Aneurysmal bone cyst of skull and vertebrae in children. Analysis of own material and review of the literature

Pawel Daszkiewicz¹, Marcin Roszkowski¹, Wiesława Grajkowska²

¹Department of Neurosurgery, Children’s Memorial Health Institute, Warszawa, Poland
²Department of Pathology, Children’s Memorial Health Institute, Warszawa, Poland

Aneurysmal bone cyst (ABC) is a benign, expansive, osteolytic lesion, consisting of blood-filled cysts, capable both of rapid enlargement and spontaneous resolution. Asymptomatic cases have been reported too. The aim of this paper was to analyse the outcomes of surgical treatment of ABC in children and a review of pertinent literature.

We adopted the method of retrospective analysis of medical documentation of 10 patients with ABC. These were patients at the Department of Neurosurgery of the Children’s Memorial Health Institute (Warsaw, Poland) from 1980 to 2002. There were 2 cases of cranial lesions and 8 cases of vertebral lesions. All the patients underwent surgical treatment only. Total lesionectomy was obtained in 7 cases, subtotal — in 3 cases. With the mean follow-up time of 5.1 years, good outcome (no neurological deficits) was noticed in 5 cases, moderate disability in the form of paraparesis — in 4 cases and full paraplegia — in 1 case. The following perioperative complications were noticed: transient paraplegia (1 case) and recurrence of ABC requiring reoperation (1 case). Among the 8 patients with vertebral lesions, progressive scoliosis requiring instrumental stabilization of the spine was noticed in 5 cases (4 vertebral body lesions and 1 laminar lesion).

Primary infiltration of vertebral body by an ABC may lead to subsequent progressive scoliosis, which requires instrumental stabilization of spine. This demands careful planning and development of a comprehensive treatment program. ABC in children is a predominantly aggressive lesion, but even subtotal excision does not entrain a recurrence. Localization of lesion at the D3–5 levels is associated with an increased risk of postoperative neurological deterioration. Patients should be treated surgically before the development of severe deficits, which later may prove irreversible.

key words: aneurysmal bone cyst, children, surgical treatment

INTRODUCTION

Aneurysmal bone cyst (ABC) is a benign, expansive, osteolytic lesion of bone, composed of blood-filled cysts, capable both of rapid enlargement and spontaneous regression. Asymptomatic cases have been described too. Most reports of ABC are in orthopedic literature due to frequently associated disorders of motion and instability. Neurosurgeons become interested in ABC when it causes compression of neural structures or cosmetic problems within calvarial bones.

The aim of this paper is to present our experience in this field, and to discuss current opinions and data from the literature.
MATERIAL AND METHODS
From 1980 to 2002, we have treated 10 children with ABC. This group consisted of 4 girls and 6 boys, whose mean age was 9.6 years (range 7–14 years). In this study, we adopted the method of retrospective analysis of medical documentation (medical reports, radiologic images and histopathologic slices). Additionally, we have presented a review of the available literature.

RESULTS
In the above-mentioned group of 10 children, 2 had lesions in cranial bones and 8 — in the vertebrae. Symptoms were closely related to localisation of lesions and severity of compression of the neural tissue. These were: local pain (6 patients), deformation of calvarial bones or palpable tumour of the vertebrae (1 case each), progressive scoliosis (1 patient), slowly progressive paraparesis (5 patients), neurogenic bladder (1 patient), radicular signs (1 patient), dysequilibrium and vomiting (1 case of occipital location). A minor trauma elicited symptoms in 2 patients. One patient had been treated at our institute 5 years earlier for arterio-venous malformation of the spinal cord at the level of the present ABC.

Localisation of the lesions was as follows: frontal bone (1 patient), occipital bone (1 patient), T5 (2 patients), L4, T4–T6, T7–T9, T10, C7–T1 and S2 (1 case each).

CT-scan revealed in all cases an expansive avascular, polycyclic, osteolytic lesion, with delicate septations, sometimes with fluid levels. MR imaging showed a sharply delineated lesion, which was characterised by a heterogeneous signal in all sequences and non-homogeneous contrast enhancement.

All the patients were operated on and no adjuvant treatment was given. Total excision of lesion was accomplished in 7 cases and subtotal — in 3. Radical, one-stage procedure was performed in 8 cases, while 2 patients underwent staged surgery (biopsy or partial excision) due to initial histopathologic diagnosis of a sarcoma. Only after correction of pathologic diagnosis were these children reoperated with a more radical surgery performed as second-stage procedure.

The child with a lesion in the occipital bone (with severe compression of posterior-fossa structures and acute hydrocephalus at presentation) underwent urgent shunt placement as first-line treatment. The child with his lesion in the frontal bone underwent craniectomy and plasty of bone-defect as one-stage procedure.

