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Aggressive Giant Extraskeletal Aneurysmal Bone Cyst of the Thigh: Overcoming Challenges with a Multidisciplinary Approach

Quisto Ósseo Aneurismático Extraesquelético Agressivo da Coxa: Ultrapassando Desafios com uma Abordagem Multidisciplinar

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ABSTRACT

Aneurysmal bone cysts are vascular benign fibroblastic lesions usually found in bone that are locally destructive, with a greater incidence in the first and second decades of life. Patients usually undergo curettage or, less frequently, surgical resection, which may lead to growth disturbances and deformities in cases of large or complex lesions. Minimally invasive techniques such as sclerotherapy and endovascular embolization have been developed as an alternative or complement to surgery, with promising results. The authors present a rare case of an extraskeletal aneurysmal bone cyst successfully treated with minimally invasive techniques followed by surgical resection and provide a literature review of the current treatment options.
Keywords: Bone Cysts, Aneurysmal; Embolization, Therapeutic; Radiology, Interventional; Sclerotherapy

RESUMO

O quisto ósseo aneurismático é uma lesão benigna, de origem vascular fibroblástica, localmente destrutiva, que se forma tipicamente no osso, em indivíduos nas primeira e segunda décadas de vida. As lesões são tipicamente submetidas a curetagem ou, menos frequente, ressecção cirúrgica, que apresenta risco de complicações como deformidades ósseas ou distúrbios de crescimento, sobretudo em lesões complexas ou volumosas. Técnicas minimamente invasivas como a escleroterapia ou a embolização arterial têm sido desenvolvidas como alternativa ou complemento à cirurgia, com resultados promissores. Os autores apresentam um caso de quisto ósseo aneurismático extraesquelético tratado com sucesso através da combinação de técnicas minimamente invasivas e seguido de ressecção cirúrgica, e revisão da literatura a respeito dos tratamentos atualmente disponíveis.
Palavras-chave: Embolização Terapêutica; Escleroterapia; Quisto Ósseo Aneurismático; Radiologia de Intervenção

INTRODUCTION

Aneurysmal bone cyst (ABC) is a benign fibroblastic tumor that is usually found in bone, with locally aggressive potential due to expansion and destructive nature towards surrounding tissue.[1] It usually affects patients below the age of 20, and 70% of cases are of primary nature. The remaining 30% are associated with other tumors.

In the past, treatment consisted of open resection, which has the potential for 0% recurrence but high morbidity.[2] There has been a current shift to less invasive procedures to treat ABC in order to reduce morbidity and growth disturbance. Curettage and bone grafting remain the gold standard for managing ABC,[3] but several minimally invasive tools have shown efficacy and safety, such as sclerotherapy, radionuclide ablation and endovascular embolization.[4]

In this report, the authors discuss the role of interventional radiology in conjunction with orthopedic surgery in treating a challenging case of peripheral extraskeletal ABC.

CASE REPORT

A 28-year-old Nepalese male patient with a one-month history of progressive posterior left thigh pain and claudication was referred to an Orthopedic surgery clinic. Prior medical history was unremarkable.

On physical examination, a palpable mass was detected in the posterior thigh, as well as range-of-motion limitation performing left knee flexion. There was no evidence of neurovascular compromise.

Contrast-enhanced computed tomography (CT) was performed, which revealed a giant soft tissue mass located in the deep layer of the left posterior compartment of the thigh, with peripheral calcifications and multiple cysts of variable size (Fig. 1). The cyst-like mass had a total volume of 569 cc and was moderately vascularized from several branches of the deep femoral artery, as depicted on CT angiography. No soft-tissue mass component was identified within the multiple cysts besides the multiple septa, but extensive periosteal reaction was evidenced at the proximity with the posterior cortex of the femur.

