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Case report

Malignant Transformation of an Aneurysmal Bone Cyst to Huge Fibroblastic Osteosarcoma without Radiation Exposure: Proper Surgical Management is More than a Challenge

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Abstract

In this study, we present a case of aneurysmal bone cyst presenting as pathologic fracture of the left proximal femur. The patient, who had history of multiple surgeries, was referred to us with a giant mass. The patient was found to develop osteosarcoma without radiation exposure. External hemipelvectomy was performed.

Appropriate surgical management of a huge tumor that repeatedly operated is more than a challenge like this case. Aneurysmal bone cysts rarely present as pathologic fractures initially. In addition, to the best of our knowledge, the number of aneurysmal bone cysts (ABCs) exhibiting malignant transformation to osteosarcoma without radiation exposure is less than ten in the literature.

Keywords*: aneurysmal bone cyst, osteosarcoma, malignant transformation, hemipelvectomy.*

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Introduction

Aneurysmal Bone Cyst (ABC) is a vascular tumor-like cystic formation that expands and sometimes completely destroys the bone. ABCs have a wide spectrum of clinical manifestations ranging from a dormant lesion to an aggressive behavior mimicking sarcoma. Chief complaints are swelling and pain due to the superficial nature of the lesion. Although very rare, a pathologic fracture can also be encountered. Moreover, ABC can sometimes be a secondary component of a benign tumor or an osteosarcoma [1]. Although ABC is a benign lesion, there is a possibility of malignant transformation as a result of radiation exposure. Malignant transformation without radiation exposure is extremely rare [2,3]. The number of malignant transformation cases reported is highly limited so far. In addition, most of the reported cases had transformation to osteosarcoma [4]. In this case report, we present a case of ABC that was diagnosed with a rare manifestation, i.e., pathologic fracture, and exhibited malignant transformation without radiation exposure.

Case report

Twenty-three-year-old male patient was referred to our clinic with a giant mass extending from the left hip to the femoral region. The Syrian patient had history of two surgeries performed on the same region at the same center.

According to the detailed medical history of the patient, he presented with pain in the left hip nearly 4 years ago and was found to have a pathologic fracture in the left proximal femur with direct radiography (Figure 1a). MRI showed a cystic lesion filling the trochanteric region and extending to the femoral neck and inferior border of the lesser trochanter (Figures 1b-1c).

