Aneurysmal bone cysts: Case report and literature review

Loraine Quintana-Pajaro1,2, Cristian Camilo Blanco-Teheran1,2, Daniela López-Cepeda1,2, Luis Rafael Moscote-Salazar2-3*, Alfonso Pacheco-Hernandez2,3, Amit Agrawal1, Harsh Deora3, Alexis Rafael Narvaez-Rojas3

BACKGROUND

Aneurysmal bone cysts (ABC) are benign lesions with great vascularization and expandability. They represent approximately 1% of all bone tumors. These lesions are characterized by having multiple cavities filled with blood. In most cases, they are located in the thoracic and lumbar region. In this article, we aimed to present a clinical case and followed by review of relevant literature regarding pathophysiology, clinical, diagnostic and therapeutic aspects of this pathology.

CASE PRESENTATION

A 41-year-old woman came with chief complaint of developing pain in the left hip, right gluteal region, and right posterior thigh for last 10 months. We performed hip plain x-ray, which suggested a left hip injury. Further investigation using computed tomography and magnetic resonance imaging were performed and revealed tumor of the left hip (Figure 2). A biopsy guided by computed axial tomography was performed. The histopathologic examination revealed numerous small vascular spaces lined with endothelial cells, abundant pools of red blood cells were seen, and hemosiderin pigment was seen at places along with giant cells, which was suggestive of ABC.

CONCLUSION: Aneurysmal bone cysts are lesions that represent a small percentage of the total osseous tumors. Sacral localization is rare and the manifestations are nonspecific.

Keywords: Aneurysmal bone cysts, bone tumor, iliac, sacral

*Corresponding to: Luis Rafael Moscote-Salazar; Facultad de Medicina, Campus de Zaragoza, Universidad de Cartagena, Cartagena, Colombia; mineurcirujano@aol.com

INTRODUCTION

Aneurysmal bone cysts (ABC) are benign lesions, non neoplastic, with great vascularization and expandability. They represent approximately 1% of all bone tumors and were first described by Van Arsdale.1,2 Generally, patients with these lesions have neurological and somatic pain symptoms. There may be rigidity or even the presence of a palpable mass.3–7 Initially, x-rays are used for initial diagnosis but they are unspecific and do not reveal ABC appearance. Therefore, other specialized studies are performed such as computed tomography, magnetic resonance, bone scan and biopsy.3

The treatment is still controversial, so we do not yet have a preference toward another. However, surgery is the most common treatment with the complete resection having the least recurrence rate. It can also be managed with adjuvant treatments such as radiotherapy, selective arterial embolization, among others.3,8–11

In this article, we aimed to present a clinical case and followed by review of relevant literature regarding pathophysiology, clinical, diagnostic and therapeutic aspects of this pathology.

CASE PRESENTATION

A 41-year-old woman came with chief complaint of developing pain in the left hip, right gluteal region, and right posterior thigh for last 10 months which affected her daily activities. She was managed with analgesics and anti-inflammation medication without obvious improvement. The pain was worsening and followed by increased analgesic doses. On physical examination, we found normal vital signs and hydrated oral mucosa. Tenderness over right hip was negative.

We performed hip plain x-ray, which suggested a left hip injury (Figure 1). Further investigation using computed tomography and magnetic resonance imaging were performed and revealed tumor of the left hip (Figure 2). A biopsy guided by computed axial tomography was performed. The histopathologic examination revealed numerous small vascular spaces lined with endothelial cells, abundant pools of red blood cells were seen, and hemosiderin pigment was seen at places along with giant cells, which was suggestive of ABC.

Figure 1. Preoperative x-ray examination. A. Anteroposterior pelvis x-ray. Well-defined lesions and edges were observed with a honeycomb pattern at the level of the left iliac bone near the sacroiliac joint. B. Anteroposterior x-ray of the left hip. Aneurysmal bone cysts were seen in the left iliac bone. Notice the pattern of destruction. The bone was expanded but the lesions were marginalized by cortical bone.
local vascular alteration, failure in bone maturation and chromosomal modifications (translocation of t(16;17) (q22;p13)). The local vascular alteration may be due to secondary to preexisting bone tumor such as giant cell tumor, osteoblastoma, osteosarcoma, and chondroblastoma; failure in vascular repair due to traumatic injury; and vascular malformation in bone. In the 1990s, Kransdorf and Sweet proposed that ABCs were the secondary changes to a vascular malformation which was caused by evolutionary alteration of a preexisting bone tumor. It led to the formation of a second lesion in the bone that masked the primary bone pathology that triggering it.

