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CASE REPORT

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Solid variant of aneurysmal bone cyst: a case report with bilateral involvement of the distal femoral metaphyses

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Abstract An extremely rare case of bilateral, symmetrical involvement of distal femoral metaphyses by the solid variant of aneurysmal bone cyst (ABC) in a boy aged 13 years is described. Although there is no difference between the conventional ABC and the solid variant in terms of clinical and radiological presentation, the lesion is solid, composed of fibrohistiocytic cells with abundant giant cells and/or areas of florid, heterotopic ossification, while aneurysmal channels are sparse or absent. The lesion needs to be differentiated from giant cell tumour of bone, when the osteoclastic component predominates, while fibrous dysplasia, osteoblastoma and even osteosarcoma need to be excluded any time ossification is prominent. Careful evalution of the clinical, radiological and pathological findings is necessary.

Key words Bone tumour-like lesions · Solid variant aneurysmal bone cyst · Giant cell tumour · Differential diagnosi

Introduction

Aneurysmal bone cyst (ABC) is an expansive, non-neoplastic, tumour-like lesion, arising either primarily or developing as a secondary change to other benign or malignant bone tumours [10, 12, 14, 19]. The typical primary ABC is characterized by cavernous channels, spindle celled stroma with osteoclast-like giant cells and osteoid or bone production with or without osteoblastic rimming [10]. Apart from the purely cystic lesions, the existence of a so-called solid variant was described by Sanerkin et al. in 1983 [16]. This is characterized by the scarcity of cavernous channels and the fibrohistiocytic, giant cell reaction and/or osteoblastic proliferation.

Both variants of ABC can be mistaken for a variety of bone tumours and other tumour-like bone lesions [1, 7, 12, 14, 16, 19].

We present a unique case of bilateral solid ABC.

Case report

A 13-year-old boy was admitted to a hospital complaining of pain in his left knee. Radiographic examination revealed a lytic lesion at the left distal femoral metaphysis (Fig. 1). Surgical treatment was recommended, but the patient's parents refused surgery. At the age of 16 years, after a pathological fracture, the lesion was completely excised and the fracture treated by osteosynthesis using an autologous fibular bone graft. The pathology report de-

