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Case Report

A rare case of huge aneurysmal bone cyst of the pelvis

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ABSTRACT

A 21-year-old female presented with a huge painful aneurysmal bone cyst (ABC) involving the left ilium, acetabulum, and ischium, resulting in a limp and activity limitation. After serial selective arterial embolization, intravenous bisphosphonates, and surgical curettage with bone grafting and cementing, she could achieve painless, nearly full range of motion at 2-year and 3-month follow-up with no recurrence. Her quality of life using the RAND 36-item Health Survey (version 1.0), the Musculoskeletal Tumor Society Scoring System (MSTS), and the Toronto Extremity Salvage Score indicated a good outcome. The aim of reporting this case was to show that it is safe to surgically approach a large pelvic ABC after devascularization with scleroembolic procedures augmented with intravenous bisphosphonates.

Keywords: Aneurysmal bone cyst, Bisphosphonates, Curettage, Embolization, Pelvis

INTRODUCTION

Aneurysmal bone cysts (ABCs) are rare benign lytic lesions of the bone.[1,2] Primary ABC accounts for 1%–2% of all primary bony tumors and usually presents in the first two decades of life.[3-7] A study describing the anatomical distribution of 897 ABCs found pelvic lesions in 11.6% of the cases, the fourth most frequent site in descending order after tibia, femur, and vertebra.[8] Despite accounting for nearly half of all flat bone lesions,[9] pelvic ABCs are extremely rare. Management of pelvic ABCs requires careful consideration of special factors, such as the risk of bleeding, proximity to neurovascular structures, relative inaccessibility, and damage to the integrity of weight-bearing structures such as the acetabulum or sacrum.[5,10,11] Potential treatment modalities include observation, direct injection of the cyst with a fibrosing agent, embolization, resection, and intralesional curettage with or without local adjuvant therapies.[10] We present the rare case of a huge ABC extending over three pelvic zones: the ilium, ischium, and acetabulum. In this case, the challenges were its considerable size, location, proximity to the neurovascular bundle, and involvement of weight-bearing areas. We used a combined neoadjuvant and surgical approach that resulted in a successful outcome. The aim of reporting this case was to show that it is safe to surgically approach these lesions once devascularization with scleroembolic procedures augmented with intravenous bisphosphonate neoadjuvant treatment has taken place.

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CASE REPORT

A 21-year-old female teacher presented to our department in February 2012 complaining of pain in the left hip joint for the past 4 years. At the onset, the pain was dull but progressed in intensity over time. It aggravated with movement and improved with rest and oral analgesics. She gradually started limping and was using a cane to walk. In 2010, she had a biopsy taken in another hospital and was diagnosed with an ABC. During the following 2 years, she visited several hospitals in the country but was refused treatment due to the challenging nature of her tumor. Her physical examination showed a restricted and painful range of motion in the left hip joint. She also had a slight bulge at the left inguinal region, which was mildly tender to touch and with a hard consistency. Our initial radiological assessment included plain radiographs, computed tomography scan (CT), three-dimensional (3D) CT scan, and magnetic resonance imaging (MRI). The images revealed a 16 cm × 11 cm × 7 cm expansile lytic cavitary lesion over the left ilium, ischium, and acetabulum with characteristic multifluid levels. The mass fell short of the left sacroiliac joint but reached up to the midline of the pelvis displacing the uterus, cervix, and bladder toward the right. The outer cortex of the lesion showed thinning, occasional areas of the cortical break, a clear plane of separation, and no evidence of soft-tissue invasion. The femoral neurovascular bundle was well spared and away, the hip joint was congruent, and the cartilage and the proximal femur appeared normal. The intravenous gadolinium contrast-enhanced T1-weighted MRI showed evidence of linear septal-type enhancement within the lesion and no effusion in the hip joint [Figure 1]. The stage of the lesion was Enneking 3.

Considering the location and size of this ABC, the treatment plan consisted of devascularization of the lesion, increasing the bone density and filling the bone cavity after curettage. From April 2012 to April 2014, the patient underwent six sessions of selective arterial embolization (SAE) with polyvinyl alcohol particles every 3–6 months by an interventional radiologist. In parallel, she received intravenous bisphosphonates to promote bone consolidation (pamidronate 7.5 mg/kg/dose) at 6-month intervals.

There was gradual and significant bony consolidation of the lesion after consecutive sessions of SAE and intravenous bisphosphonates [Figure 2], but the patient had persistent pain. In August 2014, we decided to perform the curettage of the lesion but retaining the bony shell and filling the void with bone cement. To maximize its devascularizing benefits, her surgery was planned 4 months after her last SAE session. We used a left ilioinguinal approach in a supine position. Upon reaching the tumor, the pelvic contents were retracted medially, and a 3 cm × 3 cm window was created

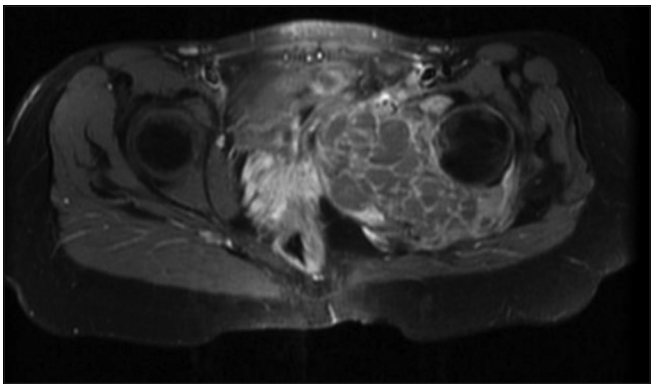

Figure 1: Magnetic resonance imaging with contrast axial section showing the well-defined aneurysmal bone cyst mass displacing the pelvic viscera, femoral neurovascular bundle is spared and reduced muscle bulk.

