]	17	16	3	3 (up to 3)	14	0	3
Garg [35]	10	2	1	3 (up to 2)	7	3	0
Guibaud [41]	16	5	1	6 (up to 3)	13	1	2
Falappa [33]	13	8	2	9 (up to 4)	13	0	0
Topouchian [76]	15	5	4	4 (up to 3)	9	2	4
Sales de Gauzy [68]	12	5	0	1 (up to 2)	6	3	3
Total	83	41	11	26 (up to 4)	62	9	12

[a] The first number is the number of patients who needed supplementary injections and the number in brackets is the maximal number of Ethibloc® injections

of a bone paste made of autogenic bone marrow and allogenic bone powder. Successful healing was observed in eleven cases after a mean duration of follow-up of 3.9 years. The rationale underlying this intralesional treatment was that the bone grafting material might reverse ABC expansion by promoting ossification through a bone induction mechanism. The concept of this treatment was to retain the ABC tissue, using its own intrinsic osteogenic potential to promote healing [27, 31]. Successful osteoinduction requires not only an adequate release of bone morphogenetic proteins (BMP) but also the presence of responding cells. ABC contains reactive repair tissue that has the intrinsic capacity of self-healing. Since demineralized bone matrix and bone marrow were used in combination, it is impossible to determine whether the effects were due to the inductive properties of the matrix or the introduction of osteogenic cells or growth factors contained in the marrow. By triggering new intralesional bone formation, the bone paste represented an effective means of reversing the expanding phase of ABC. The particulated bone allograft was easy to handle and to introduce into an irregular cavity [27, 31]. As no curettage is required, the proposed treatment avoids extensive surgery and blood loss and is convenient for the treatment of poorly accessible lesions such as those occurring in the pelvis.

Classical methods of treatment

Selective arterial embolization

The aim of embolization is occlusion of the vascular supply to the lesion without interfering with the vascularity of surrounding tissue or structures [26]. Selective arterial embolization (SAE) has been reported as effective treatment and can be considered in lesions whose site (spine, pelvis) or size make other types of treatment difficult or hazardous [13, 16]. This is particularly helpful at sites where it is not possible to use a tourniquet [40]. SAE has several advantages over other forms of treatment. First, it is a non-surgical technique that may be effective as primary treatment but, in the event of failure, does not preclude the possibility of intervening surgically. SAE may cause complications: ischemia to neural structures and vital structures, such as viscera, are the main concern [40].

Curettage or saucerization

In a review of the literature of some series with 20 cases or more (1,134 ABC's), the recurrence rate of the 669 ABC's treated with curettage ± bone graft was of 31%. Recurrence rates after curettage varies widely (Table 2). It is important to differentiate curettage with a small hole in the cyst wall [15], curettage through a large cortical window that allows excellent observation of the interior of the cyst [25] (usually 70–90% of the diameter of the involved bone) and saucerization [42] to understand the important variation in the recurrence rate. Whether the graft is autogenous or allogenous makes no significant difference in the recurrence rate [75]. On account of the high risk of recurrence after curettage alone, several authors used various forms of adjunctive therapy, such as phenol [12], polymethylmethacrylate [59], liquid nitrogen [50, 52, 70] or the use of a high-speed mechanical burr [36] to decrease the rate of local recurrence. In Table 2, the lower recurrence rate with adjunctive therapies shows the efficiency of these treatments.

Resection

En bloc resection is the treatment with the lowest risk of local recurrence (Table 2): 7 recurrences for 175 resections. All the recurrences were in marginal resections and not for wide ones. En bloc resection is the treatment of choice with low morbidity in eccentric

Table 2 Different types of initial treatment and recurrence rates of ABC in general series (the first number is the number of cases and the number in brackets is the number of recurrences)

Authors	Number of ABC	Curettage ± grafting	Curettage and cementation	Cryotherapy ± curettage	Curettage + radiation therapy	Curettage with a high-speed burr	Radiation therapy alone	Marginal or wide resection
Campanacci [9]	161	91 (19)	–	–	15 (3)	–	8 (2)	47 (0)
Vergel de Dios [77]	153	124 (27)	–	–	12 (2)	–	1 (0)	16 (0)
Mankin [51]	150	110 (24)	20 (5)	–	–	–	–	20 (1)
Marcove [52]	106	44 (26)	–	51 (9)	–	–	11 (1)	–
Ruiter [67]	105	82 (28)	–	–	–	–	2 (0)	21 (4)
Osaki [59]	65	30 (11)	35 (6)	–	–	–	–	–
Biesecker [5]	63	44 (26)	–	7 (1)	–	–	4 (1)	8 (0)
Dormans [25]	45	–	–	–	–	44 (8)	–	1 (0)
Gibbs [36]	40	–	–	–	–	34 (4)	–	6 (0)
Nobler [57]	33	18 (6)	–	–	1 (0)	–	6 (1)	8 (2)
Ramirez [65]	29	24 (8)	–	–	–	–	–	5 (0)
Arlet [3]	27	11 (4)	–	–	–	–	2 (0)	14 (0)
Schreuder [70]	27	–	–	27 (1)	–	–	–	–
Cole [15]	25	22 (7)	–	–	1 (0)	–	–	2 (0)
Bollini [6]	24	12 (5)	–	–	1 (0)	–	–	11 (0)
Clough [14]	21	15 (6)	–	–	2 (0)	–	1 (0)	3 (0)
Koskinen [43]	20	14 (2)	–	–	1 (0)	–	–	5 (0)
Server Perez [71]	20	17 (4)	–	–	1 (0)	–	–	2 (0)
Farsetti [34]	20	11 (2)	–	–	3 (0)	–	–	6 (0)
Total	1,134	669 (205)	55 (11)	85 (11)	37 (5)	78 (12)	35 (5)	175 (7)[a]

