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Case Report and Review of the Literature

Malignancy of Aneurysmal Bone Cyst in Osteosarcoma and Angiosarcoma: Series of 2 Cases and Literature Review

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ABSTRACT

Aneurysmal bone cyst (ABC) is a rare benign bone tumor characteristic of the first decades of life. Malignization of ABC in the absence of risk factors such as radiotherapy is extremely rare, with few cases published in the literature. There are malignant tumors such as telangiectatic osteosarcoma with an initial clinical, radiological and anatomopathological resemblance that requires special attention in their differential diagnosis. We present two patients with an initial histologic diagnosis of ABC who developed true malignancy, in the absence of risk factors, into osteoblastic osteosarcoma and epithelioid angiosarcoma. In spite of its exceptional nature, in the presence of an aggressive type of ABC malignant degeneration should always be suspected and therefore, an open biopsy and wide resection performed for a correct diagnosis to avoid residual microscopic disease in case of aggressiveness. Epithelioid angiosarcoma from an ABC tumor malignancy has never been described before.

Background

ABC can be considered as a primary or secondary lesion, the latter term being reserved for those which are coexisting with another lesion, either benign or malignant. The progression of an ABC to a malignant tumor is an extremely rare event, being somewhat more frequent after the application of radiotherapy within a primary tumor. Likewise, it is essential to be alert to the appearance of an aneurysmal bone cyst, since, although very infrequently, there is the possibility of being faced with a tumor of aggressive characteristics with similar initial clinical-radiological manifestations. Few cases of malignant tumors developed from ABC have been described in the literature. All of them demonstrating their benignity through biopsy.

Clinical Case 1

The first case is a 32-year-old male, followed up since childhood for an aneurysmal bone cyst in the right humerus, who suddenly began to experience intense pain, with a conventional radiographic examination showing the presence of a pathological fracture and radiological progression (Figure 1).

In view of the clinical and radiological worsening, it was decided to perform an open biopsy where the diagnosis of ABC was confirmed in the absence of malignant cells. Following this biopsy, the patient began to experience repeated intramuscular bleeding (Figure 2).

Given the torpid lesion evolution, it was decided to perform a wide resection including the soft tissue mass. A reconstruction with structural allograft and endomedullary humerus nailing fixation was performed subsequently (Figure 3).

The anatomopathological diagnosis was an aneurysmal bone cyst at bone level, coexisting with an infiltration of malignant cells in the perilesional musculature compatible with the diagnosis of epithelioid angiomyosarcoma.

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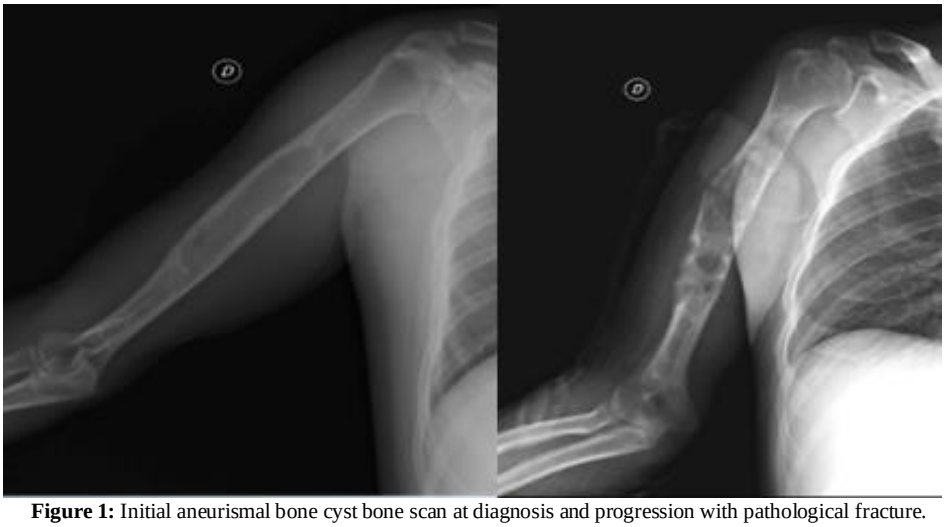

Figure 1: Initial aneurismal bone cyst bone scan at diagnosis and progression with pathological fracture.

