

Aneurysmal Bone Cyst Treated with Sclerotherapy

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Abstract

Aneurysmal bone cyst (ABC) is a rare benign osteolytic tumor that predominantly affects children, adolescents, and young adults. Their pathophysiology is not fully understood, but genetic mutations have been identified in some cases. ABCs commonly present with pain, swelling, and potential functional impairment and may be associated with pathological fractures. Diagnostic imaging, including x-ray, MRI, and CT, plays a crucial role in the evaluation and diagnosis of ABCs. On imaging, a characteristic soap-bubble appearance and fluid-fluid levels may be present. Surgical resection, curettage with or without bone grafting, sclerotherapy, embolization, and radiation therapy are treatment options.

Keywords: bone, benign neoplasm, pelvis, bone intervention

Case Summary

A middle-childhood patient with right hip pain. Pelvic radiography and MRI were performed, showing an expansile lesion of the left ischium. On MRI, the lesion was hyperintense on a T2 sequence. Subsequent orthopedic consultation led to CT-guided sclerotherapy with doxycycline on two occasions. On follow-up, the child showed dramatic improvement radiographically and clinically and was asymptomatic.

Imaging Findings

Right hip radiograph reveals an expansile lesion of the right ischium (arrow) that is confirmed on MRI (Figure 1). The lesion involves the right ischium and posterior acetabulum with internal septations and fluid-fluid levels (Figure 2). An axial CT image

from CT-guided sclerotherapy shows the needle placement, positioned in the lesion via a transgluteal approach (Figure 3). Follow-up MRI 12 months after sclerotherapy showed near-complete resolution of the lesion (Figure 4).

Diagnosis

Aneurysmal bone cyst (ABC).

The differential diagnosis includes unicameral bone cysts, giant cell tumor, and telangiectatic osteosarcoma. Osteoblastoma and chondroblastoma may also be mistaken for ABCs depending on their location.

Discussion

ABCs are benign osteolytic tumors with an incidence rate of 0.14 per 100,000 individuals.¹ These tumors are

more commonly diagnosed in children, adolescents, and young adults, with 75-90% of cases occurring in patients under 20 years old.² After the age of 30 years, the occurrence of ABCs becomes rare, and it is exceptionally rare after the age of 50 years. While ABCs can develop in any skeletal structure of the body, they often affect the metaphysis of long bones and the spine. In the spine, the lumbar vertebrae are the most frequently affected location.¹ ABCs tend to involve the posterior aspects of the vertebrae and can lead to vertebra plana or significant compression fractures, which are characterized by the loss of the entire height of the vertebral body.³

The pathophysiology of ABCs remains unknown, but several proposed mechanisms have been put forth. One of the earliest theories suggests that ABCs originate from a vascular anomaly, leading to local disturbances in blood flow within the bone and the subsequent formation of "cystic," blood-filled spaces. Another theory implicates previous trauma or injury as a trigger for ABC formation, although it is important to note that trauma is not consistently observed in cases.¹ Recent studies have made significant advancements by identifying genetic

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Figure 1. Pelvic radiograph showing expansile lucent lesion of right ischium.

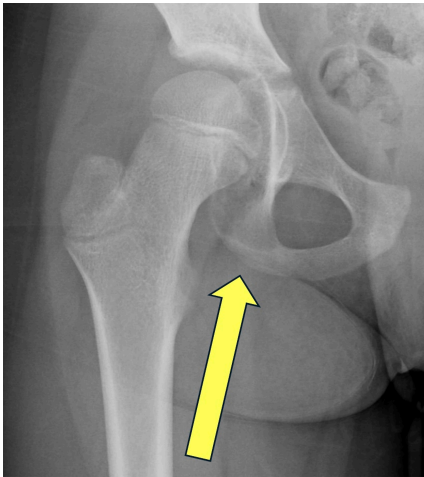


Figure 2. Axial T2-weighted MRI showing hyperintense expansile lesion of posterior acetabulum/ischium (arrow).

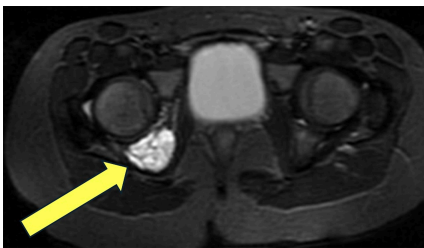
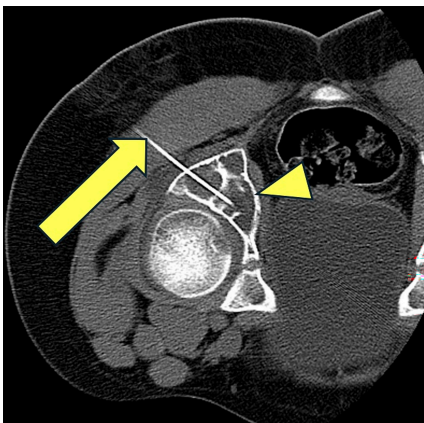
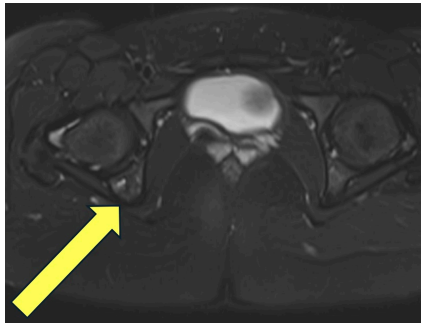


Figure 3. CT-guided placement of two needles (arrows) into the lesion, which were used to inject doxycycline to sclerose the lesion.



mutations within the USP6 or CDH11 genes in a significant portion of ABC cases. These genetic mutations distinguish

Figure 4. Pelvic MRI and radiograph 4 years post-sclerotherapy showing radiographic and MRI resolution with only a small cystic lesion remaining on axial T2 MRI (arrow).