Increase of local vascular pressures triggers an osteolytic-osteogenic process in the trabeculae of the affected bone which produces the appearance of multiloculated cavities filled with blood. This process consisted of 4 phases. In the first stage, the osteolysis predominates and in second phase, the lesion grows and expands which cause thinning of the bone. The third and fourth phase are characterized by soap-bubble pattern and healing process, respectively. The latter is distinguished by the ossification of the lesion and only a small group of these patients reaches this stage.

Therefore, at the end of the 1990s, the study by Panoutsakopoulos et al. showed that some ABCs had a genetic component that could trigger the appearance of these tumors. These chromosomal mutations consisted of translocations of chromosomes 16 and 17. These findings were similar to study by Oliveira et al. in 2004 who analyzed 8 cases and found that recurrent translocations in these chromosomes cause the juxtaposition of CDH11 and USP6 (ubiquitin-specific protease), increasing the transcription of this protease.

ABCs usually occur between the first and second decade of life and predominantly in women. They represent 1% of all primary bone tumors. The predilections are femur, tibia, humerus, spine, pelvis, ribs, small bones of the hands and feet. As many as 10 – 30% occurs in spine with predominance of the lumbar spine and equal occurrence in thoracic and cervical spine.

At present, the pathophysiological process of aneurysmatic bone cysts formation is still not completely elucidated. Multiple theories have been proposed that can be grouped into vascular spaces lined with endothelial cells, abundant pools of red blood cells were seen, and hemosiderin pigment was seen at places along with giant cells, which was suggestive of ABC.

We proceed to perform excision-curettage and bone-grafting surgery. Patient evolves satisfactorily and discharged after 10 days.

**DISCUSSION**

Aneurysmal bone cyst was firstly described by Van Arsdale in 1893 as ossifying hematomas, and in 1942 the term of ABC was started to being used. These lesions are benign, tumor-like, locally aggressive and highly vascular. They grow quickly and predominantly inside long bones with filled cystic cavities and multiple internal septations of varying thickness.

ABC is usually found in young patients between the first and second decade of life and predominantly in women. They represent 1% of all primary bone tumors. The predilections are femur, tibia, humerus, spine, pelvis, ribs, small bones of the hands and feet. As many as 10 – 30% occurs in spine with predominance of the lumbar spine and equal occurrence in thoracic and cervical spine.

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We proceed to perform excision-curettage and bone-grafting surgery. Patient evolves satisfactorily and discharged after 10 days.
Table 1. Summary of literature reporting case of aneurysmal bone cyst in the sacrum

| Authors             | Year | Case | Age | Gender | Clinical manifestations                                                                 | Duration of symptoms |
|---------------------|------|------|-----|--------|----------------------------------------------------------------------------------------|-----------------------|
| Zileli, et al.       | 2013 | 6    | 29  | M      | Low back pain                                                                          | 24 months             |
|                     |      | 32   |     | F      | Left leg pain, left monoparesis                                                         | 12 months             |
|                     |      | 43   |     | M      | Low back and leg pain                                                                  | 1 month               |
|                     |      | 15   |     | M      | Low back and left leg pain, paraparesis                                                 | 3 months              |
|                     |      | 13   |     | F      | Low back and leg pain                                                                  | 12 months             |
|                     |      | 22   |     | F      | Low back pain, paraparesis                                                             | 12 months             |
| Pogoda, et al.       | 2003 | 1    | 17  | F      | Moderate abdominal pain and intermittent weakness of right leg                         | 4 weeks               |
| Codd, et al.         | 2006 | 1    | 8   | M      | Low back pain                                                                          | 4 months              |
| Mulhall, et al.      | 2003 | 1    | 15  | M      | Low back pain                                                                          | 2 months              |
| Aydin, et al.        | 2016 | 1    | 14  | F      | Sacral and leg pain                                                                    | 24 months             |
| Brastianos, et al.   | 2009 | 10   | 64  | F      | Low back pain, weakness of the right foot loss of sensation and tingling                | 8 – 10 years          |
|                     |      |      | 26  | F      | Low back pain                                                                          | 3 months              |
|                     |      |      | 39  | M      | Constipation and bladder dysfunction                                                    | 1 year                |
|                     |      |      | 16  | F      | Low back pain                                                                          | 3 months              |
|                     |      |      | 32  | M      | Low back pain and radiation to right leg                                                | -                     |
|                     |      |      | 60  | F      | Low back pain with weight loss                                                         | 7 months              |
|                     |      |      | 57  | M      | Low back pain and difficulty walking                                                    | 5 months              |
|                     |      |      | 10  | F      | Left leg pain                                                                          | -                     |
|                     |      |      | 5   | F      | Back pain and difficulty walking                                                       | 3 months              |
|                     |      |      | 17  | F      | Low back pain and bowel and bladder dysfunction                                         | -                     |
| Jevtich              | 1988 | 1    | 60  | M      | Low back pain, radiating along the right leg                                            | -                     |
| Simm, et al.         | 2013 | 1    | 8   | M      | Left posterior thigh pain                                                              | 3 weeks               |
| Skubitz, et al.      | 2015 | 1    | 27  | M      | Low back pain                                                                          | -                     |
| Vester, et al.       | 2010 | 1    | 19  | F      | Lower back pain also affecting thigh and knee                                          | 6 months              |
| Ghermandi, et al.    | 2016 | 1    | 42  | M      | Lumbar back pain                                                                       | -                     |
| Nas, et al.          | 2015 | 1    | 32  | M      | Pain radiating to thighs and anal region, numbness and incontinence                     | -                     |
| Doss, et al.         | 2014 | 1    | 16  | M      | Sacral and leg pain                                                                    | 4 months              |
| Yildirim, et al.     | 2007 | 1    | 13  | M      | Left hip and low back pain, related to accident                                         | 4 months              |
| Rajadhyaksha, et al. | 2003 | 1    | 13  | F      | Pain in the right buttock and posterior thigh.                                          | -                     |
| Derinkuyu, et al.    | 2016 | 1    | 13  | F      | Difficulty in walking, left leg pain, and lower back pain                               | 1 month               |

