Aggressive Aneurysmal Bone Cyst of the Spine. Case report and Literature Review

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ABSTRACT

Introduction: An aneurysmal bone cyst is an expansive, lytic, pseudotumoral lesion that consists of blood-filled spaces separated by septa. It represents 15% of primary spinal tumors. An aggressive presentation is even rarer. Different therapeutic options are described depending on its stage and recurrence rate. We present the case of a patient with neurological involvement due to the aggressive behavior of an aneurysmal bone cyst in the thoracic spine, which required surgical treatment. **Conclusion:** Treatment options for aneurysmal bone cysts must adapt to each case, depending on its characteristics.

Keywords: Aneurysmal bone cyst; thoracic spine; myelopathy.
Level of Evidence: IV

Quiste óseo aneurismático vertebral agresivo: presentación de un caso y revisión bibliográfica

RESUMEN

Introducción: El quiste óseo aneurismático es una lesión pseudotumoral lítica, expansiva, compuesta por espacios llenos de sangre separados por tabiques. En columna representa el 15% de los tumores primarios, y su presentación de comportamiento agresivo es aún más infrecuente. Se han descrito diferentes opciones terapéuticas en función de su estadío y tasa de recurrencia. Presentamos el caso de un paciente con compromiso neurológico por la presencia de un quiste óseo aneurismático en columna torácica, de comportamiento agresivo, que requirió resolución quirúrgica. **Conclusión:** Las opciones de tratamiento del quiste óseo aneurismático se deben adecuar a cada caso en particular, según sus características.

Palabras clave: Quiste óseo aneurismático; columna torácica; mielopatía.
Nivel de Evidencia: IV

INTRODUCTION

The aneurysmal bone cyst (ABC) was first described by Jaffe and Lichtenstein in 1942 as an intraosseous and osteolytic lesion, differentiating it from hemangiomas and other giant cell tumors.[1] It is an expansive, lytic pseudotumoral bone lesion composed of blood-filled spaces separated by connective tissue septa formed by reactive bone tissue, fibroblasts, and osteoclast-type giant cells.[2]

ABC is a benign tumor and represents between 1% and 1.4% of all primary bone tumors.[3] Most cases have been identified in the first two decades of life; it is rare to find them after the age of 30. In the spine, they represent 15% of primary tumors.[4]

Their behavior can vary in the different stages, from latent to aggressive, as described by Enneking,[5] in such a way that its clinical presentation can involve anything from an imaging finding to compressive symptoms such as pain, neurological deficit, spinal instability or deformity. [6]

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Complementary studies reveal pathognomonic images of fluid-fluid levels. The definitive diagnosis is obtained by biopsy. Different therapeutic options have been described according to their behavior, location and recurrence rate.

Because there are few reported aggressive case reports of spinal ABC, diagnostic and treatment algorithms remain controversial and variable. Treatments have ranged from simple curettage, with or without bone graft, injection of a fibrosing agent, complete resection surgery, radiotherapy and selective arterial embolization to a combination of these methods.[7]

Although treatment outcomes are usually good, local recurrence is described in all therapy protocols, which cannot be accurately predicted, since in the published literature these rates are highly variable, even reaching figures of up to 25%.[8]

We present the case of a patient with a diagnosis of spinal ABC with aggressive behavior, with symptoms of spinal cord injury, treated by surgical approach.

CLINICAL CASE

A 32-year-old male patient, with no relevant clinical or surgical history, consulted at our institution due to back pain of 1 month's evolution. He did not report any history of trauma.

The medical history and physical examination revealed hypoesthesia in both feet, paresthesia, and Achilles clonus in the lower limbs. His strength was decreased, 4/5 according to the MRC (Medical Research Council) scale of muscle strength.[9]

On admission, imaging studies were requested. The radiograph showed the winking owl sign at the level of the tenth thoracic vertebra (T10) (Figure 1).