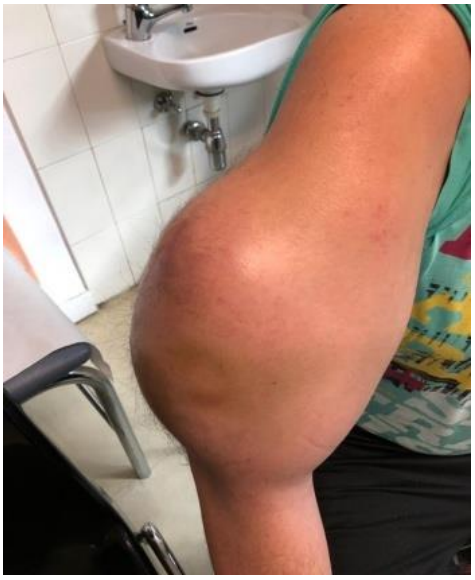

Figure 2: Intramuscular bleeding after open biopsy.

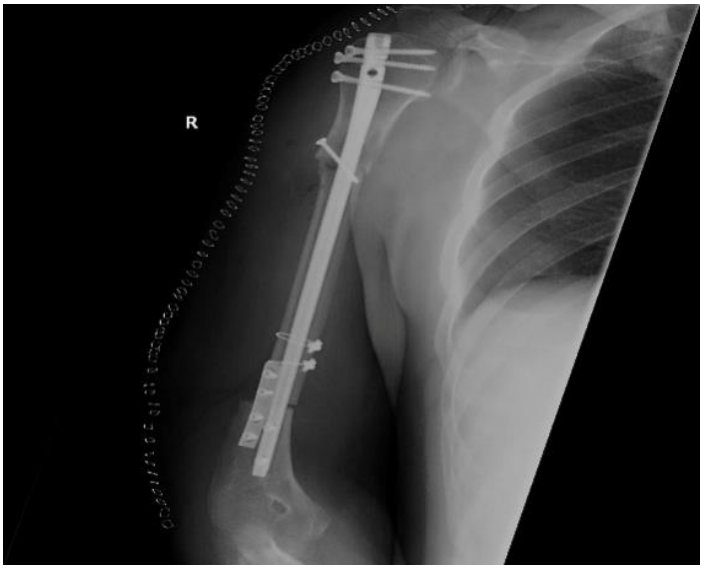

Figure 3: Structural allograft with endomedullary fixation after wide tumoral resection.

After discussing the case in the Tumor Committee, adjuvant chemotherapy and radiotherapy were decided. After treatment, the patient suffered a relapse at the proximal level, which required an interscapulothoracic amputation and intraoperative radiotherapy (Figure 4). To date, the patient has relapsed and is currently undergoing palliative treatment.

Figure 4: Interscapulothoracic amputation.

Clinical Case 2

The second case described is a 16-year-old male patient who, after a trauma and several days of pain, is diagnosed of rotator cuff tendinopathy in the absence of radiological control in the emergency department. After conventional radiological control, he is transferred for a suspicious lytic-blastic tumor at the level of the epiphyseal-metaphyseal proximal humerus (Figure 5). The patient is preferentially referred to the Musculoskeletal Tumors Unit, where a CT scan, MRI and open biopsy were urgently requested. The open biopsy pathological anatomy was finally reported as an aneurysmal bone cyst in the absence of malignant cells.

