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## Case Report

# Spontaneous rapid regression of a juvenile primary aneurysmal bone cyst of the skull: A case report and literature review ☆☆☆

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## ABSTRACT

Aneurysmal bone cyst (ABC) is a benign lesion that often starts off the metaphysis of long bones and which, as it grows, may blow out bone. Only 3%-6% of cases are located in the skull. Spontaneous recovery has been reported. These cases occur more often in adults and in pelvic locations. Spontaneous regression at the skull level remains a very rare entity and few cases were described in the literature. Here, the authors report another case of spontaneous rapid regression of ABC of the skull in a 7-year-old boy revealed by gradually increasing painless hard swelling in the right frontal bone region with rapid spontaneous regression within 15 days. The authors will also proceed with an overview concerning this rare entity.

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## Introduction

Aneurysmal bone cyst (ABC) is a benign lesion that often starts off the metaphysis of long bones and which, as it grows, may blow out bone. This pseudotumor can also affect the pelvis and the spine. Only 3%-6% of cases are located in the skull [1,2,3]. This lesion may cause pain of varying intensity as well as a swelling that becomes apparent if the tumor is blowing. Although a standard x-ray may sometimes be sufficient to make diagnosis, magnetic resonance imaging (MRI) is bet-

ter for differential diagnosis. Biopsy is necessary because the cyst is sometimes secondary to a malignant lesion such as telangiectatic osteosarcoma. Spontaneous recovery has been reported [2,14]. These cases occur more often in adults and in pelvic locations. Spontaneous regression at the skull level remains very rare and few cases were reported in the literature [16].

Here, the authors report another rare case of spontaneous rapid regression within 15 days of ABC of the skull in a 7-year-old boy revealed by gradually increasing painless hard swelling in the right frontal region. The authors will proceed with an overview concerning this rare entity

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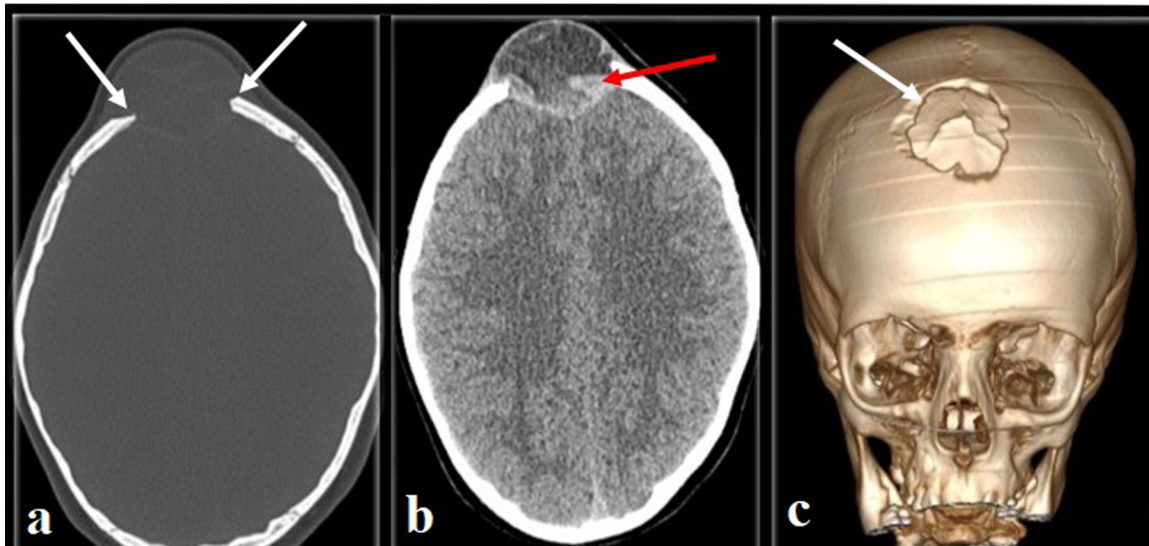
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**Fig. 1 – Axial brain CT scan in bone (A) and soft tissue window (B) with a three-dimensional (3D) reconstruction in anterior view (C) showing a local cortical destruction of both outer and inner frontal bone cortex (white arrows). Note the spontaneously hyperdense signal indicating hemorrhagic sediment (red arrow) (Color version of the figure is available online.)**

## Case report

A 7-year-old boy with no particular medical or surgical history presented to our department with a 3-month history of gradually increasing painless hard swelling in the right frontal bone region just behind his natural hairline. Physical examination revealed a painless and non-tender mass,  $4 \times 4.5 \times 4$  cm in size, which was firm to hard, and fixed to bone with smooth surface. The skin over this swelling had a normal appearance with no redness, heat or itchiness. There was no neurologic palsy. General condition examination was without abnormalities. All routine investigations were within normal limits.