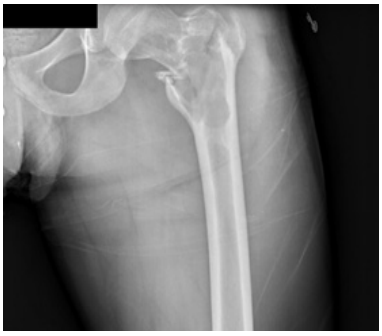

Figure 1a - Direct radiograph at the initial presentation of the patient

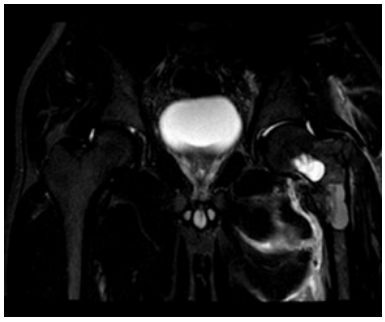

Figure 1b - T2 slice on MRI at the initial presentation of the patient

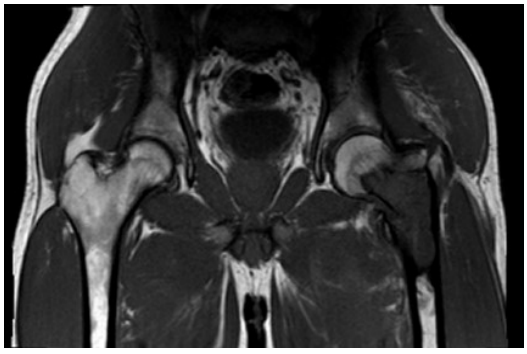

Figure 1c - T1 slice on MRI at the initial presentation of the patient

Surgery was planned and intraoperative frozen section examination showed that the lesion was compatible with ABC. Therefore, it was understood that the patient had undergone lesion curettage + cautery and adjuvant therapy with alcohol + allografting + anatomical plate fixation of the proximal femur (Figure 2a). Result of the histopathological examination was reported as ABC. It was reported that the patient did not show up for follow-up visits and presented with pain in the left hip at postoperative month 12. Direct radiographs showed recurrence (Figures 2b-2c). The patient was planned to undergo sclerotherapy and it was reported that the patient then underwent open surgery, which involved curettage+cautery and adjuvant therapy with alcohol + allografting, since it was found that the sclerosing

agent could not be sufficiently dispersed throughout the lesion. Result of the histopathological examination was compatible with recurrent ABC. It was reported that the patient did not show up for follow-up visits, had pain in the left hip at his initial presentation nearly 9 months after the second surgery and that direct radiographs were compatible with recurrence (Figure 2e). The patient rejected denosumab therapy, which he was recommended, and presented to the hospital nearly 19 months later. It was reported that the cystic lesion grew further and extended to the pelvis and femoral diaphysis (Figure 2f). The patient was then referred to our clinic.

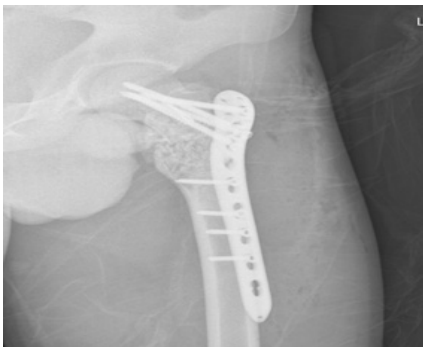

Figure 2a - Postoperative radiograph following the first curettage-grafting and fixation