In the 8 children with vertebral lesions, one-stage total excision was accomplished in 5 cases, while subtotal and partial excision — in 3 cases. One of these children had the lesion totally removed at reoperation. In this group, 5 out of 8 patients required instrumental stabilisation due to progressive scoliosis. These were 4 cases of lesions localised in vertebral body and 1 case of ABC in the lamina and pedicle.

For the whole group, the mean follow-up time is 5.1 years (range 1–12). Good outcome (no neurological deficits) has been obtained in 5 patients, moderate disability (paraplegia with preserved ambulation) in 4 cases and severe disability (paraplegia and neurogenic bladder) in 1 case. It should be emphasised, however, that compared with preoperative baseline status, an improvement in pain and deficits was noticed in 9 patients. Only patient was paraplegic even before the surgery; there was no improvement in this case.

The following complications were noticed: progressive scoliosis (5 cases), transient paraplegia (1 case) and recurrence of ABC requiring reoperation (1 case). Cases of scoliosis have been discussed in more detail before. Transient neurologic deterioration to near paraplegia occurred in a child with ABC at the level of T5, infiltrating spinal canal and compressing spinal cord, who presented severe paraparesis, which disappeared after a few days of post-surgery and at present the child is ambulatory with mild monoparesis of left leg. Recurrence of ABC was diagnosed 7 months after seemingly radical excision of a lesion at the S2 level. This child underwent reoperation and now, after 6 months, is recurrence-free. In all the remaining cases, after both total and subtotal excision (2 cases), no recurrence has been observed. A child treated before at our institute for extensive arteriovenous malformation of the spinal cord of T2–T10, now presenting ABC at the T10 level, operated on while fully paraplegic, no improvement was obtained (Fig. 1–4).

DISCUSSION
According to the WHO definition, aneurysmal bone cyst (ABC) is a benign, expansile, osteolytic lesion, consisting of blood-filled cysts, separated by septations of connective tissue, bony trabeculae and giant cells of the osteoclastic group [1]. In 1942, Jaffe and Lichtenstein for the first time described ABC as a distinct nosologic entity in two young men (17- and 18-years old), with lesions located in pubic bone and at the level of C2. They believed that many cases of ABC might have been misdiagnosed as both benign or malignant bone lesions [4]. Differential diagnosis of ABC includes: low-grade osteosarcoma, teleangiectatic osteosarcoma or giant-cell tumour.

ABC is a rare condition, constituting 1–6% of all primary bone tumours. Their incidence in general popula-
Aneurysmal bone cyst (ABC) is about 1.4: 1 million, with a slight female predominance [4]. In our material, a slight male dominance was found (6:4). Familial occurrence of the disease has been reported, suggesting a possible genetic cause [3, 19]. ABC usually manifests itself in the second decade of life (70–85% of patients are under 20, and the mean age of patients in most reports is 13–17.7 years) [4]. ABC may arise in any bone. In our material, it was localised in skull and spinal column, giving rise to specific symptoms in agreement with the profile of our Department.

The aetiology and pathogenesis of ABC remains unclear [4]. Essentially, venous congestion due to disturbed blood flow within the bone leads to increased pressure within the cyst and its expansion, with subsequent compression, destruction and resorption of bone.
ysis) of adjacent bone. ABC may both enlarge rapidly, destroying bone and compressing neighbouring structures, as well as may undergo spontaneous regression [9]. According to current concepts, ABCs may be subdivided into:

1. Secondary ABCs, arising as a reaction to trauma or other bone disease (in 23–32% of cases, ABC may co-exist with primary bone tumours, metastases, vascular malformations, etc.);
2. Primary ABCs, arising de novo, with no other co-existing pathology.

Symptoms associated with enlarging ABC are: local pain, deformation of bone, limitation of movement in joints, neurological deficits (usually caused by vertebral lesions compressing spinal cord), pathologic fractures (8–21% of cases), vascular murmur and hyperaemia of skin overlying the lesion [1, 2, 10, 12]. Fully asymptomatic cases have been described (Eastwood, 2003).

Lab tests usually yield normal values, except for the activity of alcaline phosphatase, which may be slightly elevated [1, 10].