Magnetic resonance imaging (MRI) was performed for further assessment, confirming a cystic mass with multiple cysts with fluid-fluid levels (Figs. 2A and B), exerting significant mass effect and displacing the surrounding soft tissue structures, including the sciatic nerve. In a 45-day span between CT and MRI assessments, the mass had nearly

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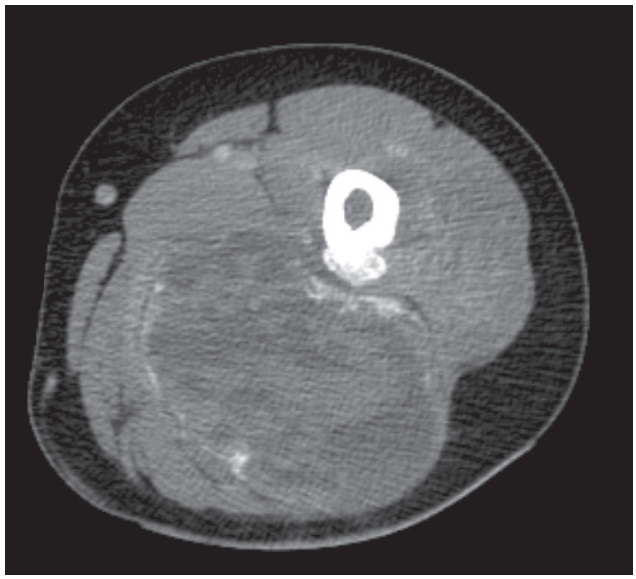

Figure 1 – Computed tomography of the thigh reveals a soft-tissue mass located in the posterior thigh, with thin rim calcifications (axial plane)

doubled in size (936 cc), and the patient had developed increased pain and claudication with the need for a walking aid.

An ultrasound-guided biopsy was performed in-between CT and MRI imaging, with soft-tissue extraction of the cyst septa and fluid aspiration of different cysts for cytologic evaluation. Pathology results heralded findings suggestive of ABC with no signs of underlying malignant tissue. After MRI evaluation, a second biopsy was performed, targeting the soft tissue adjacent to the posterior femoral periosteal reaction, with a similar result.

A multidisciplinary decision-making process opted for minimally invasive treatments to halt progression, induce size reduction, and serve as a bridge for subsequent surgical removal if technically feasible.

The patient underwent arterial embolization, which was achieved with a femoral contra-lateral approach, superselective catheterization of the feeding vessels from the deep femoral artery and embolization with polyvinyl alcohol particles (355 - 500 µm) until stasis was obtained (Fig. 3). An intra-procedural arterial CT was performed which revealed further increase of the mass to 1256 cc.

A follow-up CT one-month post-embolization documented stability of the ABC size and progression to peripheral bone mineralization Combined therapy was proposed, and two sessions of fluid aspiration and subsequent percutaneous polidocanol injection on the largest cysts were performed, each session focusing on a different segment of the mass. Follow-up CT revealed a reduction in the size of the treated cysts and further mineralization, with a small reduction in global tumor size (1105 cc) (Fig. 4).

After discussion with the orthopedic surgery team, surgical removal was deemed feasible and proposed. A second session of pre-operative arterial embolization was done to reduce bleeding during surgery, and the ABC was successfully removed (Figs. 5A and B). Histological analysis revealed a multiloculated cystic lesion without epithelial lining, septs with macrophages and giant multinucleated cells of osteoclast type, bony trabeculae with immature bone and osteoblastic activity, as well as myofibroblast tissue proliferation. The findings were compatible with aneurysmal bone cyst.

Patient was referred for physical rehabilitation. A follow-up MRI one month later documented a large seroma at the