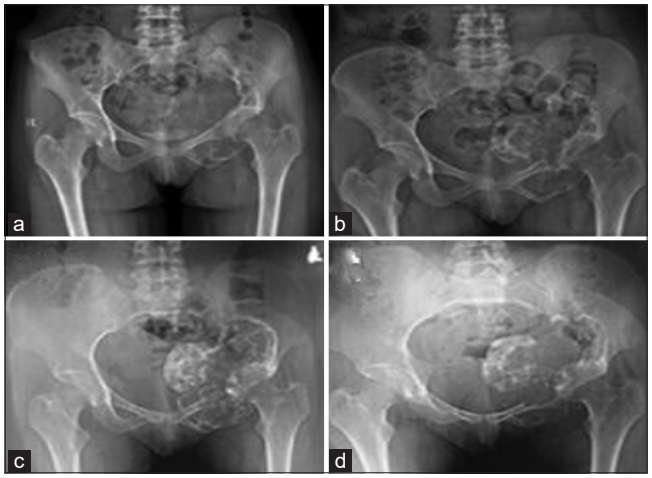

Figure 2: Radiographs taken after one (a), two (b), three (c), and six (d) sessions of selective arterial embolization showing progressive bony consolidation of the aneurysmal bone cyst shell in the left hemipelvis.

on the tumor surface. We anchored two long cancellous screws in the cavity (to increase the bone cement surface area for anchorage) after curettage and then filled it with polymethylmethacrylate. A layer of autologous cancellous bone graft harvested from the ipsilateral iliac crest was placed in the supra-acetabular area of the cavity as well. The intraoperative blood loss was 250 ml. Figure 3 shows the immediate postoperative radiograph and after 2 years and 3 months of follow-up.

The patient was progressively mobilized 6 weeks after surgery from supported walker to independent weight-bearing. She had radiographic follow-up every 3 months for the 1st year and then every 6 months the following year. The lesion's progression by 3D CT scans taken at various stages of her management is shown in Figure 4. There was no evidence of recurrence at her last follow-up 2 years and 3 months after

surgery (December 2017). The patient had minimal pain and could walk without walking aids. She was also able to perform independently her activities of daily living such as cooking, climbing stairs, going for groceries, cleaning, and squatting. We measured her quality of life using the RAND 36-item Health Survey version 1.0.[12] Her aggregate score was "good," 70 out of 100.[13-15] We also used two disease-specific instruments that evaluate the functional outcomes of patients with extremity tumors, the Musculoskeletal Tumor Rating Scale (MSTS),[16] and the Toronto Extremity Salvage Score (TESS). The MSTS score was 25 out of 35, whereas the TESS was 81 out of 100, both indicating good outcomes.[17,18] The patient was 8 months into her first pregnancy at her last visit and reported feeling accomplished and enthusiastic.

DISCUSSION

Treatment of large ABCs of the pelvis (size > 5 cm) is a challenge.[5] The published literature is limited to case reports and case series, and therefore, little evidence to the best treatment options for this type of cases is currently available. The case presented here is peculiar in many aspects. First, it was a large endopelvic mass, which we decided to treat with six SAE sessions combined with intravenous bisphosphonates over 2 years. This unique approach resulted in significant consolidation of the lesion. Second, the extensive devascularization by SAE rendered it unnecessary to control the feeding vessels intraoperatively and therefore minimizing the risk of massive intraoperative bleeding that could result in death as reported in a case of a 25 cm × 25 cm × 20 cm tumor.[19] Third, most cases of large ABCs undergo marginal or wide excision instead of curettage.[5] However, the advantage of retaining the bony shell is that a near anatomical construct is preserved and extensive reconstructive procedures can be avoided in young patients.

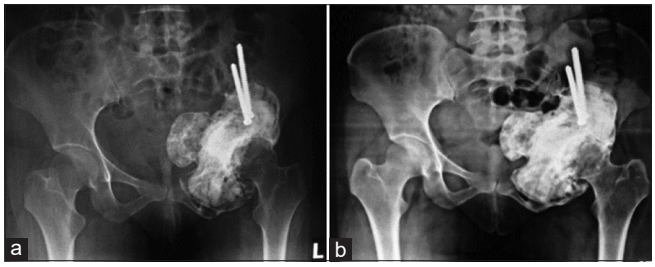

Figure 3: Immediate postoperative radiograph (a), and after 2 years and 3 months of follow-up (b). Long cancellous screws can be appreciated in the cavity of the lesion, as well as the homogenous distribution of bone cement.