On radiography, ABC shows as an eccentric, cystic, osteolytic bone lesion. Central or subperiosteal lesions are rare. Bone has a characteristic “soap bubble” structure, while the lesion may be surrounded by an osteosclerotic ring (“egg-shell” sign). CT-scan may reveal cystic, osteolytic lesions with internal septae, sometimes with fluid-levels. It is well known, that CT is a “golden standard” for diagnosis of bone lesions, while MRI is superior in documenting the degree of infiltration of adjacent soft-tissues [2, 20]. Bone scan may show enhanced accumulation of isotope around the lesion, while angiography may reveal a vascular “blush” around ABC with an area of pooling of contrast, but without distinct feeding arteries nor draining veins. In many authors suggest that the appearance of ABC on contemporary imaging studies is specific, enabling a fairly reliable diagnosis based on them alone. In cases of doubt, biopsy is warranted in order to rule out any co-existing pathology. Others recommend open or needle biopsy in all cases [8].

Macroscopic appearance of ABC is that of a blood-filled sponge. The shell of new bone encloses blood- or serum-filled cysts of various sizes, separated by brownish septa. Microscopic examination reveals that septa are composed of loosely arranged spindle cells and giant polyynuclear cells, proliferating capillaries and areas of osteoblastic activity leading to formation of new bone and bony trabeculae. Spindle cells show mitotic activity but no cytologic atypia. Areas of new bone formation, reactive bone and mitotic figures are present [11]. There are solid forms of ABC, with no cystic component [10, 13]. The name “solid ABC” has been adopted for this entity.

ABC may take one of the following clinical forms defined by Enneking [4]:

1. Occult (inactive form), asymptomatic and diagnosed accidentally. Pathologic fractures and other disorders are rare, the lesion is painless, remains circumscribed, does not progress and does not cause deformation of bone. Radiography may show a small, circumscribed lesion, surrounded by mature bone with no signs of expansion or deformation of adjacent bone. There is no accumulation of isotope on bone scan, neither neovascularisation on angiography. CT-scan shows a homogenous, well-circumscribed lesion, which does not cross cortical bone. Microscopic study reveals low cell/stroma ratio, mature and well differentiated stroma, and encapsulation of lesion by mature connective fibrous tissue or cortical bone.
2. Active form, may cause discomfort, pathologic fractures or dysfunction of extremity, shows progressive enlargement with expansion of bone. The lesion is still fully encapsulated. Radiography shows a well circumscribed lesion with internal septae. Bone scan may show an enhanced perilesional uptake of isotope. Microscopic examination reveals a still normal cell: stroma ratio, a distinct capsule composed of mature fibrous tissue and spongious bone and a narrow area of reactive inflammatory and vascular tissue between the capsule and adjacent normal tissue. Expansion is due to bone resorption by osteoclasts, although the area of resorption is irregular, leading to a corrugated cyst-bone interface.
3. Aggressive form, which, although histologically benign, behaves as a low-grade neoplasm. Symptoms are due to enlargement of the pathologic mass, compression of adjacent structures and pathologic fractures [2]. The lesion is usually painful on palpation, crosses natural barriers of cortical bone, fasciae and cartilage, while its capsule is often penetrated by finger-like extrusions of pathologic tissue into adjacent structures. Radiography shows a lesion with irregular margin, destruction of cortical bone and expansion towards soft tissues. Bone scan shows an increased uptake of isotope and angiography— a distinct zone of neovascularisation with pooling of contrast. CT-scan shows a heterodense lesion with irregular margin. Microscopic examination reveals high cell/stroma ratio, areas of variable maturity, more mitotic figures, infiltration of vessels and foci of ABC in neighbouring tissues, sometimes seemingly with no contact with primary lesion.
All the cases in our material are consistent with the aggressive (7 cases) or at best active form (3 cases). Occult or inactive forms were not present in our material. We may then conclude that most cases of ABC in children are aggressive forms.

Currently, rare cases of occult lesions are diagnosed incidentally. When the diagnosis is certain and the lesion itself is stable, most authors recommend close follow-up because of spontaneous regression of ABC [13].

The mainstay of treatment of symptomatic ABC is surgical resection [1], if only classic requirements of “anatomical accessibility and functional plausibility” are fulfilled. Recommended scope of surgery varies from simple curettage, or curettage with resection of inner lining of bony cavity using rongeurs or high-speed drills. Some recommend additional topical application of cytotoxic agents, like phenol, liquid nitrogen, methylmethacrylate or absolute alcohol [6, 8] or endovascular embolisation when possible [15]. The use of these agents is questionable, as unequivocal proof of their effectiveness is lacking, and their use is associated with a real risk of irreversible damage to surrounding structures. At our institute, we do not use such adjunctive measures, so we do not have any experience in this field. In the case of recurrence, most authors recommend reoperation and “en bloc” resection. Obviously, such procedures are often mutilating and, in the case of spine, often destabilising.