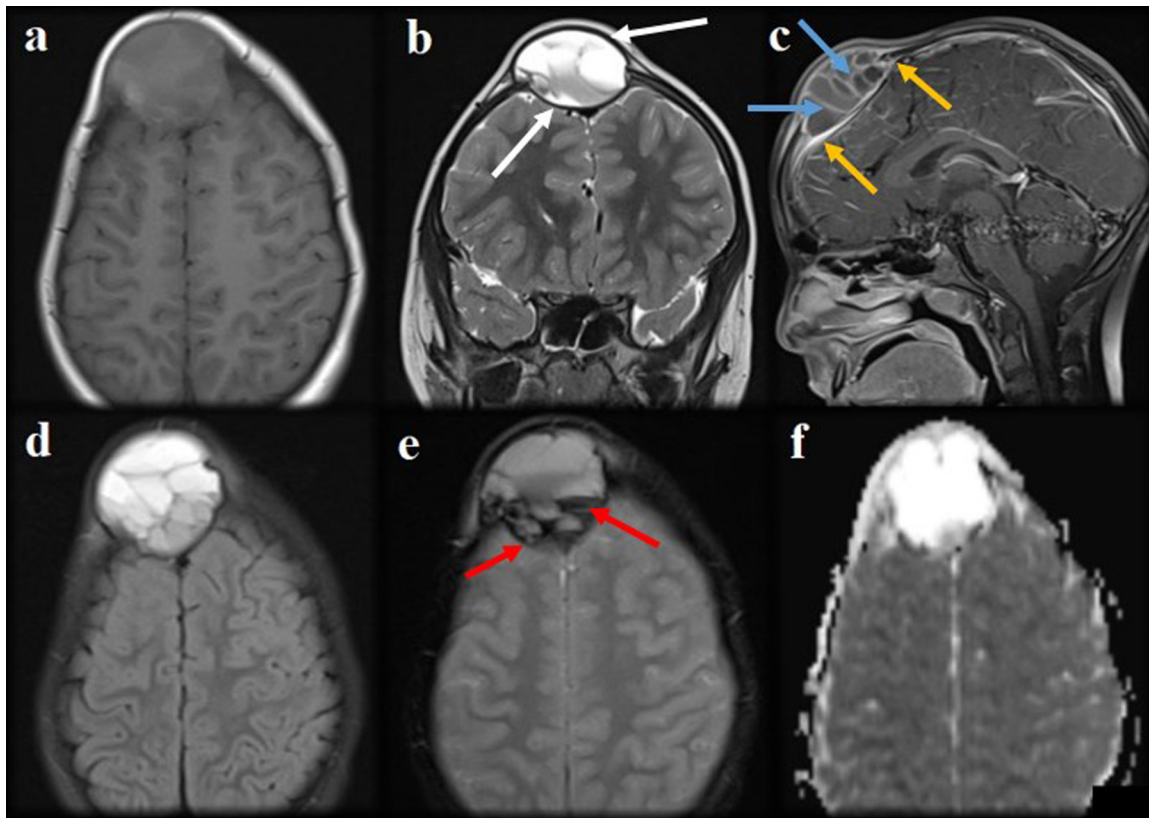
Plain X-ray skull was not performed and a computed tomography (CT) scan with and without contrast enhancement showed an expansile extraaxial osteolytic heterogeneous mixed density mass of the right frontal bone of 4.5 cm in diameter causing break and thinning of both outer and inner cortex. It was in direct contact with the superior sagittal venous sinus with no evident sign of tumor involvement or thrombosis. This lesion had internal septation giving a soap-bubble aspect, taking irregular variegated enhancement. Some cysts are spontaneously hyperdense indicating hemorrhagic sediment (Fig. 1). No solid nodule was detected.