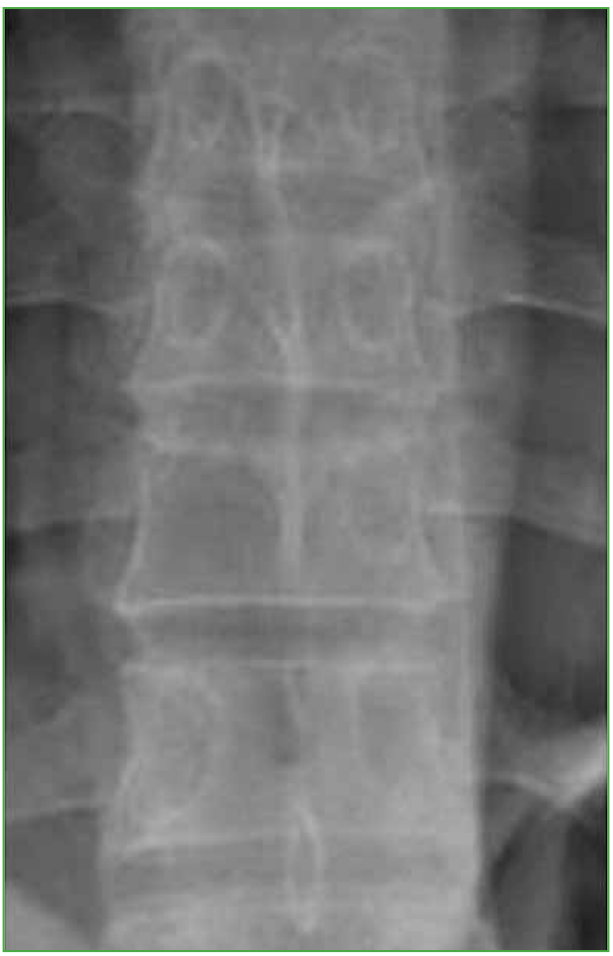

Figure 1. Anteroposterior dorsal spine radiograph. Lytic lesion centered on the right pedicle of the T10 vertebra. Winking owl sign.

The computed tomography showed an expansive lytic image, with disruption of the cortex, which compromised the pedicle, transverse process, posterior arch (predominantly on the right), and body of T10 (<50%) (Figure 2).