When contemplating surgical treatment of ABC, the following issues must be kept in mind:
1. **ABC of the skull** creates mainly neurologic and cosmetic problems.
2. **ABC of the extremities** in close contact with epiphyses and joints, as well as surgical procedures in this area, may result in arrest of growth and motion of the affected extremity — patients and their relatives should be informed about this potential sequelae.
3. **ABC of spine** may lead to neurologic deficits (spinal cord compression) and instability; spinal surgery may be associated with sizeable blood loss [of importance in small children].

In our material, progressive scoliosis, which required instrumental stabilisation, occurred in 5 cases (out of 8 spinal lesions). In 3 remaining cases, external brace (in the postoperative period) and rehabilitation effectively preserved spinal alignment. Noteworthy is, that in the group of 5 children requiring instrumentation, the lesion was localised in vertebral body in 4 cases and in laminae in 1 case only. This is understandable in view of the generally accepted theory of spinal biomechanics [16]. In the setting of spinal trauma, the cornerstone of treatment is the “three columns theory” elaborated by Denis (the first column — first half of vertebral body, anterior longitudinal ligament and first half of fibrous ring; the second column — posterior half of vertebral body, posterior longitudinal ligament and posterior half of fibrous ring; the third column — vertebral laminae, intervertebral joints, interspinous and yellow ligaments). Spine is considered unstable if 2 out of the three above mentioned columns are damaged. However, in the setting of spinal tumours (pathogenesis and biomechanics of ABCs make them more similar to spinal tumours), the Denis concept is not applicable, as trauma and tumor vary greatly in the extent of damage to vertebrae, intervertebral disks, ligaments and joints, the quality of adjacent tissues and their potential to heal. Kostuki and Errico proposed a system of evaluation of spinal stability in patients with spinal tumours. According to their theory, spine is composed of 6 columns: the 3 columns by Denis, further subdivided into the right and left halves. Spine is considered stable if 1 or 2 columns are damaged, moderately unstable if 3 or 4 columns are disrupted and highly unstable if 5–6 columns have been destroyed. Co-existent angulation of spinal axis of over 20 degrees and pain on movement, signify instability [16].

Radiotherapy has been used in the past to treat ABCs, but now it’s use is subject to much criticism due to the risk of induction of secondary neoplasms and gonadal and growth-plate damage. Nevertheless, its application may be considered if other treatment modalities (surgery) are impractical [5].

The following complications of surgical treatment of ABC are quoted [1, 4, 13, 18]: recurrence, intraoperative haemorrhage or postoperative haematoma, infection with wound dehiscence and osteitis, damage to adjacent structures (inferior vena cava, dura, spinal cord and nerves) with concordant sequels (haemorrhage, neurologic deficit, liquorrhoea), pulmonary embolism, chronic back pain and scoliosis. We noticed the following complications: transient paraplegia (1 case), recurrence (1 case) and progressive scoliosis (5 cases). Intraoperative blood loss was typical for spinal surgery and even in small children (the youngest was 7 years old) it was easily controlled with adequate fluid and blood transfusions. No other complications were noticed.

Prognosis in ABCs is usually excellent, and overall proportion of cure is 90–95%. The incidence of recurrence is 0% after extensive resection, about 7% after resection of cyst with capsule, about 12% after curettage with radiotherapy and nearly 30% after simple curettage [4]. Nearly all cases of recurrence reported to-date, occurred within 2 years after surgery. However, a 5-years follow-up is recommended (in children — until puberty). In this paper, the mean follow-up time is 5.1
years, so it exceeds the recommended span and our observations are reliable. Until now, our only case of recurrence was seen 7 months after a seemingly total excision of ABC. On the other hand, no signs of disease progression were seen in two other patients after incomplete excision and 2 and 3 years of follow-up. In all other cases, generally very good results of treatment seen in our material only confirm literature data.

We must keep in mind, that in about 30% of cases of ABC, particularly in adults, there is a co-existing pathology (usually bone dysplasia or giant-cell tumour), where both treatment strategy and prognosis depend on the nature of underlying disease [14, 21].

A survey of Polish scientific literature concerning ABC, yielded mainly case reports. Depending on location of lesion, leading signs and profile of professional interest of authors, particular papers deal with neurological [10, 22], orthopaedic [7, 23], and oto-rhino-laryngologic [10, 22] aspects of the disease.

CONCLUSIONS

- Primary localisation of ABC in vertebral body will usually lead to progressive scoliosis requiring instrumental stabilisation, thus the need for careful planning of treatment strategy;
- Most cases of ABC in children are of the “aggressive” type, but even incomplete excision of the lesion makes possible a long, progression-free survival;
- Lesions at the T5 level are associated with high risk of neurologic deterioration after surgery;
- Surgical excision remains the cornerstone of treatment of ABC, and should be instituted before the appearance of potentially irreversible neurological deficits.

REFERENCES