MRI revealed a  $58 \times 45 \times 30$  mm well-defined frontal cystic mass of soap-bubble appearance with well-defined hypointense capsule. This multilocular cystic mass was well circumscribed. It was laminating both the inner and outer tables of the cranial vault and exerting pressure over the superior sagittal venous sinus conserving its permeability. The frontal lobe of brain parenchyma was not compressed. Most

of the cyst's internal contents had low signal intensity on T1-weighted images and high signal intensity on T2-weighted images and T2-Fluid-attenuated inversion recovery (FLAIR) sequences. It was associated with multilocular zones with fluid-fluid levels having a high intensity signal on T1-weighted images and an intermediate intensity signal on T2 sequence indicating hemorrhage. The cyst was heterogeneously enhanced after gadolinium chelates injection including its septa. No restricted signal was found on diffusion-weighted image (DWI). On the GRE T2\*-weighted image, hyposignals predominating on the posterior, and sloping side of the lesion were also seen (Fig. 2).

A selective transfemoral angiographic study of both external carotid arteries was performed to identify the main blood-supplying vessel and allow its embolization prior to surgery. The finding was that tumor derived its blood supply mainly from both anterior and posterior frontal branches on selective injection of the superficial temporal artery with no other contribution to this circulation (Fig. 3). Surgery without embolization was therefore decided through a horseshoe frontal skin flap. We planned a craniectomy with coagulation of the supplying artery associated to an en bloc removal of the lesion with a narrow margin of surrounding normal bone followed by a cranioplasty repairing the skull vault defects.

Surgery was, unfortunately, postponed because of the severe acute respiratory syndrome CoV (SARS-CoV-2 or COVID-19) crisis in our hospital. The boy was thus discharged from our department with an appointment at our outpatient clinic. He presented 2 weeks later at our outpatient clinic for spontaneous regression of his painless swelling. The MRI showed that the overall size of the cyst had hugely decreased (Fig. 4). Three months later, the pa-



**Fig. 2 – Brain MR. (A) axial T1-weighted image; (B) coronal T2-weighted image; (C) sagittal plane after enhancement; (D) axial T2 FLAIR weighted image; (E) axial GRE T2\*-weighted image; (F) axial diffusion-weighted image (DWI). They show a well-defined frontal extradural mass with internal hypointensity in T1 and hyperintensity on FLAIR and T2. A thin rim of low signal intensity capsule surrounds the nodule (white arrows). The cyst represses the superior sagittal venous sinus without thrombosis (yellow arrows). There was a septal enhancement after gadolinium chelates injection (blue arrows). The image shows multilocular zones with fluid-fluid levels on GRE T2\*-weighted image. A hypointense hemosiderin deposits are also seen on the posterior and declining zones (red arrows). There was no signal restriction on DWI (Color version of the figure is available online.)**

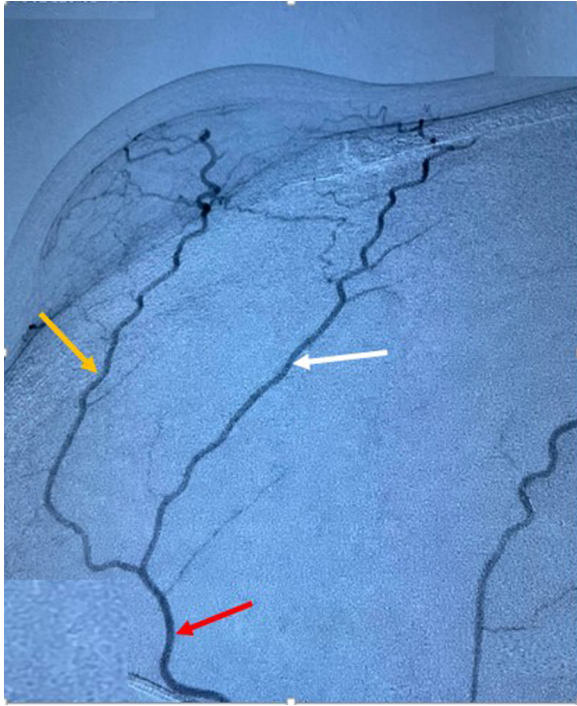
tient was free of symptoms, and there was no evidence of recurrence.

## Discussion

The aneurysmal bone cyst (ABC) was originally described in 1942 by Jaffe and Lichtenstein in 2 adolescent patients as lesion with a "soap bubble" aspect. ABCs were extensible and showed signs of adjacent bone erosion and tissue invasion. Upon surgical exploration of the lesion, a thin bone wall with hematic fluid content has been described [1]. It is a benign, osteolytic, expandable lesion of blood-filled spaces of different sizes, separated by connective tissue partitions with trabeculae or osteoid tissue and osteoclasts. The stretchable nature of these lesions may cause local pain, edema, deformity, and even pathologic fractures [1,2]. This is a pathologic entity with rare presentation; it is representative of 6% of its primitive bones. It is in fact difficult to estimate the incidence due to spontaneously regressive cases or silent clinics. The mean

age of presentation is 13-17.7 years, with the same distribution by sex, although some authors have observed a higher incidence in women. It most often affects long bones, vertebrae, and flat bones. Only 3% to 6% of cases are located in the skull [1,2,3].