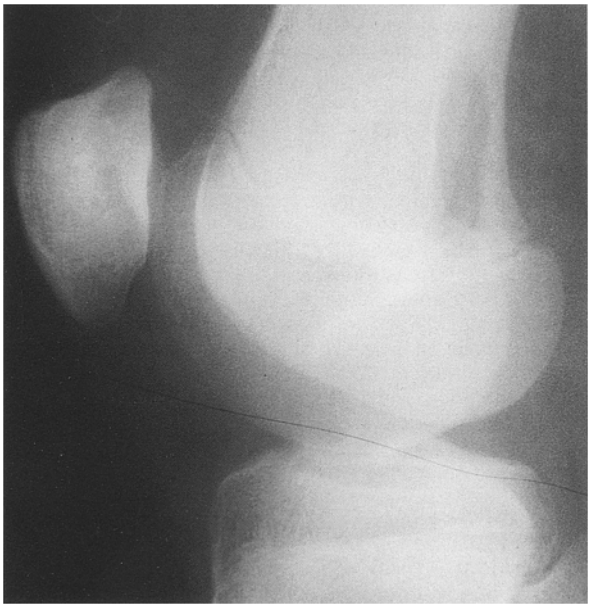

Fig. 1 Radiograph of the osteolytic lesion of the distal left femoral metaphysis

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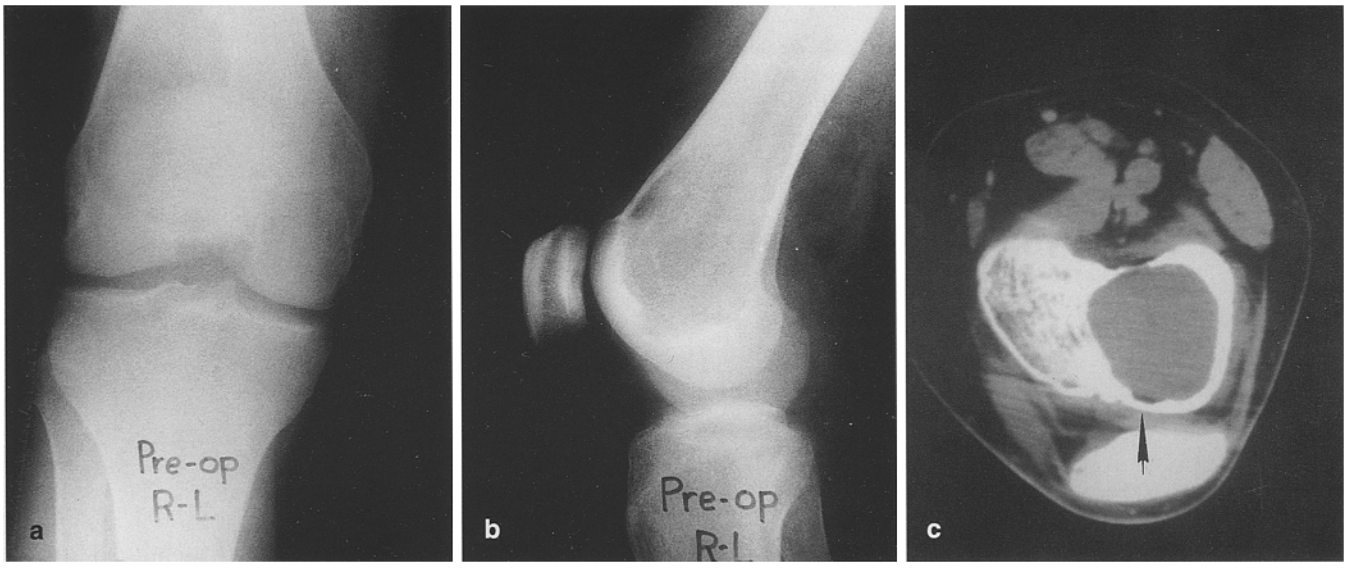

Fig. 2a–c The radiological appearance of the second bone lesion. Phase (**a**) and profile (**b**) aspect of the metaphyseal lesion of the right femur. **c** CT scan of the lytic lesion of the right femur presenting a soft tissue density. Only a small area of low density was noticed. (*arrowhead*)

scribed the lesion as fibrous dysplasia. No further radiographic skeletal survey was done to the patient who, soon after his mobilization, started complaining of pain in his right knee which gradually worsened. On X-ray, a lytic lesion appeared in the distal metaphysis of the right femur, which on CT scan was well demarcated without cortical involvement (Fig. 2a, b, c). Re-evaluation of the histological slides from the first lesion revealed that the lesion was solid, composed of fibroblastic stroma with few areas of hyalinization. Interlacing bone trabeculae with a rim of proliferating osteoblasts were observed throughout the stroma showing a tendency toward maturation (Fig. 3a, b). Focally, foci composed of osteoclast-like giant cells as well as a few small aneurysmal spaces were also found. A final diagnosis of a solid ABC was made, which was confirmed by other opinions. Surgical treatment of the second lesion in the right femur was undertaken and after removal of the thin cortical shell, the osteolytic cavity was found to be filled up with solid, fleshy and friable tissue with scattered haemorrhagic areas. An intralesional excision (curettage) was undertaken and the cavity was then washed with alcohol and tightly packed with bone graft.