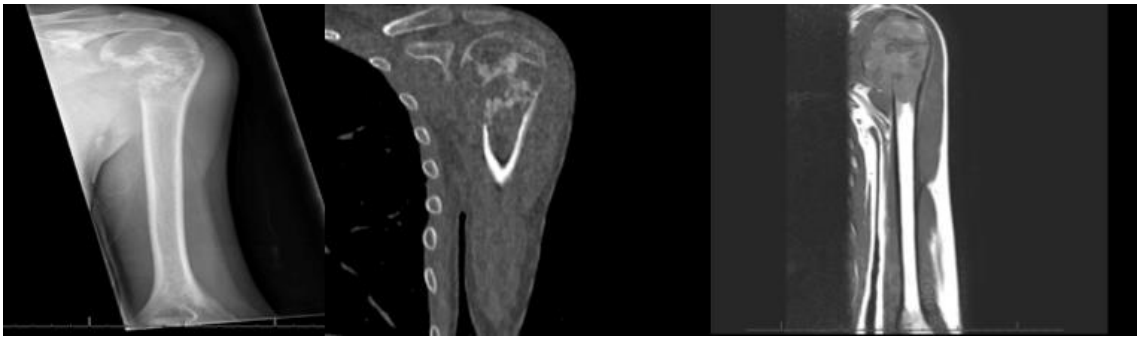

Figure 5: Lytic-blastic tumor.

Given the absence of proximal bone remnant, it was decided to perform as definitive treatment a marginal resection of an aneurysmal bone cyst of the proximal left humerus and partial tumor prosthesis reconstruction, where the pathological anatomy was finally reported as an aneurysmal bone cyst together with 10% malignant cells compatible with osteoblastic osteosarcoma (Figure 6).

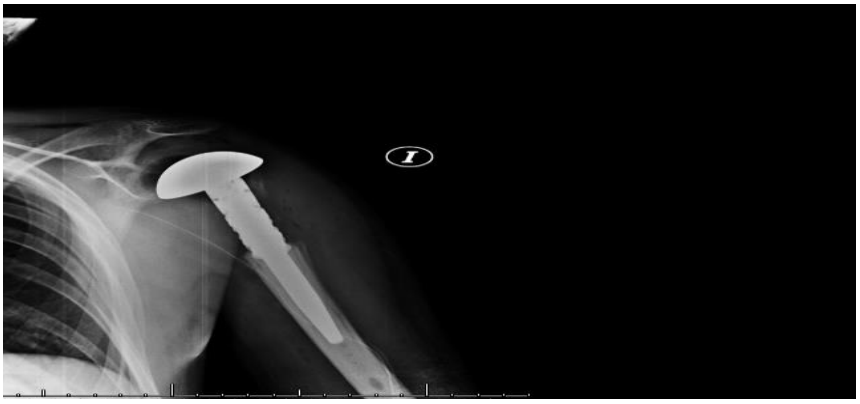

Figure 6: Marginal tumor resection with partial tumor prosthesis reconstruction.

After adjuvant chemotherapy and being disease free for 2 years, in January 2020 he began with discomfort in the left shoulder with pathological uptake in PET-CT, and an open biopsy was performed with the diagnosis of recurrence ABC and osteosarcoma at the level of the rotator cuff of 0.5 × 1cm. In view of the local recurrence, it was decided to perform a new wide resection including the prosthesis, rotator cuff and a large part of the deltoid and a new reconstruction with a tumor prosthesis. In the resection piece analysis, there was no evidence of osteosarcoma remains. Subsequently, 6 cycles of adjuvant chemotherapy were given.

Discussion

The aneurysmal bone cyst is a benign expansive osteolytic lesion, typical of the first three decades of life, characterized by the presence of blood-filled spaces at the macroscopic level and channels divided by connective tissue septa that may contain osteoid tissue and osteoclasts at the microscopic level. They are rare bone lesions with a prevalence of only 1-2% of all tumors [1, 2].

Despite the existence of several theories linking the origin of an aneurysmal bone cyst (ABC) to lesions such as an arteriovenous shunt or post-traumatic bleeding over a previous bone lesion, the pathogenesis of ABC remains unknown. Recent studies argue for the existence of chromosomal abnormalities within ABC, suggesting that they are true neoplasms [3, 4].