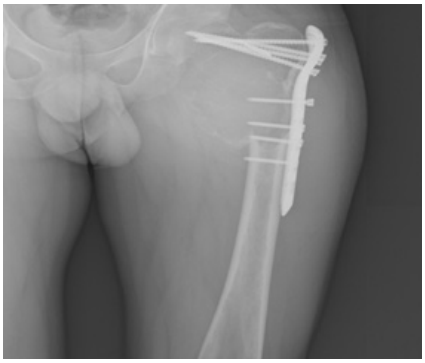

Figure 2b - AP radiograph 1 year after the first surgery

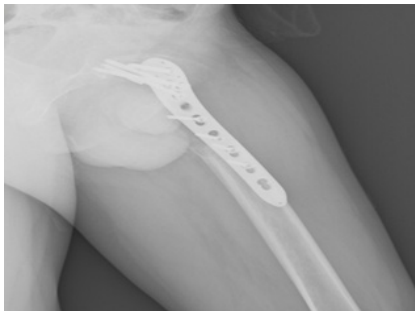

Figure 2c - Lateral radiographic view 1 year after the first surgery

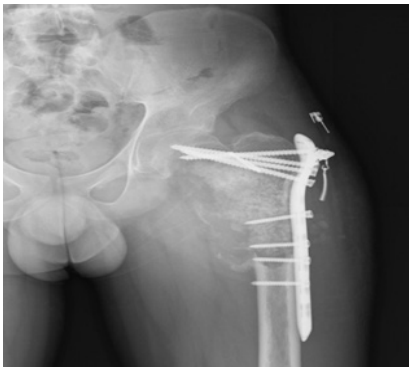

Figure 2d -Postoperative radiograph following the second curettage-grafting

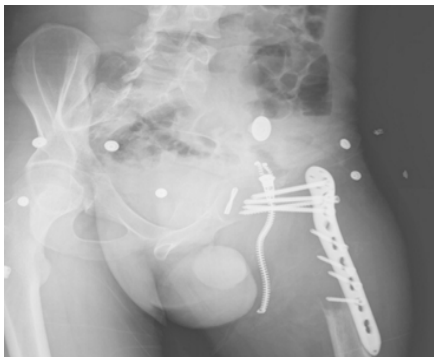

Figure 2e - Direct radiograph 9 months after the second surgery

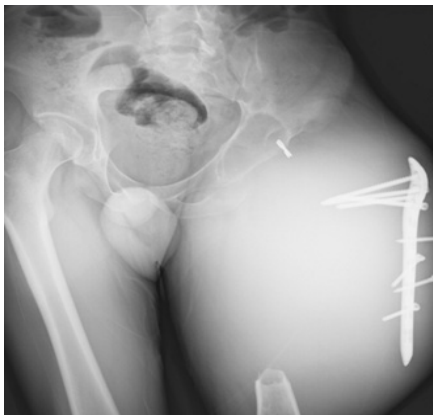

Figure 2f - Direct radiograph 19 months after the second surgery

His physical examination showed a giant mass measuring nearly 70x110 cm extending from the left hip to the femoral region (Figure 3a). Considering the laboratory parameters, the patient did not have any pathologies requiring an acute intervention except low hemoglobin (Hgb) level. According to the direct radiographs and computed tomography (CT) images, the patient had a mass lesion measuring nearly 50x28 cm in the left proximal femur that contained coarse calcifications and destroyed the proximal femur, left iliac wing, left side of the sacrum and superior and inferior pubic rami, and the lesion exhibited heterogenous contrast uptake. There was marked pressure on the left iliac arteries and femoral artery due to the lesion (Figures 3b, 3c). No other foci were detected in thoracic-abdominal CT scans and whole-body bone scintigraphy.

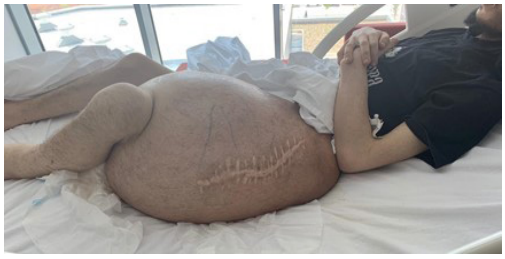

Figure 3a - Clinical view at admission

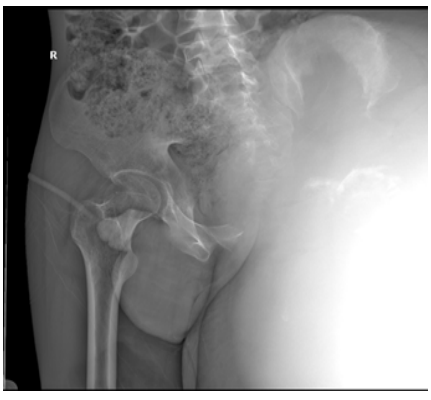

Figure 3b - Direct radiograph at admission

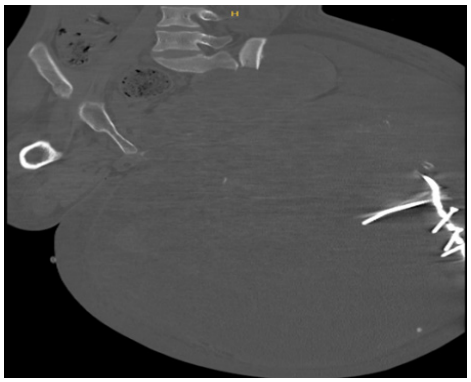

Figure 3c - Computed Tomography image at admission

The patient was admitted to the ward. In order to reduce the fluid component of the mass and facilitate the surgery to be performed, 2 drainage catheters were placed into the 2 deepest points of the mass with the interventional radiology department. In total, approximately 2500 cc of fluid was drained from both catheters. The catheters were blocked, and thus removed on day 3 (Figure 4).