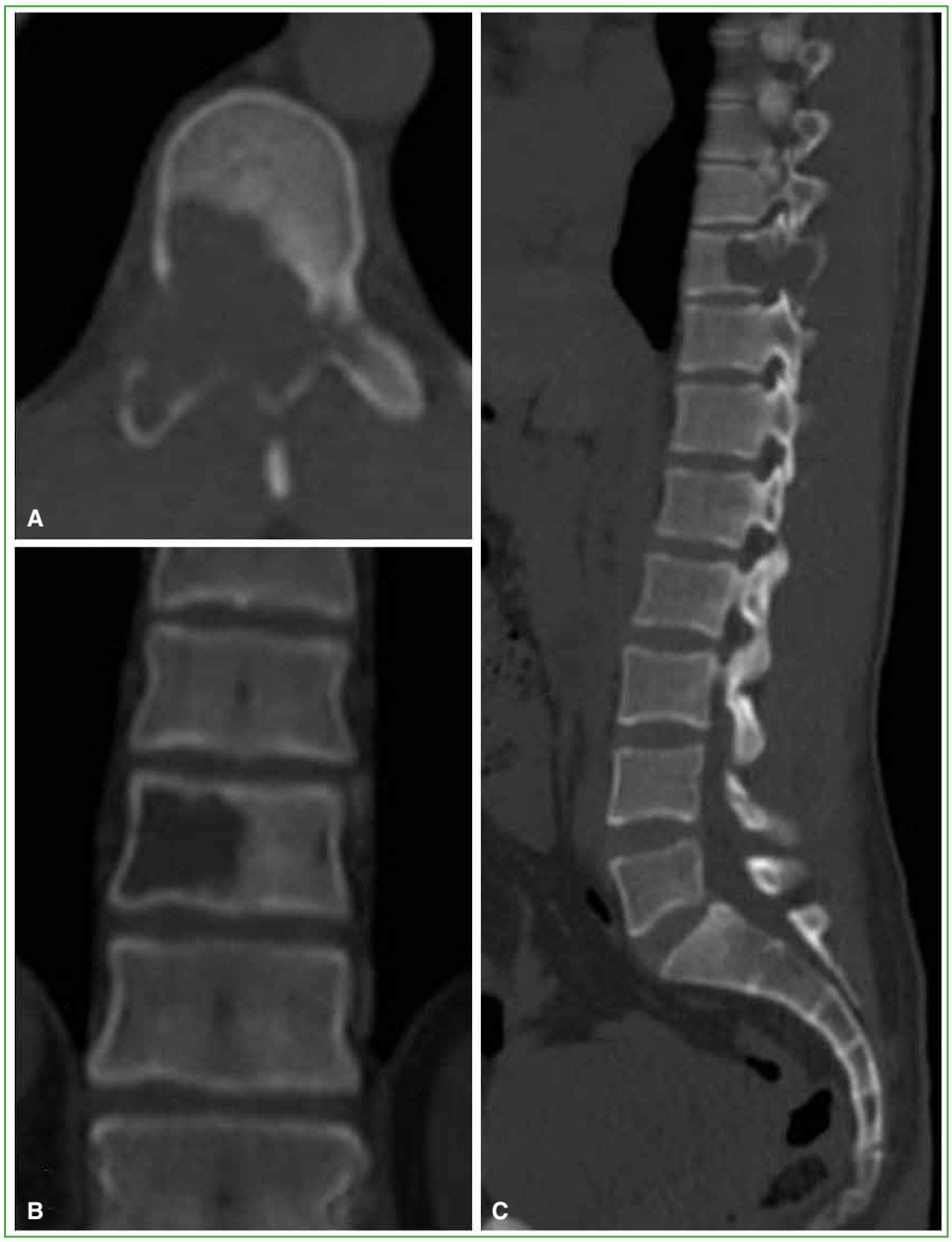

Figure 2. CT scan of the lumbosacral spine. **A.** Axial section. **B.** Coronal section. **C.** Sagittal section. A lytic, expansive image, with cortical thinning and interrupted edges compromising the pedicle, right transverse process, posterior arch, and vertebral body.

In the thoracic spine MRI, the same expansive lesion was identified, with liquid content and extension to the thoracic canal, which generated spinal cord compression and stenosis of the canal, obstructing the passage of cerebrospinal fluid (CSF) (Figure 3).