Its etiology is still unknown. Through a review of literature, we can classify it as primary, when there is no evidence of another lesion, and secondary to associated tumors in 23%-32%. The most widespread theory considers that ABC is a reaction process to intraosseous or subperiosteal hemorrhage. This latter is thought to be due to a local circulatory anomaly [4] and blocked venous drainage leading to an increase in venous pressure and resulting in a dilation of the local vascular network. When there is an initial vascular injury, pressure increases, causes expansion, erosion, and reabsorption of adjacent bone tissues [1]. The most common associated neoplasm is the giant cell tumor. Other benign or malignant tumors may be found, such as fibrous dysplasia, osteoblastoma, chondrosarcoma, and metastatic carcinoma. Trauma has been described as another probable cause [1,3]. In our patient, there was no evidence of prior head injury including trauma lead-



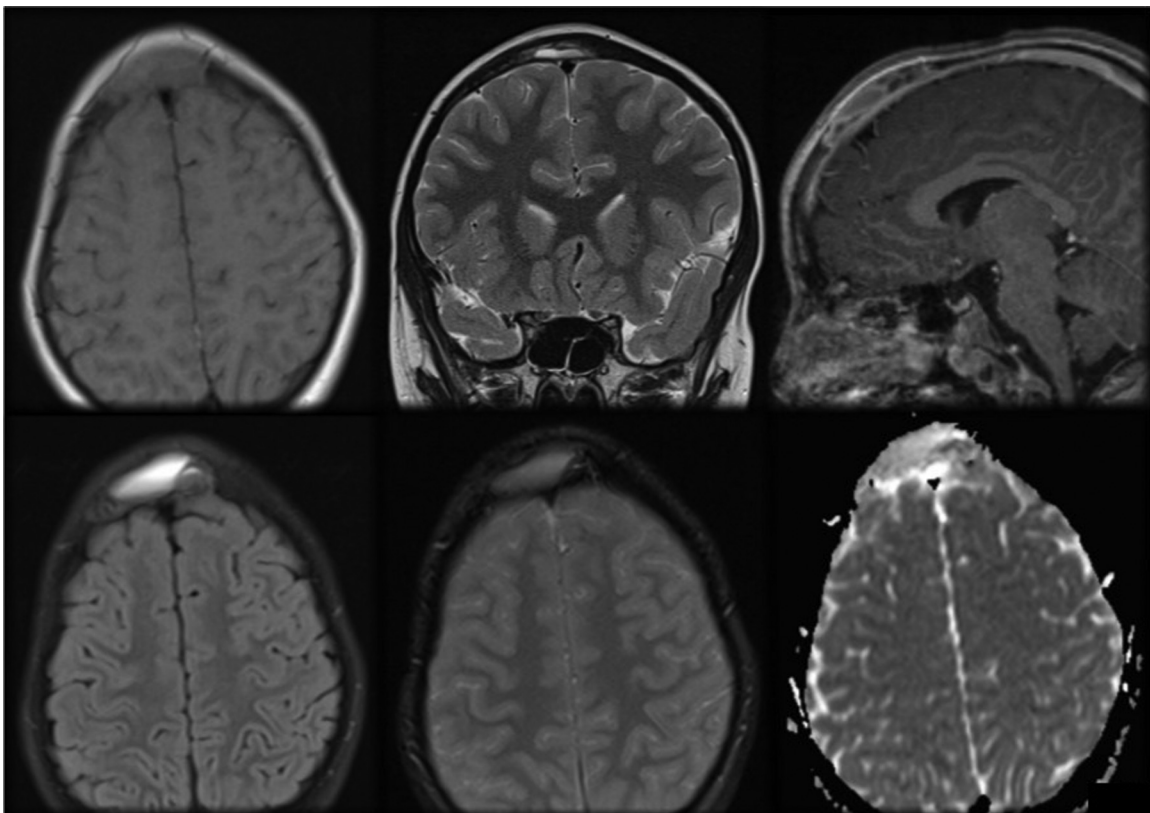
**Fig. 3 – Selective angiogram of the superficial temporal artery (red arrow), arterial phase, lateral view showing blood supply mainly from both anterior (yellow arrow), and posterior frontal branches (white arrow) (Color version of the figure is available online.)**

ing to hemorrhage. That's why we can consider our case as a primary cyst.