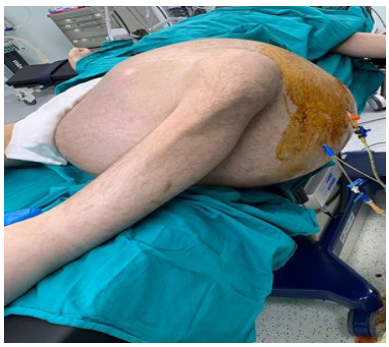

Figure 4 - Placement of drainage catheters

The patient was evaluated by the multidisciplinary tumor council of our hospital and an external hemipelvectomy was planned.

Preoperative embolization was performed on the day of surgery to reduce intraoperative bleeding. The superficial femoral artery, deep femoral artery, common femoral artery, external iliac artery, internal iliac artery, superficial femoral vein, common femoral vein, external iliac vein and internal iliac vein were embolized (Figures 5a, 5b).

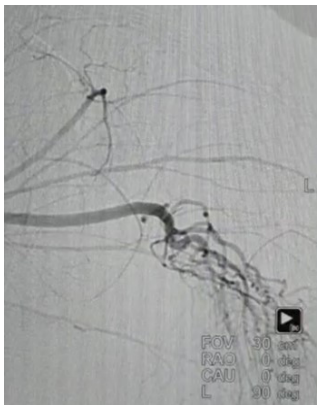

Figure 5a - Embolization procedure

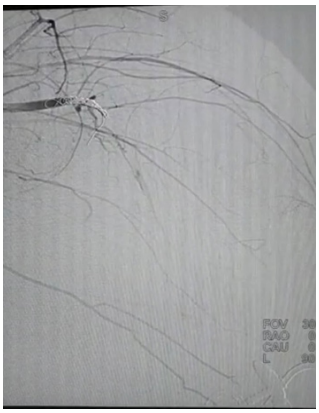

Figure 5b - Embolization procedure

The surgery was performed by a multidisciplinary team consisting of an orthopedist, cardiovascular surgeon, plastic surgeon and general surgeon. The patient was placed in supine position (Figure 6). After anesthesia was induced, the first step was the determination of the flap required for stump closure following external hemipelvectomy by the plastic surgeon. The incision was made and the fluid component and tissue pieces within the mass were removed. In total, approximately 30 liters of fluid, tissue component and fixation material were removed. Distal extremity was moved away from the field. Soft tissues and bone fragments were excised to complete the hemipelvectomy in the proximal aspect. Following bleeding control, 2 Hemovac drains were placed, and stump closure was performed by the

plastic surgery department. The patient was administered 4 units of erythrocyte suspension (ES) and 5 units of fresh frozen plasma (FFP) intraoperatively. The patient exhibited intraoperative hemodynamic instability. Therefore, he was intubated and transferred to the intensive care unit after the surgery.

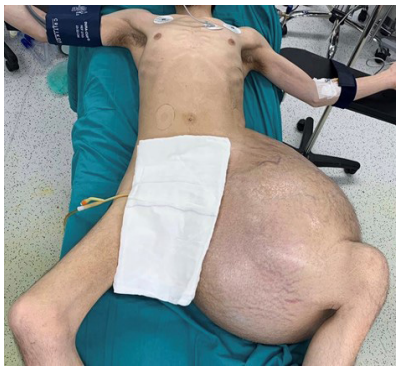

Figure 6 - View of the patient before surgery

During intensive care follow-up, the patient had cardiac arrest at 12 hours postoperatively. CPR was performed and a pulse was obtained at minute 25, after which vital signs became stable. The patient received 6 units of ES replacement during the 3-day stay at the intensive care unit. The patient was then admitted to the Orthopedics Ward. Drainage catheters were followed up for 48 hours and then removed. 1000 cc of hemorrhagic drainage was collected throughout the said 48-hour time period. The patient was hemodynamically stable after admission to the ward and he was mobilized with crutches. According to the histopathological examination, the diagnosis was reported as fibroblastic osteosarcoma (Figures 7a-c).