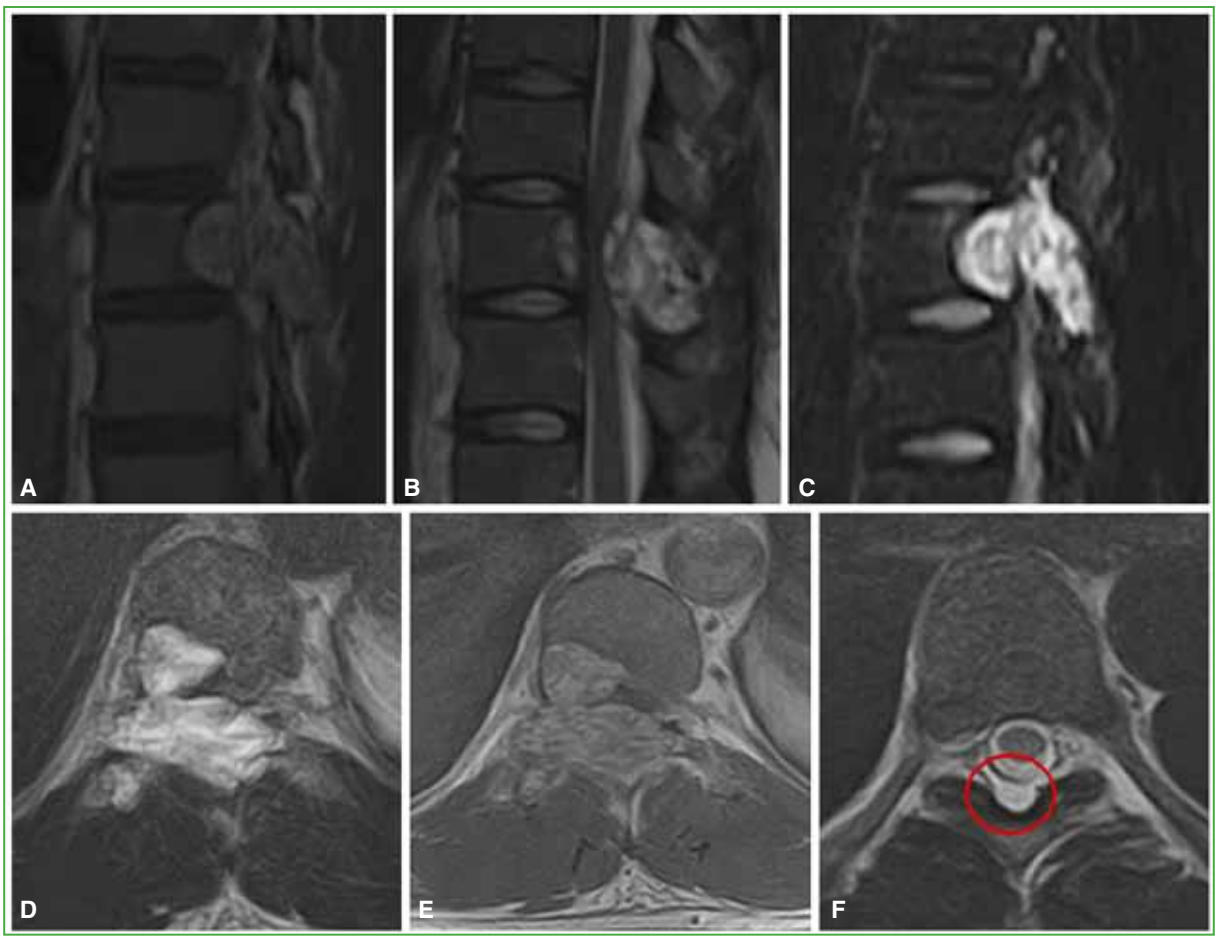

Figure 3. Thoracic spine MRI. **A.** T1, sagittal section. **B.** T2, sagittal section. **C.** STIR, sagittal section. **D.** T2, axial section. **E.** Axial section, with contrast. Expansive lesion with fluid signal intensity (hyperintense on T2 and STIR sequences and hypointense on T1 sequences), with septa inside. Complete stenosis of the medullary canal without passage of CSF. **F.** Epidural tumor cuff.

These sequences evidenced the compromise of somatosensory pathways in the lower thoracic segment/thoracolumbar junction.

A CT-guided punch biopsy was performed through a right T10 pedicle access; 3 cm^3 of hematic material for the cytological study and a bone plug fragment for the histological study were obtained. There were no complications during the procedure. In the histological sections of all the samples, fibrous septa with reticular bone, oval and spindle-shaped mononuclear cells without atypia, and osteoclast-like giant cells were observed, which led to the final diagnosis of ABC (Figure 4).