Table 2. Summary of treatment options for aneurysmal bone cyst

| Treatment options          | Description                                                                 | Reference |
|----------------------------|-----------------------------------------------------------------------------|-----------|
| Surgery                    | It was considered the main treatment and the recurrence rate depends on resection, or curettage with bone graft or without it. Resection provided lower rate of recurrence. | [3], [4], [8], [9], [10], [12], [16], [28], [33] |
| Radiotherapy               | In most cases, it was used as an adjuvant treatment. However, there was risk of developing a malignancy. The dose was really important for the outcome of this treatment. | [3], [4], [7], [8], [9], [12], [13], [16], [34] |
| Selective arterial embolization | It is performed as a preoperative treatment to reduce bleeding in the surgery. However, it could have some complication such as the risk of embolizing arterial supply to adjacent normal tissue and embolic complications. | [1], [3], [9], [11], [12], [13], [15], [16], [35] |
| Bisphosphonate injection  | First reported in 2013 by Simm, et al. when administered in children and adolescents with ABC. Its effectiveness to inhibit osteoclastic activity and reduction of inflammation was demonstrated. However, it was considered lack of evidence for its use due to adverse effects of osteonecrosis. | [3], [23] |
| Percutaneous cryoablation | It should be considered when the goal of treatment was to reduce tumor size. | [3] |
| Denosumab therapy         | This was a human monoclonal antibody that bind to and inhibit receptor-activator of nuclear kappa beta ligand (RANKL). RANKL was expressed in specific cells of ABC. It was recommended to patients who do not respond to selective arterial embolization and contraindicated for surgery. | [36], [37] |
as an adjuvant treatment. However, there is risk of inducing malignancy in the ABC due to radiotherapy. It is believed that radiotherapy should only be prescribed in patients where surgery is contraindicated. This type of treatment is completely contraindicated for those young patients with lesions near the spine due to the risk of developing myelopathies and the potential to induce spinal deformity.4

For the treatment of ABC, we found some methods that are minimally invasive, such as selective arterial embolization, sclerotherapy, bisphosphonate injection, cementoplasty, percutaneous ablation, among others that have been used recently.3,9,10 The summary of treatment option for ABC can be seen in Table 2.

In present time, ABC has an excellent prognosis. However, it is recommended to monitor these patients continuously there is still possibility of recurrence. In addition, it must be emphasized that early and adequate diagnosis of these patients leads to better treatment and therefore a lower recurrence rate and better prognosis.3,9,15

CONCLUSION
Aneurysmal bone cysts are lesions that represent a small percentage of the total osseous tumors. Sacral localization is rare and the manifestations are nonspecific. It is very important to rule out the presence of a primary bone lesion such as osteosarcomas and the possibility of recurrent bone cysts.

CONFLICT OF INTEREST
None of the authors of this paper has a financial or personal relationship with other people or organizations that could inappropriately influence or bias the content of the paper.

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