The multiple tissue pieces measured 5.5×4.5×4 cm. They were mainly soft and granular with few haemorrhagic areas; some were firm and gritty in consistency. The tissue was fixed in buffered formalin, for 24 h and embedded in synthetic paraffin through the usual procedure. Decalcification was performed on a few tissue pieces using a 7% solution of nitric acid.

Histologically the lesion presented large areas reminiscent of giant cell tumour of bone. These areas were separated by broad bands of fibroblastic or fibrohistiocytic tissue with foci of osteoblastic proliferation and osteoid deposition. Fibromyxoid tissue was found focally, while aneurysmal vascular spaces were readily identified (Fig. 4a, b).

Immunohistochemistry was performed on serial 4 μm thick sections from representative, undecalcified, tissue blocks, mounted on poly-L-lysine (Biomakor) coated glass slides. Monoclonal antibodies for the detection of vascular endothelium (CD31 clone JC/70A, DAKO) [5] and smooth muscle cells α-smooth muscle actin [(α-SMA) clone 1A4, Biomakor] were applied overnight at optimal dilution (CD31 1:50 and α-SMA 1:3000) at 4° C in a moist chamber. Pre-digestion with porcine pancreatic trypsin type II 0.1% (Sigma) for 20 min at 37° C was performed before the application of CD31. All immunoreactions were developed with the streptavidin-biotin-immunoperoxidase system (Immunon/Shandon) in the Sequenza semi-automated immunostainer (Shandon). The immunoprecipitate was visualized with 3′3′-diaminobenzidine tetrahydrochloride as a chromogen (Sigma) checking the results under the microscope. The blood vessels of the tissue served as positive controls, while in control sections the primary antibodies were omitted and the sections were incubated with buffer solution. Besides the wall of the stromal vessels, many stromal cells particularly around the few small cystic spaces reacted strongly positive with α-SMA, while most of the flattened cells lining these spaces expressed CD31 immunoreactivity (Fig. 3a, 4c).