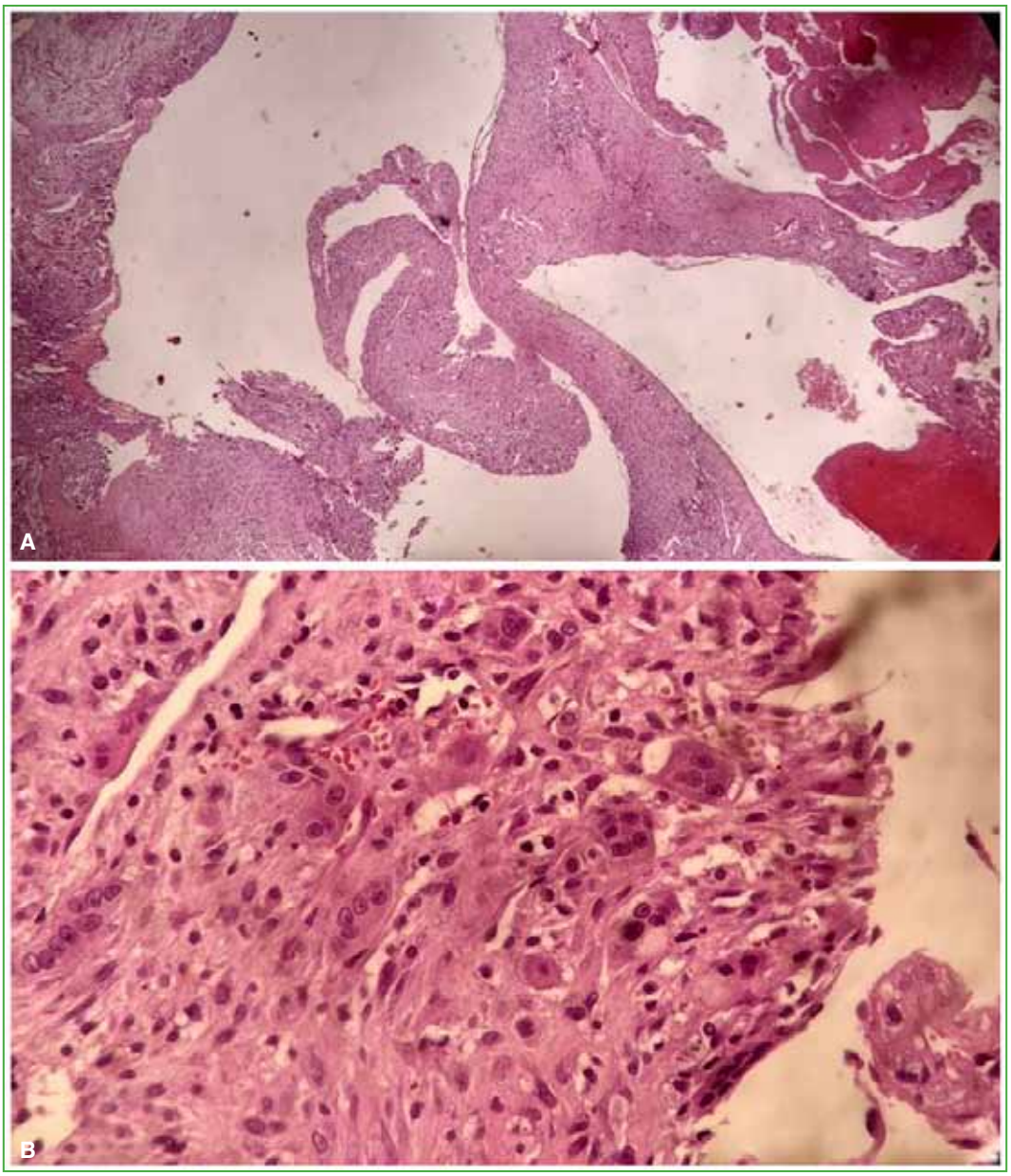

Figure 4. Pathological anatomy analysis. **A.** 40x. **B.** 100x. Histological sections show fibrous septa with trabecular bone, oval and spindle-shaped mononuclear cells without atypia, and osteoclast-like giant cells. These morphological findings are linked to an aneurysmal bone cyst.

Based on the patient's diagnosis and symptoms, it was decided to perform a surgical procedure. A digital spinal cord angiography was performed under general anesthesia (Figure 5), which showed right T10-dependent hypervascularization. In the same stage, endovascular embolization treatment with embospheres was performed.

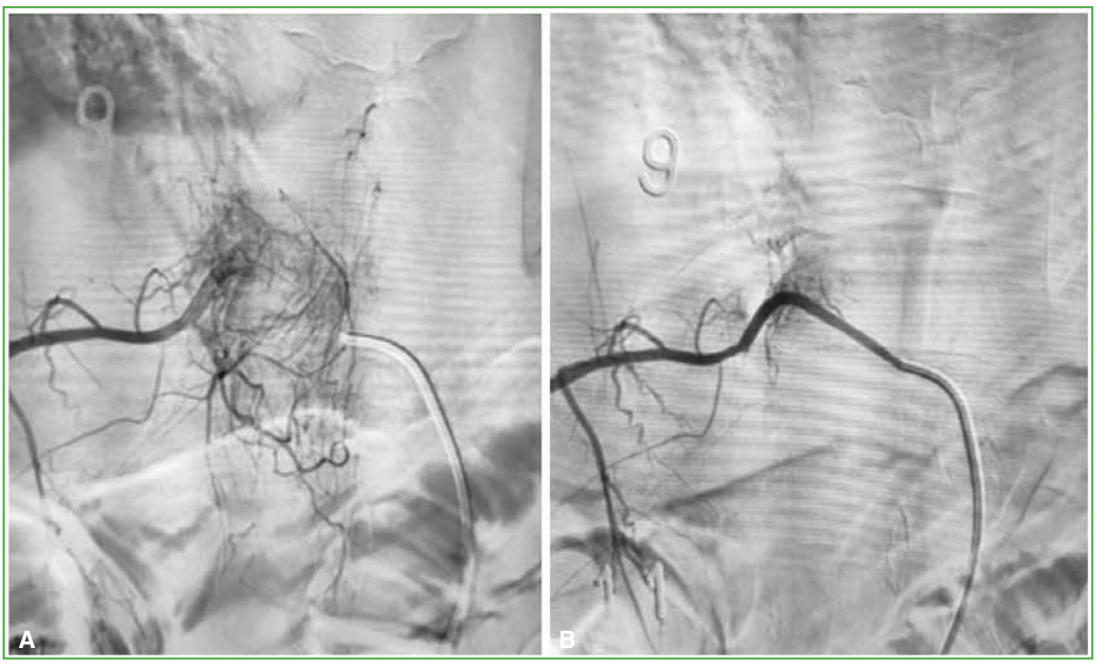

Figure 5. Digital spinal angiography. **A.** T10 vertebra preembolization. **B.** T10 vertebra postembolization.