ABCs as primary tumors, setting them apart from secondary tumors that lack these specific mutations.⁴ Primary ABCs develop independently as a benign, “cystic” bone lesion with blood-filled spaces. In contrast, a secondary ABC is formed as a response to another bone lesion. Some examples of secondary ABCs include fibrous dysplasia, chondroblastoma, non-ossifying fibromas, and telangiectatic osteosarcoma.

ABCs often present with pain and swelling. The intensity of pain can vary from mild to severe, potentially limiting movement depending on the size and location of the tumor. Additionally, swelling and the tumor’s expansive growth can lead to bone or joint deformity.⁴ In rare cases, an early indication of the disease may be a pathological fracture. If the lesions occur in the spine, they can result in conditions such as torticollis, scoliosis, vertebral plana, and neurological symptoms.²

Various imaging studies can assist in the diagnosis and characterization of ABCs. The initial study often involves plain radiography, which reveals lobulated and expansile radiolucent lesions with well-defined borders. The internal septations give the tumor its characteristic “soap-bubble” appearance.¹ These lesions are typically found in the metaphysis of bones and are eccentrically located, although they can also occur in the

diaphysis. In more aggressive forms, a periosteal reaction resembling the Codman triangle may be present.¹

MRI is the imaging examination of choice due to its superior ability to visualize soft tissues, allowing for precise assessment of the lesion’s extent. It also provides information about the internal composition of the tumor. The presence of an expansile lesion with fluid-fluid levels strongly supports the diagnosis of an ABC.² MRI can also detect perilesional edema in either the surrounding soft tissues or bone marrow, which is indicative of rapid growth.¹ CT has lower sensitivity in detecting fluid-fluid levels compared to MRI but can be valuable in constructing a two- or three-dimensional lesion map in anatomically complex areas.² While other imaging modalities may not be diagnostically useful, they can play a role in the treatment phase.

The treatment options for ABCs can vary depending on factors such as the lesion’s location, size, and extent. There are several therapeutic approaches that can be used, including surgical resection, sclerotherapy, embolization, and radiation therapy. Currently, image-guided sclerotherapy is the treatment of choice.¹

Recent meta-analyses have revealed that even simple curettage can achieve high healing rates. However, it is associated with a higher risk of disease recurrence, approximately 22%. This recurrence rate can be mitigated by utilizing adjuvant therapies such as cryotherapy, cancellous, or synthetic bone grafting, in combination with the management approach.⁵

Sclerotherapy is a minimally invasive treatment option that involves injecting a sclerosing agent directly into the lesion. The aim of sclerotherapy is to induce sclerosis and fibrosis of the “cystic” spaces, leading to the collapse and stabilization of the ABC. Sclerotherapy is particularly useful for ABCs located in challenging anatomical locations and as an option for patients who are not suitable candidates or refuse surgery. Sclerotherapy with alcohol or

doxycycline has comparably high healing rates as minimally invasive interventions but requires multiple treatments before healing is achieved.⁵ Radiological findings post-treatment include an overall decrease in cystic area of bone and an increase in cortical thickness, although these changes may take 10 months or more to become evident on imaging.⁶

Embolization and radiation therapy are also considered alternative treatment modalities for ABC. Embolization involves the selective occlusion of the feeding arteries supplying the ABC using a variety of embolic agents. By cutting off the blood supply, embolization aims to reduce the vascularity of the lesion, leading to shrinkage of the lesion size and stabilization. Prospective studies have found comparable rates of healing between patients undergoing embolization and curettage, with few complications and low rates of recurrence at up to 2 years.³ Additionally, arterial embolization performed before subsequent curettage has resulted in lower recurrence rates and lower volumes of intraoperative blood loss.⁵ Radiation therapy, while

having acceptably good outcomes, is also associated with radiation-induced malignancies, and its use needs to be carefully evaluated by a multidisciplinary team.

Conclusion

ABCs are rare benign osteolytic tumors that predominantly affect children, adolescents, and young adults. Their exact pathophysiology is not fully understood, but genetic mutations have been identified in some cases. ABCs commonly present with pain, swelling, and potential functional impairment and may be associated with pathological fractures. Diagnostic imaging, including X-ray, MRI, and CT scans, plays a crucial role in the evaluation and diagnosis of ABCs. On imaging, characteristic soap-bubble appearance and fluid-fluid levels may be present. Surgical resection, curettage with or without bone grafting, sclerotherapy, embolization, and radiation therapy are employed as treatment modalities.

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