Extensive biochemical and hormonal investigation, as well as a detailed bone imaging survey were carried out scrutinizing the possibility of hyperparathyroidism but all tests were normal and no other bone lesions were identified. Six months after the second operation both lesions showed sound healing and the patient regained full activity.

Discussion

ABC is a non-neoplastic bone lesion, the precise nature of which remains unclear [12, 20]. Except for primary ABC with a characteristic clinicopathological picture, it seems that aneurysmal changes are sometimes found focally in bone lesions of divergent nature [10, 12, 14, 19]. Sanerkin et al. [16] described a variant of ABC in which the dilated sinusoidal cavities are sparse or absent, while the tumour is mainly solid and composed of fibroblastic or fibromyxoid stroma with osteoblastic and osteoclastic elements. After this first description based upon four cases, two more series comprising eight cases [19] and 15 cases [1] respectively, as well as one case report by Edel et al. [7] appeared in the literature. In all of these cases, the location of the bone lesion, the patient's age and the radiographic features were those of conventional ABC. However, the scarcity of aneurysmatic channels in combination with the fibrohistiocytic and/or osteoclast-rich areas, gave rise to differential diagnostic problems from giant cell tumour of bone [16, 19] while in some cases with extensive osteoblastic proliferation and oste-

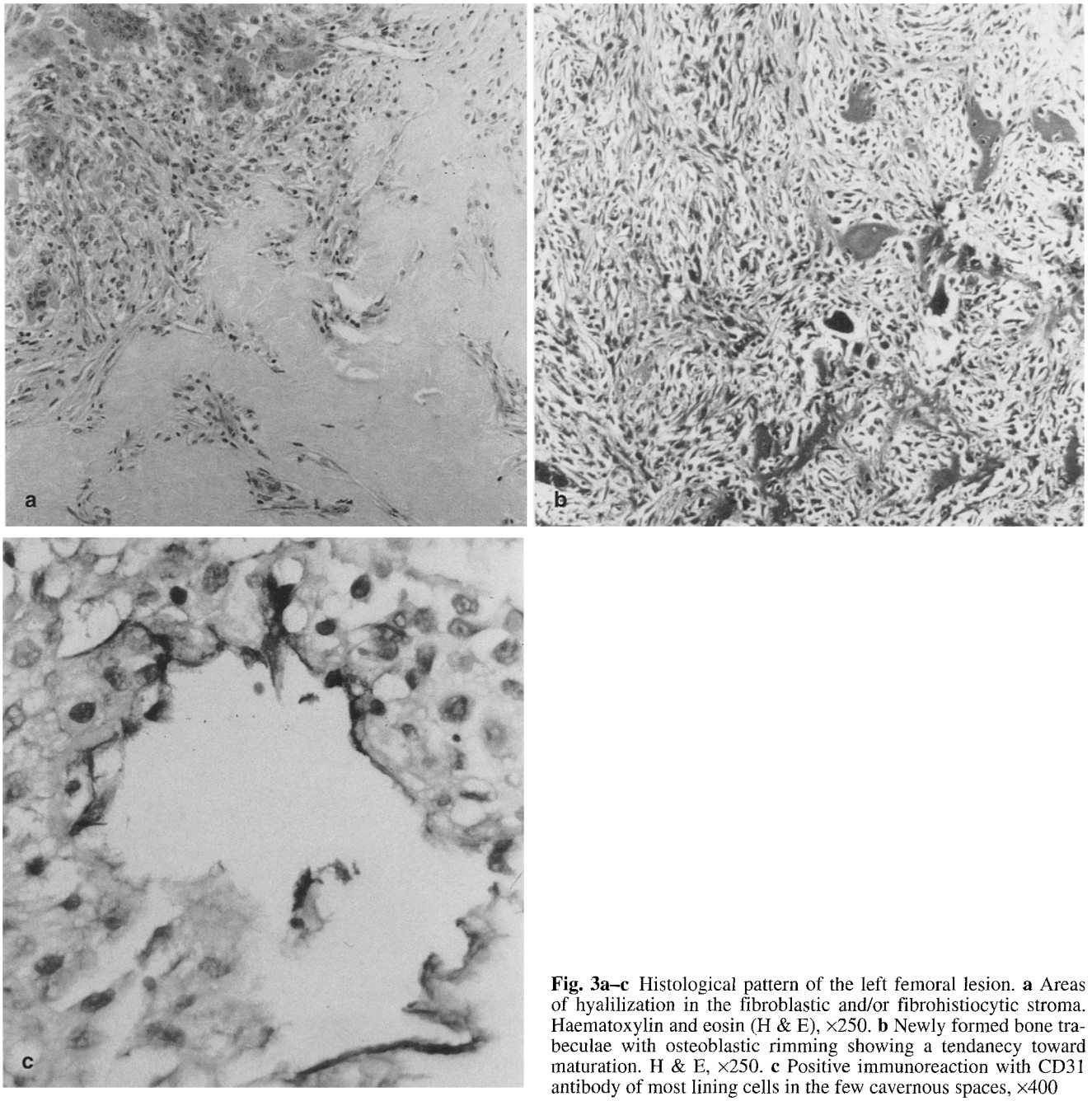

Fig. 3a–c Histological pattern of the left femoral lesion. **a** Areas of hyalilization in the fibroblastic and/or fibrohistiocytic stroma. Haematoxylin and eosin (H & E), ×250. **b** Newly formed bone trabeculae with osteoblastic rimming showing a tendanecy toward maturation. H & E, ×250. **c** Positive immunoreaction with CD31 antibody of most lining cells in the few cavernous spaces, ×400

oid production, osteoblastoma or even osteosarcoma need to be ruled out [1, 7, 16, 18, 19].

Both lesions here presented, large areas reminiscent of giant cell tumour of bone with a minor aneurysmal component. However, their metaphyseal location and the development of at least the first at the age of 13 years was against the diagnosis of giant cell tumour of bone, which is exceptional under the age of 15 years [16]. Although the diagnosis of fibrous cortical defect was highly probable in this case, extensive bone production particularly in the left femoral lesion led to the misinterpretation of the lesion as fibrous dysplasia. The pattern of ossification, with the rim of osteoblasts surrounding the newly formed bone trabeculae, as well as its tendency toward maturation were reminiscent of the zonal phenomenon characterizing myositis ossificans. Osteoblastic cells presented no signs of atypia and the few mitosis found were normal.

Involvement of two adjacent bones, usually in the vertebral column or in a vertebra and the corresponding rib is considered an important diagnostic clue for ABC and is probably the result of direct extension of the lesion [3, 10, 19]. Cases of bilateral development of classic ABC are extremely rare, with only two examples reported in the world literature, one with bilateral involvement of the maxilla [15] and the other with bilateral, symmetrical involvement of humerus and ulnar [21]. The case we re-