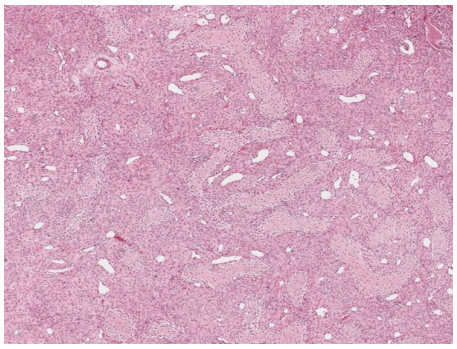

Figure 7a - Diffuse, randomly arranged osteoid formation between spindle cells with malignant appearance (HEX40)

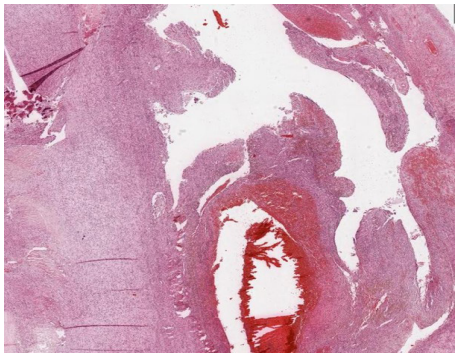

Figure 7b - Cavities that are filled with erythrocytes, adjacent to tumor structures and possessing thin fibrous walls without endothelial lining elements (HEX40)

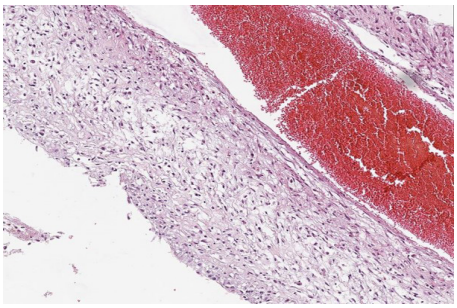

Figure 7c - Previous tru-cut biopsy of the patient. Cystic structures with thin fibrous walls filled with erythrocytes in between regions containing spindle cells (HEX100)

The patient, who was followed up for nearly 90 days at the ward postoperatively, underwent surgery twice for wound site issues and he was mobilized with crutches (Figure 8a). After complete wound healing, the patient could walk with a custom-made prosthetic leg (Figure 8b).

8a *8b*

Figure 8a - Mobilization with crutches following external hemipelvectomy
Figure 8b - Mobilization with a prosthetic leg following external hemipelvectomy

Discussion

ABC is a rare and benign bone tumor. In general, the preferred treatment is composed of opening a sufficiently sized bone window, followed by curettage of the lesion, adjuvant therapies and filling of the defect with a graft or cement. In selected patients, embolization can be performed as the primary treatment or in combination with surgery. Radiotherapy is rarely preferred as it leads to transformation to sarcoma in late follow-up. Malignant transformation of ABC is rare. Particularly, malignant transformation without radiation exposure is considerably rarer [5-7].

In 1991, Kyriakos and Hardy reported an 11-year-old female patient with an ABC located in the distal tibia who underwent proper curettage and adjuvant therapies repeatedly and exhibited transformation to pleomorphic osteosarcoma [3]. Transformation to malignancy occurred nearly 4 years later and the patient did not have radiation exposure. Similarly, in 2012, Anract et al. reported transformation to malignant fibrous histiocytosis that occurred nearly 12 years later in a patient diagnosed with ABC in the distal femur [8]. Brindley et al. presented 2 cases of ABC that exhibited transformation to osteosarcoma without radiation exposure in 2005. Although proper curettage and adjuvant therapies were performed for these lesions located in the proximal humerus and proximal tibia, development of telangiectatic and fibroblastic osteosarcoma was reported respectively in 5- and 12-year follow-ups [6]. Hsu et al. reported transformation to osteosarcoma at the end of year 6 in a patient who was diagnosed with ABC in the proximal tibia and received the treatment protocol [4]. In addition, Kansagra et al. reported malignant transformation to fibroblastic osteosarcoma within 2 years in a patient who was diagnosed with ABC after presenting with pathologic fracture following trauma in the femoral diaphysis [9]. These case reports indicate that routine follow-up is of utmost importance in patients receiving treatment for aneurysmal bone cysts. Patients should be informed about the fact that these follow-ups are not only required to detect disease recurrence but also to check for possible transformation to malignancy.