Rossi *et al.* described a case similar to ours in a 13-year-old boy who underwent SAE four times in 5 months but without bony consolidation.[20] The tumor was treated with extended curettage, and the void was filled with Plexur M™, a moldable bone graft substitute. However, additional measures during the surgery had to be taken to control the bleeding. Similar to our case, the patient had a near-normal range of motion, no recurrence, and restored walking after 2 years of follow-up.

Ozdemir *et al.* reported a tumor of 30 cm × 30 cm × 20 cm at the second sacral vertebral level in a 14-year-old. They achieved marginal excision with a combined anteroposterior approach after ligation of the internal iliac arteries. Although they had no clinical nor radiological recurrence and no dysfunction of bowel and bladder at 84 months of follow-up, they used 14 pints of blood transfusion and postoperative radiotherapy which added significant morbidity to the patient.[21] The experience narrated by Wathiong *et al.* in managing an 11-cm iliopubic ABC with a single session of SAE followed by curettage and bone grafting without major blood loss (800 ml) is an example of a similar successful treatment strategy.[7] However, our treatment method remains unique as we found no mention of the use of intravenous bisphosphonate for large pelvic ABCs, which we believe played a pivotal role in consolidating the lesion.

Biphosphonates are known to inhibit osteoclastic bone resorption (osteoclastogenesis) as well as have antitumor and antiangiogenesis effects.[22] It results in an inhibition of protein prenylation and RAS signaling in osteoclasts leading to a decrease in bone resorption.[23] Cornelis *et al.* in their series of eight patients with symptomatic unresectable benign bone lesions, such as an ABC, Langerhans cell histiocytosis, osteoblastoma, and a giant cell tumor, used bisphosphonate as an alternative treatment. They concluded to have achieved

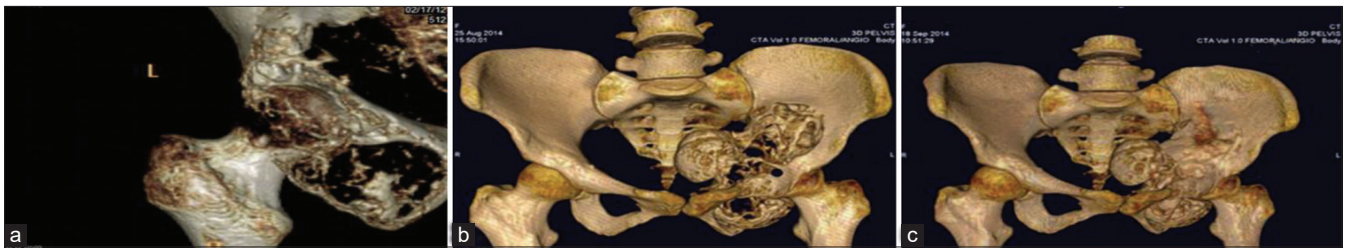

Figure 4: Three-dimensional computed tomography scan of the patient from February 2012 at the time of diagnosis (a), preoperative after six sessions of selective arterial embolization and intravenous bisphosphonates in August 2014 (b), and postoperative in September 2014 showing filling of the bony cavity with bone cement after curettage (c). The images show the progressive bony consolidation over time.

ossification without adverse effects as an alternative to surgical treatment in difficult anatomical locations of the spine and pelvis.[24] In our case, the patient received seven bisphosphonate sessions over a period of 2 years and significant radiological consolidation was achieved.

Our treatment approach for this rare case resulted in restored function, good quality of life, and patient satisfaction. However, her MSTS, TESS, and short-form (SF)-36 scores were lower than those reported by Novais *et al.* in 13 children with pelvic ABCs who had a mean age of 12.9 years (range: 4.1–17.5 years) and a minimum follow-up of 5 years.[10] They reported a mean TESS of 95, MSTS was 93%, and SF-36 was 87% of the total points. The difference with our results might be explained by the patients' younger age, who presumably had an earlier diagnosis and a longer follow-up.

Our case report has the limitation that no validated outcome assessment of the patient was made preoperatively or during the early postoperative phase, so the exact amount of functional improvement could not be measured in a reliable way. A long-term follow-up of the patient will further validate the outcomes of the management plan devised for her.

CONCLUSION

Large pelvic ABCs with an endopelvic mass can be treated with scleroembolic procedures before undertaking major surgery. Since ABC is a benign tumor, all available minimally invasive resources should be exhausted before proceeding to surgery. Curettage, along with filling of bony voids with a synthetic substance and intravenous bisphosphonates, appears to be a good option for these cases.

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AUTHORS' CONTRIBUTIONS

AS conceived and designed the case report, conducted patient management, provided research materials, and collected and organized data. AHC and ER analyzed the case and formatted the figures. AS wrote the initial and final draft of the article. SMA conducted and planned patient management and provided logistic support. All authors have critically reviewed and approved the final draft and are responsible for the manuscript's content and similarity index.

ETHICAL APPROVAL

Informed consent was taken from the patient and Institutional Review Board for the publication of this case report.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published, and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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