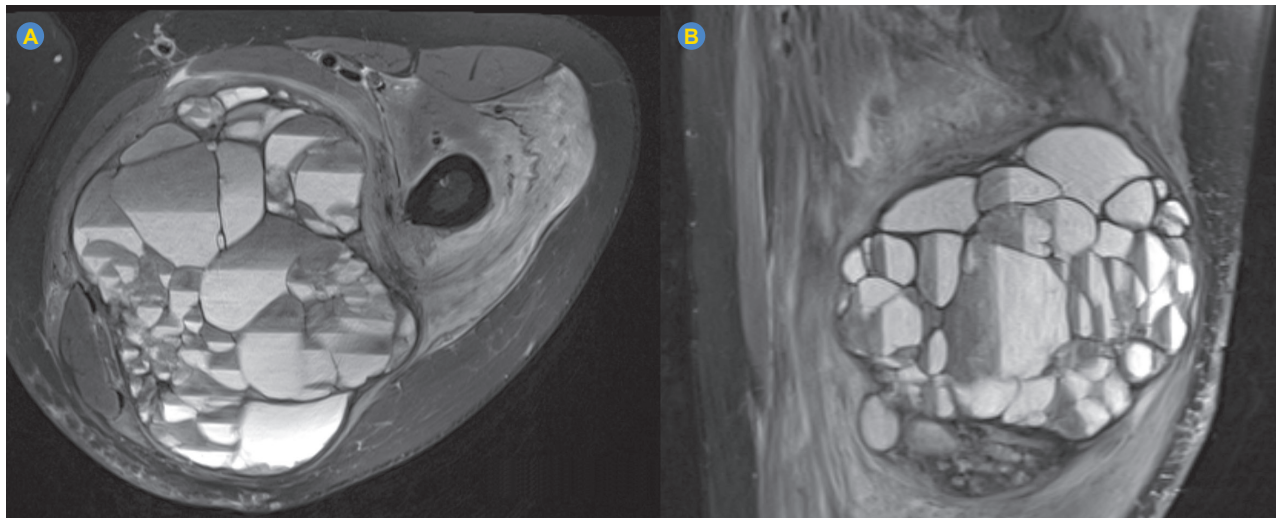

Figure 2 – Magnetic resonance imaging of the thigh revealing a multiloculated soft-tissue mass with fluid-fluid levels [axial (A) and sagittal (B) fat-saturated proton density sequences]

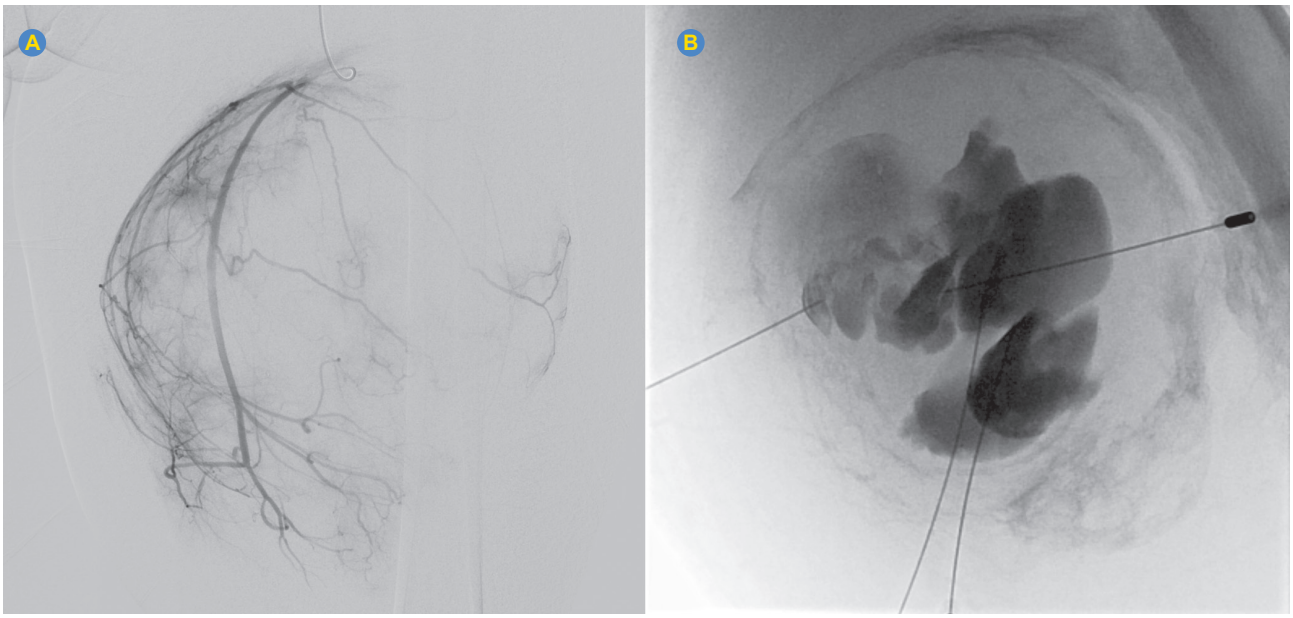

Figure 3 – Arterial embolization of the tumoral vascular supply arising from branches of the deep femoral artery (anteroposterior fluoroscopic image) (A). Percutaneous sclerotherapy using four 22G needles placed in different internal loci of the ABC (B).

resection site and clinical evaluation showed progressive improvement of mobility and pain reduction (Fig. 6). At the time of publication, patient is continuing physical rehabilitation and follow-up by the orthopedic oncology team.