In various publications, it was reported that 25% to 50% of ABCs could be a component of another bone lesion [10,11]. In a study conducted with 75 patients with osteosarcoma, 8 patients were found to exhibit secondary aneurysmal bone cyst regions [12]. Particularly, the histologic and radiologic similarities between the telangiectaticsubform of osteosarcoma and ABC lead to a failure to diagnose osteosarcoma, which in turn results in patients with osteosarcoma receiving treatment for ABC instead [13].

In a study, Brindley et al. presented two ABC cases with malignant transformation and analyzed the pathology data from previously reported relevant cases [6]. They hypothesized that malignant transformation of the fibroblastic cells remaining after curettage of the ABC was the reason for malignant transformation occurring years after the treatment of ABCs. The fact that the patient presented here exhibited malignant transformation to fibroblastic osteosarcoma is supportive of the thesis that the patient had malignant transformation secondary to an ABC.

Conclusions

In the present case the patient was found to develop a huge tumor and he had history of multiple surgeries. Appropriate surgical management of repeatedly operated a huge tumor like this case is more than a challenge. In such cases, decisions should be made by a multidisciplinary council and interventions should be made by a multidisciplinary team. In this case firstly an embolisation was performed and surgery was performed by a multidisciplinary surgical team.

Ethical aspects. The patient received informed consent.

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Сүйектің аневризматикалық кистасының радиациялық сәулеленудің әсерінсіз алып фибробласты остеосаркомаға алмасуы: дұрыс хирургиялық тактика таңдау — оңай сынақ емес

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Түйіндеме

Бұл қолжазбада біз сол жақ сан сүйегінің проксимальды бөлігінің патологиялық сынуына алып келген аневризмалық сүйек кистасының сирек кездесетін клиникалық жағдайын ұсынамыз. Алып көлемді ісігі бар науқасқа бірнеше рет ота жасалған. Тексеру барысында науқаста радиациялық әсерінен туындамаған остеосаркома дамығаны анықталған. Біз сыртқы гемипелвэктомия жасадық.

Алып көлемді остеосаркомасы бар, бірнеше ота жасалған науқасты емдеуде дұрыс хирургиялық тактика таңдау күрделі мәселе болды. Жалпы алғанда аневризмалық сүйек кисталары патологиялық сынықтар сипатында өте сирек кездеседі. Сонымен қатар, біздің білуімізше, қолжетімді әдебиет көздерінде радиациялық сәуленің әсерінсіз қатерлі ісікке, яғни остеосаркомаға трансформацияланған аневризмалық сүйек кисталарының бүгінде тіркелген жағдайларының саны 10-нан аз болып табылады.

Түйін сөздер: сүйектің аневризмалы кистасы, остеосаркома, қатерлі ісікке трансформациялану, гемипелвэктомия.

Злокачественная трансформация аневризматической костной кисты в гигантскую фибробластную остеосаркому без лучевого воздействия: Надлежащее хирургическое лечение — больше, чем просто вызов

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Резюме

В данной рукописи мы представляем редкий клинический случай аневризматической костной кисты, представляющей собой патологический перелом проксимального отдела левой бедренной кости. У пациента с гигантским образованием в анамнезе было несколько операций. Установлено, что у пациента развилась остеосаркома без лучевой нагрузки. Нами выполнена наружная гемипельвэктомия.

Надлежащее хирургическое лечение пациента с гигантской опухолью, которому неоднократно было проведено оперативное вмешательство - больше, чем просто вызов. Аневризматические костные кисты изначально редко проявляются в виде патологических переломов. Кроме того, насколько нам известно, в доступной литературе количество аневризматических костных кист со злокачественной трансформацией в остеосаркому без радиационного облучения, менее десяти случаев.

Ключевые слова: аневризматическая костная киста, остеосаркома, злокачественная трансформация, гемипельвэктомия.