QOA is mainly located in long bones, especially in the proximal humerus, followed by the proximal femur; these two locations account for more than 80% of cases. Other less frequent anatomical regions are proximal tibia or diaphyseal, distal humerus and distal femur. Exceptionally, locations such as metacarpals, carpus, scapula or mandible have also been described [5, 6]. Very few cases in the literature support the existence of malign ABC tumors in the absence of risk factors such radiotherapy. There is an inversely proportional correlation between the risk of recurrence and the distance between healthy tissue neoplasia. In this context, a correct safety margin is of paramount importance in order to improve survival and prevent relapse [7].

Kyriakos M *et al*. described an 11-year-old female with a lytic lesion at the level of the distal tibia, which was diagnosed by curettage on two occasions as a ABC, 28 months later underwent malignant transformation into a pleomorphic osteosarcoma [8]. Anract *et al*. described the case of a 40-year-old man diagnosed with malignant fibrous histiocytoma 12 years after resection of a QOA on a pathological supracondylar fracture of the femur [1]. George W. Brindley *et al*. described the case of a 13-year-old male with a lytic metaphyseal lesion at the level of the proximal humerus, with radiological features of benignity and a diagnostic biopsy of an aneurysmal bone cyst. In 2001, he began with pain and radiological changes and was diagnosed by biopsy as telangiectatic osteosarcoma [9].

Conclusion

Aneurysmal bone cyst malignization is a rare event with important consequences in morbidity and mortality terms. In the presence of an aggressive ABC, secondary degeneration should be suspected and a percutaneous or open biopsy should be performed to rule out this possibility, although a negative biopsy does not completely exclude the diagnosis. In these cases it would be advisable to perform a wide resection of the lesion in order to avoid leaving microscopic disease which, in the case of a malignant lesion, would require radical surgery for local control of the disease. These cases confirm not only the malignant potential of these tumors, but also broaden the spectrum towards epithelioid angiosarcoma, not previously described in the literature.

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Competing Interests

None.

REFERENCES

1. Báez AB, Pino MÁL, Cuadrillero DA, Cerdá SS (2010) Quiste óseo aneurismático coexistente con osteosarcoma. Discusión radiopatológica. *Radiologia* 52: 247-250. [Crossref]
2. Sasaki H, Nagano S, Shimada H, Yokouchi M, Setoguchi T et al. (2017) Diagnosing and discriminating between primary and secondary aneurysmal bone cysts. *Oncol Lett* 13: 2290-2296. [Crossref]
3. van de Luijtgaarden ACM, Veth RPH, Slootweg PJ, Koster PMW, Kool LJS et al. (2009) Metastatic potential of an aneurysmal bone cyst. *Virchows Arch* 455: 455-459. [Crossref]
4. Li YJ, Zhang Y, Yin XX, Zhang WL, Tu CQ (2016) Osteosarcoma at the site of a previously treated simple bone cyst. *Transl Cancer Res* 5: 877-882.
5. Mascard E, Brouchet AG, Lambot K (2015) Bone cysts: unicameral and aneurysmal bone cyst. *Orthop Traumatol Surg Res* 101: S119-S127. [Crossref]
6. Yin JQ, Fu YW, Xie XB, Cheng XY, Yang XY et al. (2018) Telangiectatic osteosarcoma: Outcome analyses and a diagnostic model

for differentiation from aneurysmal bone cyst. *J Bone Oncol* 11: 10-16. [Crossref]

7. He F, Zhang W, Shen Y, Yu P, Bao Q et al. (2016) Effects of resection margins on local recurrence of osteosarcoma in extremity and pelvis: Systematic review and meta-analysis. *Int J Surg* 36: 283-292. [Crossref]
8. Kyriakos M, Hardy D (1991) Malignant transformation of aneurysmal bone cyst, with an analysis of the literature. *Cancer* 68: 1770-1780. [Crossref]
9. Brindley GW, Greene Jr JF, Frankel LS (2005) Case reports: malignant transformation of aneurysmal bone cysts. *Clin Orthop Relat Res* 438: 282-287. [Crossref]