24 h later, surgery was carried out, which consisted of extended excision of the posterior arch of T10. Extensive curettage, reaming, and phenolization of the cavity were performed. An embolized epidural tumor cuff was found, which was resected (Figure 6).

Figure 6. Wide decompression of the posterior arch of T10. T10 right pedicle emptying. Curettage, reaming and alcoholization of the residual cavity.

Finally, it was stabilized by implanting pedicle screws and rods from T9 to T11 (Figure 7).

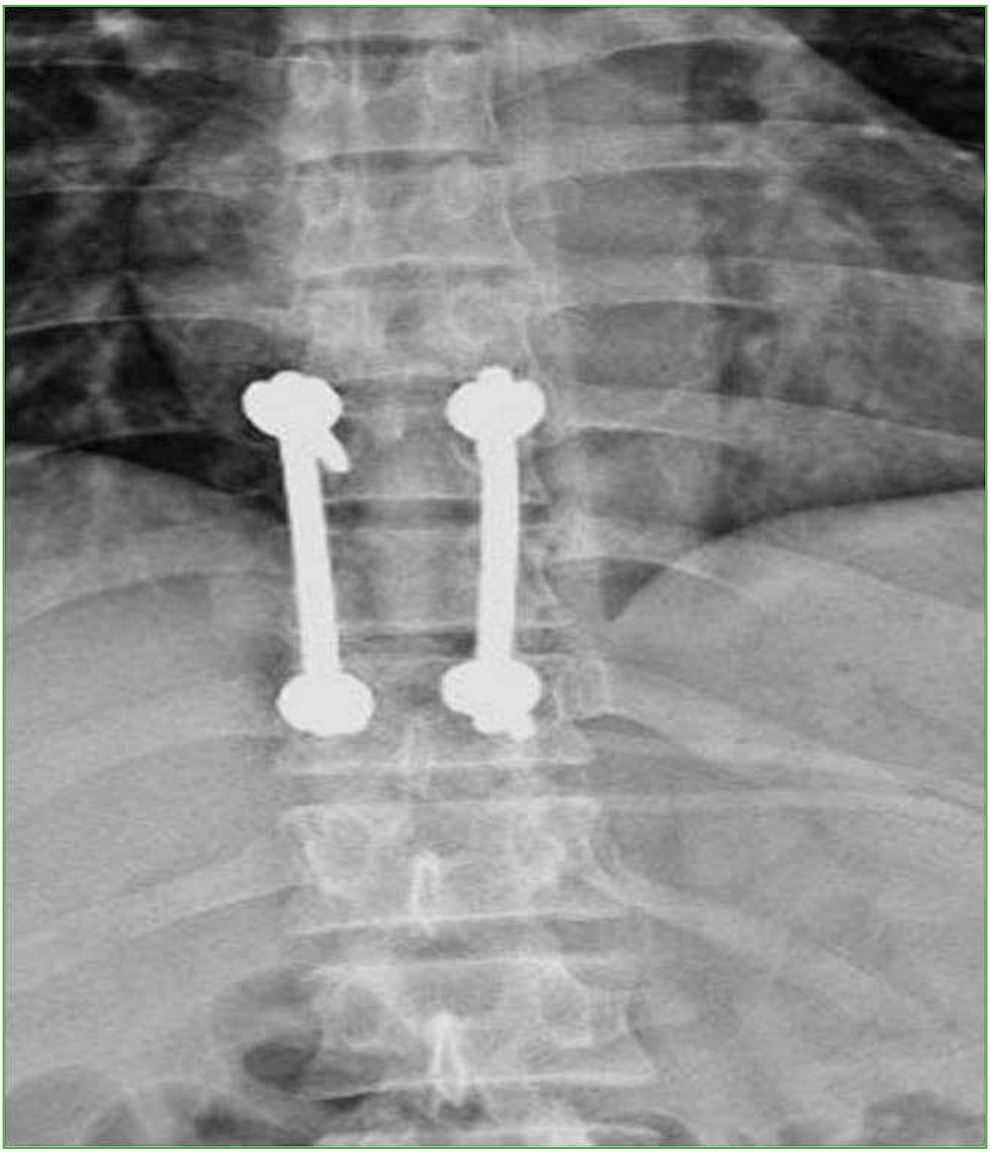

Figure 7. Anteroposterior dorsal spine radiograph in the postoperative period. Transpedicular instrumentation of T9-T11.