Tillmann's [75], Capanna's [11] and Cottalorda's series [17] are not in this table because they are earlier reports on the same patient population as respectively, Vergel de Dios' [77], Campanacci's [9] and Bollini's series [6]

[a] All the recurrences were in marginal resections and not for wide ones

lesions or in lesions arising in nonessential, expendable bones (proximal fibula, clavicle, rib, pubic ramus) [9]. In other locations, this treatment is accompanied by loss of bone and the need for reconstruction with associated morbidity [70]. In children, sub-periosteal resection is possible [6, 17]. The remaining periosteum will over several months contribute to good bone consolidation. In adults, or alternatively in some very bulky lesions, it is not technically viable to perform subperiosteal resection because the periosteum cannot be isolated. It is occasionally advisable to resort to reconstruction using a vascularised fibula bone graft [19].

Treatment option

Capanna et al. [11] described three radiographic grades. Inactive cysts have a complete periosteal shell with defined sclerotic bone limits. Active cysts have an incomplete periosteal shell and defined bone limits. Aggressive cysts have an indefinite margin and show uniform osteolysis. All the recurrences in their series were in active or aggressive tumours. There was no recurrence in the inactive types. This is important for the treatment.

The methods of treatment in the literature vary widely and treatment of an ABC remains highly controversial [23]. We believe a suspected ABC should not be treated without biopsy [23]. The management of ABC depends on the age of the patient, the location, the vascularity of the lesion, the size of the lesion and how far it has extended. Each case is unique and as such there is no "uniform" recipe. In the light of the literature however, a certain degree of consensus is emerging regarding the following propositions [23]:

- Inactive lesions can simply be monitored without any treatment [10, 22]. Sometimes, these lesions heal without recurrence with just biopsy or curettage [10, 22]. Consequently, it is not necessary to propose initially a more "aggressive" treatment and these children will be kept under supervision [20]. In the event of propitious evolution, supervision must be continued and surgery may be avoided. If the lesion increases or if it is painful, treatment must be proposed.
- For active or aggressive lesions, the surgical indication will depend on tumour location [71]. The rate of recurrence after curettage alone being high, resection offer a satisfactory theoretical solution. Marginal en bloc resection may be adopted in eccentric lesion or in expendable bones (fibula, rib, pubic ramus) with little morbidity and minimal risk of recurrence [5, 65, 77]. Exceptionally, the expansion on the lesion will require the resection of an important

osteo-articular segment. But the position of the cyst limits the indications: it is not always easy in long bones, if the tumour is juxtaposed to the growth plate, difficult in the pelvis when it is not situated in the ischium and often impossible in the spine [17]. Consequently, the most common elective treatment consists of curettage, associated or not with bone grafting. Of course, there is always a risk of recurrence, but the recurrence rate can be reduced with local adjuvants.

- In some cases, the localisation and extent of the cyst are such that operative treatment is extremely hazardous. Resection is sometimes impossible and curettage can be extremely difficult with considerable blood loss. Selective arterial embolization, in isolation or before surgery, demineralized bone particles or Ethibloc® injection makes a considerable contribution to the therapeutic solution of such cases [1, 26–28, 31, 35, 40, 41].

Conclusion

As seen previously, the cause of primary ABC appears to be multifactorial. Recent genetic and immunohistochemical studies proposed that primary ABC is a tumour and not a reactive tumour-simulating lesion. MRI is an essential step in the investigation, particularly for pelvic and spine localisations. Even if plain radiographs and MRI often support the diagnosis of ABC's, accurate histologic evaluation with correlation of radiographic and MRI findings is imperative for definitive diagnosis. Ethibloc® treatment remains highly controversial. For some authors, direct percutaneous Ethibloc® injection can be recommended as the first-choice treatment excluding spinal lesions. A minimally invasive method by introduction of demineralized bone and autogenous bone marrow is able to promote the self-healing of a primary ABC. Because ABC often affects children, aggressive surgery for this benign, sometimes self-repairing lesion should be avoided. On the other hand, its benign nature should not occult its local aggressivity which may cause neurologic complications, damage to the growth plate and therapeutic problems associated with the frequently large size of these ABCs.

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