Few cases of aneurysmal bone cyst have been studied cytogenetically, but abnormalities have been identified in the short arm of chromosome 17. The 17p karyotypic abnormality consists of a translocation and inversion [5].

The main symptoms associated with ABC are local pain and painful swelling. This pain would come from microcracks due to cortical weakening. Swelling may be significant and is due to the puffiness of the lesion. Pathologic fractures are rare with long bones and are more frequent for central ABC than for eccentric ones. Pathologic fractures are more common on the spine [6]. At this site, pain may lead to segmental stiffness with scoliosis or torticollis and neurologic symptoms are not uncommon (45% of cases) [7]. When it comes to the skull, there may be neurologic symptoms characterized by headache, proptosis, focal neurologic symptoms, and increased intracranial pressure [1,8]. The cyst's size may increase during pregnancy [8]. Our patient did not experience any of these neurologic symptoms except the gradually increasing painless hard swelling in his right frontal bone region.

Plain radiography and axial computerized tomography (CT) with bone window show the osteolytic lesion surrounded by thinned and expanded cortical bone in the shape of an eggshell without calcification of its matrix as it was seen in our patient. CT scan shows areas of decreased and increased density that are reinforced with the contrast medium. Other diseases such as simple bone cyst, giant cell tumor, chondroblas-



**Fig. 4 – Multiplanar MRI images showing a significant regression of the cyst's size compared to the first MRI.**

toma, chondromyxoid fibroma, telangiectatic osteosarcoma, and fibrous dysplasia may have the same radiological characteristics [9,10]. Magnetic resonance imaging (MRI) shows that lesion's fluid content has high T2-weighted signal and low T1 weighted signal as it was in our patient's MRI. The T2 weighting will mainly highlight the fluid content and liquid levels. T1 weighting better shows the cortical bone. Liquid levels are present in 66%-84% of cases [11]. In our patient, a fluid-fluid level was seen on GRE T2\*-weighted image. The presence of these liquid levels is by no means specific to ABC [12]. It is simply a reflection of the presence of liquids of different densities. Angiography usually shows venous malformations with prolonged persistence of the contrast agent inside the cyst. There is no arterial malformation or arteriovenous fistula [13]. This examination is usually only performed as part of preoperative or therapeutic embolization, but not for diagnostic purposes.

Regarding the natural course, cases of spontaneous recovery have been reported [2,14]. These cases occur more often in adults and in pelvic locations. This healing may also occur after biopsy [15]. In 2007, Broc-Haro et al. reported a case of a skull ABC in an 8-year-old asymptomatic female patient. The finding was made by her mother when palpating a sinking area in her right frontal region. Spontaneous regression was seen 4 months after diagnosis [16]. To our knowledge, our case is the second case reported in the literature, and the first case reporting a spontaneous regression in such short period of 15 days. No other case of spontaneous regression at the skull level had been reported.

Several cases of malignant transformation have been also reported [17,18]. Brindley et al. reported 2 cases of malignant transformation into telangiectatic osteosarcoma and fibroblastic osteosarcoma 5 years and 12 years after ABC curettage [17]. Kyriakos et al. report a case of transformation into pleomorphic osteosarcoma after several curettages [18].

Intralesional resection and curettage have been suggested as the first-line treatment, with a recurrence rate of 20% to 30%. Other treatments have been recommended, such as percutaneous injection of calcitonin and methylprednisolone [19], selective arterial embolization, local resection with a recurrence rate up to 50%; en bloc excision without dural involvement is generally curative and radiotherapy only in recurrent or unresectable cases, always with the risk of developing sarcoma [1,2,20]. Preoperative embolization has been suggested in order to reduce intraoperative bleeding [20].

In cases with ABCs that do not show symptoms, we consider it necessary to carry out serial radiological studies before definitive treatment, to document spontaneous regression with more precision, and clarity.

## Conclusion

Aneurysmal bone cyst is a benign lesion that affects children and young adults. Diagnosis can be probable with a simple x-ray, but magnetic resonance is often useful in the differential diagnosis. The biopsy is necessary since the aneurysmal cyst may be secondary to a malignant tumor such as telangiectatic osteosarcoma. Spontaneous recovery or after biopsies have already been reported but remain rare. Most often treatment is

necessary. As the tumor is benign, treatment that is too invasive, and dilapidated should be avoided. Many minimally invasive treatments are currently available.

## Patient consent

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

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