DISCUSSION

Aneurysmal bone cysts are rare expansile benign skeletal tumors that affect the bone during the growth period, with an incidence of 0.14 per 100 000 persons, accounting for 1% to 2% of all primary bone tumors.[5] Pathogenesis is associated with dysplastic vessels and neoplastic proliferation, and 80% occur before the third decade of life. In 30% of cases, an underlying tumor is present, and these are considered secondary ABC.[6]

They most commonly present as an expanding mass with a cyst-like appearance inside the bone. They are composed of multiple blood-filled cysts separated by fibrous septa containing several cell types, including giant cells with or without osteoblasts.

Common complications include pain, local edema or tumefaction, neurological compromise, movement restriction and pathologic fracture.[7]

Extraskeletal ABC are exceedingly rare; there have been anecdotal reports in the literature,[8] and they may mimic a variety of other benign and malignant tumors, such as extraskeletal (telangiectatic) osteosarcoma, soft-tissue giant-cell tumor, tenosynovial giant cell tumor, brown tumor, and myositis ossificans.

Pathogenesis is unknown but it is hypothesized that they may be associated with traumatic events or vascular changes with a pathologic reparative process underlying cystic degeneration and neoplastic formation.[8]

Although conventional x-ray is usually the first diagnostic tool employed, further imaging evaluation with CT and/

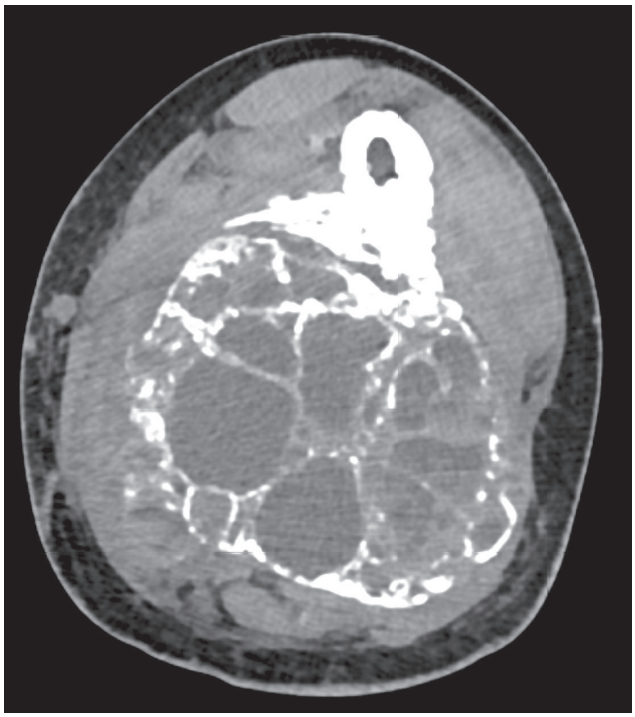

Figure 4 – Follow-up CT post-minimally invasive procedures reveals stabilization of mass size and progressive calcification and reduction of cyst volume