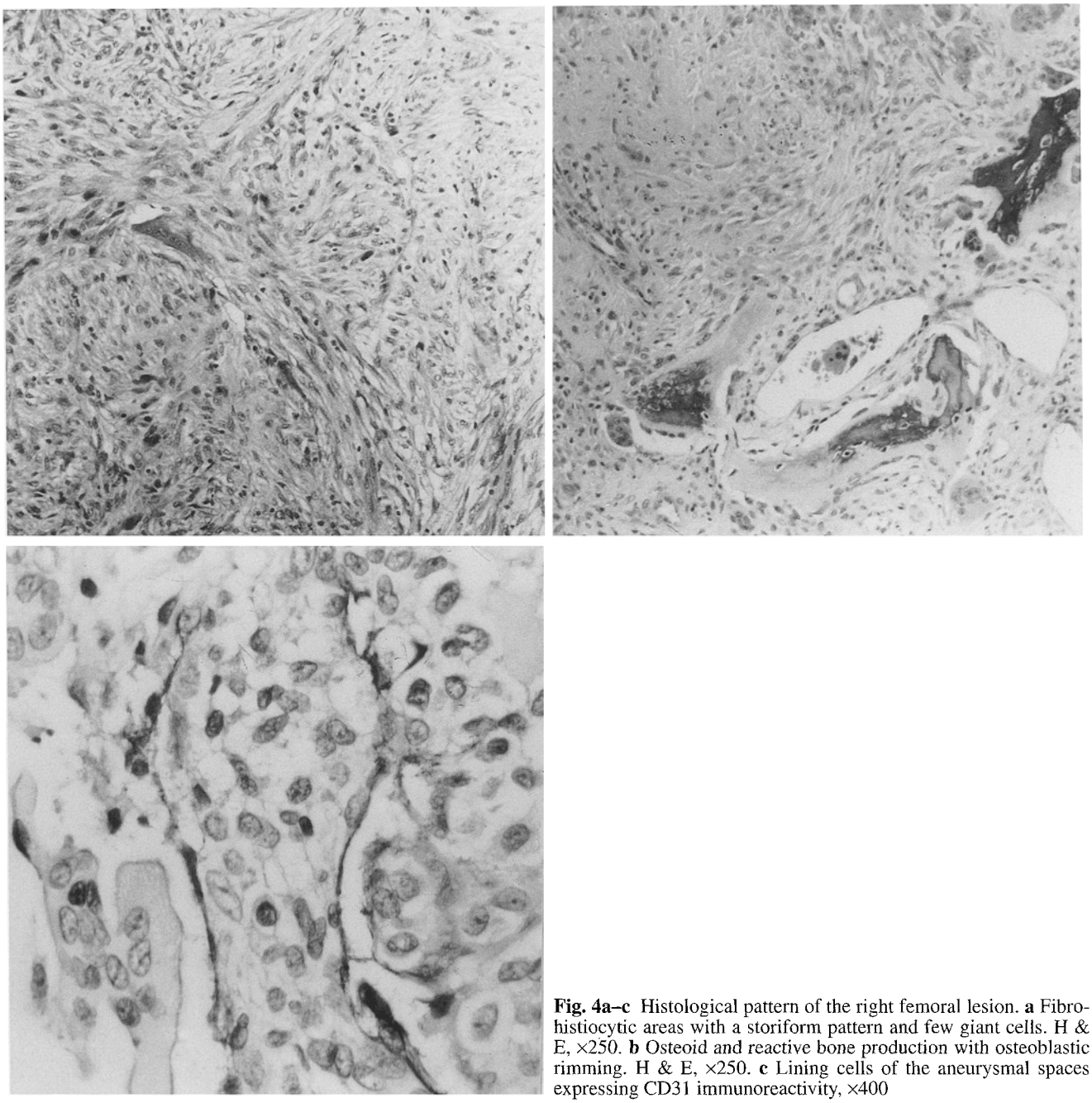

Fig. 4a–c Histological pattern of the right femoral lesion. **a** Fibrohistiocytic areas with a storiform pattern and few giant cells. H & E, ×250. **b** Osteoid and reactive bone production with osteoblastic rimming. H & E, ×250. **c** Lining cells of the aneurysmal spaces expressing CD31 immunoreactivity, ×400

port is interesting not only as an example of the rare solid variant of ABC but also as a case with lesions in two independent long tubular bones. Despite the patient's age, (initial lesion discovered at the age of 13 years) bilaterality in association with giant cell bone lesions makes the differentiation from lesions of hyperparathyroidism indispensable. Normal biochemical and hormonal blood tests, and the absence of any other bone lesions did not supported this diagnosis.

Although the entity designated by the term solid variant of ABC has gained acceptance by bone tumour oriented pathologists, most of them have some difficulties in adopting the term "solid" as an oxymoron [18]. Since both cystic and solid variants of ABC seem to be related lesions, the term solid, though sounding paradoxical should be preserved until the pathogenesis of the lesion is resolved [17, 18].

The preponderance of the cystic variant of ABC [1, 19] and the existence of extraosseous lesions with the histology of ABC in association with vascular abnormalities [13] support the view that the initial lesion might be some kind of vascular anomaly, resulting in the development of aneurysmal spaces [2, 4, 6, 8, 9]. The objection raised by Vollmer et al. [20] concerning the vascular nature of ABC seems not to be justified in our case, since many strongly positive CD31 immunoreacting endothelial cells were found lining the few aneurysmal spaces.

Spontaneous healing of ABC has already been docu-

mented radiologically [11]. However, the fibroblastic and myofibroblastic cell proliferation accompanied by an osteoclastic and osteoblastic reaction, resembles giant cell reparative lesions or reactive heterotopic ossification (myositis ossificans). The development of the solid variant of ABC might be attributed to an excessive reparative or healing process.

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