With good evolution and no progression of the neurological deficit, the patient was discharged 4 days later. Six months after surgery, the patient shows unrestricted ambulation and improvement in muscle strength, with no progression of neurological deficit on clinical follow-up examination.

DISCUSSION

ABC is included within the group of benign primary bone tumors. Although its histology defines it as such, ABC can present behavior that leads to rapid growth, with local destruction. It is a highly vascularized lesion composed of cavities of blood content, separated by connective tissue septa, and is surrounded by a layer of cortical bone, with the potential ability to expand.[10]

Common sites of occurrence are the femur, tibia, humerus, spine, pelvis, mandible, clavicle, ribs, and hand and foot bones. In the spine, the cervical region is compromised in 30-40% of cases, the thoracic spine in 25-50% and the lumbar segment in 40-45%.[11] It generally originates on the posterior elements; the vertebral body is a site with less frequent involvement.

The cause of ABC has not been discovered to date, although different hypotheses have been proposed, including the development of post-traumatic subperiosteal hemorrhage, vascular alteration of the bone, or hemorrhage from a pre-existing lesion.[12]

As in our case, the most frequent reason for consultation is patient-reported local pain.[13] With the development and expansive aggressive behavior of the lesion, neurological symptoms appear.

Imaging studies constitute a fundamental pillar in the diagnostic process of ABCs, since they present characteristic features. The lytic appearance, of an expansive nature, with fluid-fluid levels, are pathognomonic findings. The next step is the punch biopsy, which will confirm the result of the histopathology analysis.[14]

The treatment of ABCs continues to be a matter of controversy, due to the lack of defined guidelines. Multiple options have been described, to be used alone or in combination, including curettage with or without bone grafting, complete tumor resection, selective preoperative embolization, radiotherapy, chemotherapy, and intralesional injections.[15]

Curettage with bone grafting has a recurrence rate of 20%, which requires a more aggressive excision. Surgical stabilization should always be considered if a deformity develops postoperatively or due to the degree of bone resection.[16]

Radiotherapy is an option currently reserved for patients at high risk of not withstanding surgery or for those who are resistant to surgical treatment, even more so considering the potential risks of post-radiation myelopathy or sarcomatous transformation.[17,18]

Denosumab, a human monoclonal antibody that binds to the cytokine receptor activator of nuclear factor kappa B ligand, is one of the chemotherapy options. It prevents the action of agonists that act through RANKL receptors and prevents the subsequent activation and proliferation of osteoclasts. Although the reports of its use in cases of ABC are limited, chemotherapy has been considered a valid treatment option for those symptomatic ABCs that cannot be treated surgically or that have presented frequent recurrences.[9,20]

Intralesional injections of calcitonin, steroids, and concentrated bone marrow, among other options, are mentioned in the current literature, mostly in case series or reports. Although these represent a percutaneous method with few complications and adverse effects, the publications show variable results, in which partial remission predominates.[21,22]

Finally, in the face of large hypervascular tumors with a high risk of bleeding, selective preoperative embolization should be considered. As performed in our case as part of the preoperative treatment, embolization has been preferred in recent years as the first option when it is technically feasible, the diagnosis is solid, and there is the possibility of surgical intervention within 24–48 hours. Cure rates approaching 87% have been reported with this strategy.[23,24]

In this case, aggressive curettage was performed along with emptying of the lesion, associated with phenolization of the bed as a local method. It was associated with extensive decompression of the posterior arch, for which the need for instrumented fusion was imperative. Preoperative digital embolization was extremely helpful; it not only contributed to subsequent treatment, but also served to prevent massive intraoperative bleeding, since the patient had a fully thrombosed extensive epidural vascular tree, which could be seen on preoperative MRI.

CONCLUSIONS

ABC is a benign tumor with the potential to behave aggressively and with a considerable risk of local recurrence. Although there are numerous treatment options, these must be adapted to each particular case, taking into account the characteristics of the cyst.

In conclusion, surgical treatment was necessary in this case, given the clinical findings, the results of complementary studies and the tumor stage.

Conflict of interest: The authors declare no conflicts of interest.

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