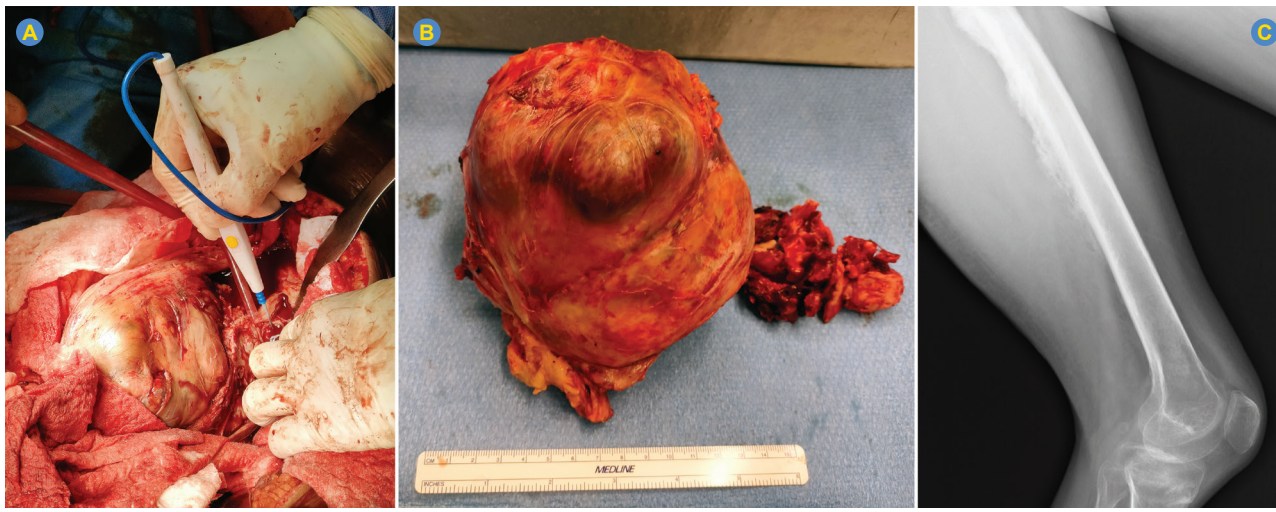

Figure 5 – Intra-operative image during surgical resection of the extraskeletal ABC (A). Surgical specimen (B). Post-surgical x-ray of the thigh, lateral incidence (C).

or MRI is fundamental for a more detailed assessment with regards to size, shape, presence of soft-tissue component, cyst size and number, presence of fluid-fluid levels, grade of mineralization and bone stock, relationship with the underlying bone or soft-tissues, and vascularity.[9,10]

Treatment options include surgical techniques, percutaneous injection or endovascular embolization. Comparative studies are missing and, currently, there is no consensus regarding the preferred type of management.[11]

The gold standard of surgical treatment is curettage with or without mechanical burring, bone grafting, cauterization, argon beam coagulation, polymethyl methacrylate (PMMA), hydrogen peroxide and internal fixation.[2] With curettage, the most commonly employed technique (with or without adjuvants), failure rates range from 0% to 40%. Supplemental use of adjuvants has been shown to decrease recurrence rates.[6] En block resection may also be employed, especially in more aggressive lesions since it has lower recurrence rates but bears greater morbidity.

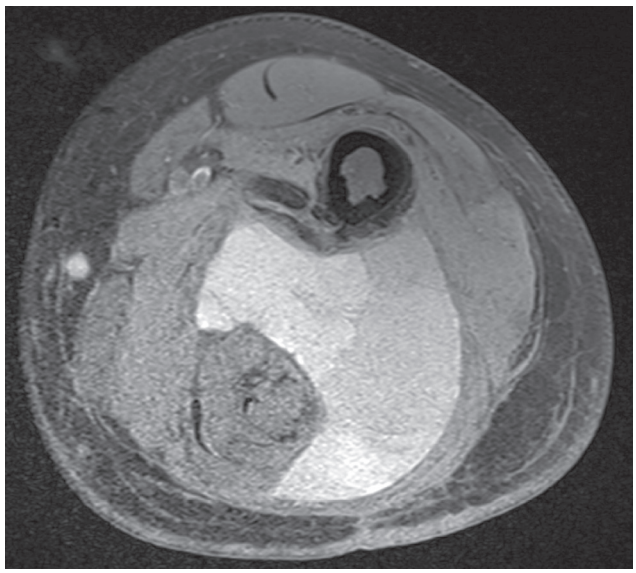

Figure 6 – Post-operative MRI after surgical resection reveals large posterior thigh seroma (axial fat-saturated proton density sequence)

Intralesional curettage with filling up of the remaining cavity with bone substitutes is generally performed in lesions to prevent pathological fractures and in lower-limb lesions with weight-bearing pain. High-speed mechanical burring is used to increase cavity size after curettage. Cauterization, argon beam coagulation and hydrogen peroxide extend the zone of necrosis and eradicate marginal tumor remnants, thus serving as adjuvant therapies.

Growth disturbance and deformity are the most common complications related to surgical treatment groups described in the literature.

Minimally invasive techniques have been increasingly employed for management of ABC, as reported in the literature.[4] Such techniques include intralesional injection of sclerosing agents and arterial embolization.

Sclerotherapy of ABC is an alternative procedure that may act as an adjuvant treatment or bridge to other therapeutic strategies. The radiological efficacy of different sclerosing agents has not been compared, and options include polidocanol, ethibloc, absolute alcohol, calcitonin and steroids, calcium sulfate, doxycycline or a combination of these agents.

Multiple treatments are often needed with injection therapy, reportedly between 1.1 to 6.4,[2] and a failure to heal or recurrence rate of 14.7% has been reported.

The most common complications regarding the most

frequently used agents include induration and hypopigmentation at the size of injection regarding polidocanol and inflammatory reaction with ethibloc.[4]

One single-center randomized controlled trial has compared injection-based therapy and surgical treatment and was not sufficiently powered to detect significant differences with healing as a primary outcome measure.[12] However, surgery was found to result in poorer functional outcomes and a higher number of complications. To the best of the authors' knowledge, there are no other studies comparing surgical and minimally invasive techniques.

Transarterial embolization is another viable option, and can be achieved with coils, polyvinyl alcohol particles or n-butil-cyanoacrilate, among other agents, and has been used for the treatment of ABC.[13] Indeed, it has been shown to halt growth, promote mineralization, and cause pain palliation in several bone tumors.[14] Pre-operative embolization has also been shown to decrease bone loss and transfusion requirements during surgery.

Denosumab is a monoclonal antibody that induces mineralization and has been reported as a useful treatment for giant cell tumors of bone. Some reports have shown efficacy in other fibro-osseous lesions, such as spinal and peripheral ABC, with promising results for local tumor control.[15]

Taking into account the pathogenesis underlying ABC, targeting the dysplastic vessels with endovascular occlusion and filling the cysts with substances that may locally destroy neoplastic cells may produce a synergistic effect. Such combined therapies have the potential to eradicate neoplastic tissue and promote bone remineralization, especially in aggressive, refractory, or unresectable tumors.[10]

The collaboration between the orthopedic oncology and interventional radiology teams at our hospital center has been ongoing for several years, with the intent of providing the best treatments for complex cases such as the one presented in this article. The authors have had good results and feedback with minimally invasive treatments for several complex ABC, successfully reducing or halting tumor growth and promoting sclerosis, which in turn leads to clinical improvement and precludes surgery.

In this case, the choice of performing minimally invasive treatments was not to preclude surgery but to effectively promote sclerosis and fibrosis, decrease tumor size and vascularity in order to facilitate surgical resection, and reduce the risk of complications.

CONCLUSION

Minimally invasive treatment with combined percutaneous and endovascular techniques has been proven to be safe and effective as an alternative or downstaging/adjuvant therapy to facilitate surgery, with significant symptom and quality of life improvement.

Although curettage is the gold standard treatment for ABC, these are often complex lesions that require a distinct case-by-case approach. In light of this, several alternative and adjuvant therapies have been developed, which should be taken into consideration when they pose as available options. However, there are currently no treatment recommendations or guidelines, and further research is needed to compare the different techniques and come up with a rationale to develop adequate therapeutic algorithms.

AUTHOR CONTRIBUTIONS

APC: Study design, data acquisition, writing of the manuscript.

TN: Data acquisition, writing of the manuscript.

CP, FVG, JP, EC, TB: Study design, critical review of the manuscript.

All authors approved the final version to be published.

PROTECTION OF HUMANS AND ANIMALS

The authors declare that the procedures were followed according to the regulations established by the Clinical Research and Ethics Committee and to the Helsinki Declaration of the World Medical Association updated in 2013.

DATA CONFIDENTIALITY

The authors declare having followed the protocols in use at their working center regarding patients' data publication.

PATIENT CONSENT

Obtained.

COMPETING INTERESTS

The authors have declared that no competing interests exist.

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