ISSN: 2640-1037

Clinics of Oncology

Case Report

Distal Radioulnar Joint Replacement with Scheker Prothesis after Enbloc Resection of a Distal Ulnar Aneurysmatic Bone Cyst: Case Report and Review of Literature

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Volume 2 Issue 2- 2019

Received Date: 15 May 2019

Accepted Date: 28 June 2019

Published Date: 06 July 2019

2. Keywords

Aneurysmal bone cyst; Scheker prosthesis; Orthopedics; Tumor

1. Abstract

Even though Aneurysmal Bone Cysts (ABCs) were first described for over 80 years, the actual nature, pathogenesis, as well as the optimal therapy of choice of ABCs remains to be unclear. In this case report we discuss a female patient who presented to our outpatient department for bone and soft tissue related tumors with an aneurysmal bone cyst at the level of the distal ulna. Un-fortunately in her case the lesion was refractory to several minimally invasive attempts and con-tinued to grow in size causing the patient to suffer from increasing pain levels as well as reduced range of motion which subsequently forced her to quit her work. After several presentations at our outpatients department and multiple counselling sessions we agreed along with the patient to perform an en bloc resection of the lesion as well as implanting a scheker prosthesis to provide her with a stable distal radio-ulnar joint (DRUJ). We follow this patient post resection for a period of one year where she reports complete pain relief and a major improvement of her range of mo-tion which in turn elevated her quality of life and allowed her to return to work and lead an active life again. In addition we provide a concise literature review of ABCS touching on the history of ABCs, the radiological findings, pathogenesis, as well as the variable treatment options available for patients and physicians alike.

3. Introduction

Aneurysmal Bone Cyst (ABCs) are a benign cystic tumor that is filled with blood and has a tendency to enlarge and expand which allowing it to present with multiple pictures of symptoms and sometimes disabilities to the patient. It is classified to be growing-related benign tumor that arises mostly childhood and 2nd decade of life in young healthy adults. It is involving mostly long-bone at sites of metaphysis and sometimes with slight ex-tension to diaphysis. Several treatment options were discussed and according to the site, enlargement and disability created by the tumor activity, the treatment option will be depending on those types of factors.

4. Case Report

A young 22 years old female patient who first started having increasing left wrist pain as well as major discomfort during the year of 2014. After several doctor presentations and several x-rays a diagnosis of Aneurysmal Bone Cysts (ABC) at the level of the left distal Ulna was reached. Accordingly, she was referred to another center specialized in treating bone Tumors for further work up of her diagnosis as well as therapy (**Image 1**). Given the age of the patient as well as the size of the ABC at the time of diagnosis an initial attempt was made to treat this lesion using mini-mally invasive therapy by means of alcohol embolization of the Cyst. However, after initiating the therapy, the patient reported no relief of her original complaints rather reported experiencing increasing levels of pain as well as reporting an increased restric-tion of the range of motion of the wrist. With her being unsatis-fied by the results of the initial therapy, the patient consequently presented at our Out Patient Department (OPD) for Bone and soft tissue Tumors for the first time on with a clinically palpa-ble bone cyst almost 10cm in size at the level of her left distal ulna (**Images 2** and **3**). In the physical examination, the patient

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Citation: S Alanaz, Distal Radioulnar Joint Replacement with Scheker Prothesis after Enbloc Resection of a Distal Ulnar Aneurysmatic Bone Cyst: Case Report and Review of Literature. Clinics of Oncology. 2019; 2(2): 1-5.

showed no soft tissue lesions or neurovascular deficits, however she displayed a major restriction of her mobility with a range of motion (ROM) in Extension/Flexion (E/F): 40/0/50°, Pronation/ Supination (P/S): 15/0/15°, as well as Radial Adduction/Ulnar Adduction (RA/UA): 15/0/0 °with instability at the level of the distal radio-ulnar Joint (DRUJ) which forced the patient to quit work. After performing initial X-ray imaging of the lesion (**Im-age 4**) and given the fact that this is a lesion that was refractory to previous therapy and showed aggressive progression and taking into consideration the age of the patient and her level of function we recommended surgical resection of the lesion. After thorough counselling of the patient she rejected surgery and expressed her wishes to wait and see as well as requesting some time to discuss surgery with her relatives. The patient then presented 3 months later with increased lesion size und progressive limitation of her ROM (**Image 5** and **6)**. With radiological evidence of lesion size progression (**Image 7**) we discussed once more the option of surgical excision of the lesion. Eventually, the patient accepted surgery. After performing the surgical planning (**Image 8)** and given the fact that we have to resect around 10cm of the distal Ulna we decided on coupling the resection with implanting a Scheker prosthesis to provide this young patient with a stable and bet-ter functioning DRUJ (**Images 9** and **15**). Immediately after the surgery the patient reported major pain relief and an increased ROM compared to her preoperative status. The patient presented 1 year postoperatively at our OPD for a follow up examination, where she reported major pain relief and a dramatically improved ROM which provided the patient with a better quality of life and allowed the patient to return to work and fulfill her role in soci-ety. Radiologically the X-rays showed no sign of ABC recurrence as well as no loosening of the Prosthesis.Clinically the patient showed an intact soft tissue layer with a dramatically improved ROM E/F: 80/0/80°, P/S: 80/0/80°, RA/UA: 30/0/20° (**Images 15** and **22**).

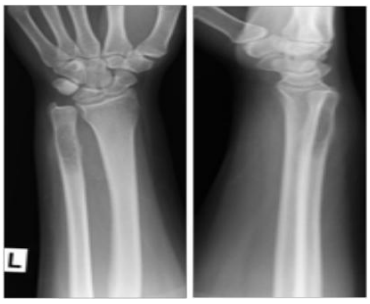


Image 1: X-Ray findings at time of the first diagnosis during October 2014.

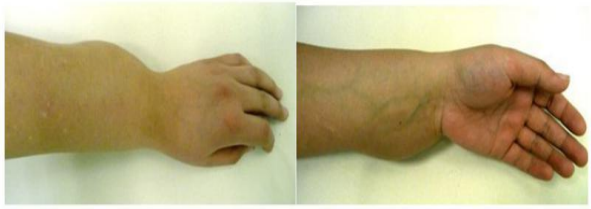


Image 2 and 3: Clinical images of the patient's left ulna during the first presen-tation in our OPD during May 2015.

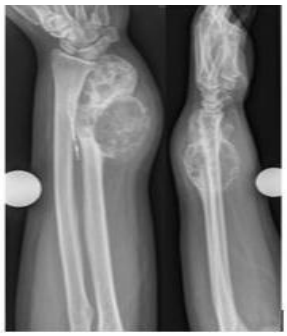
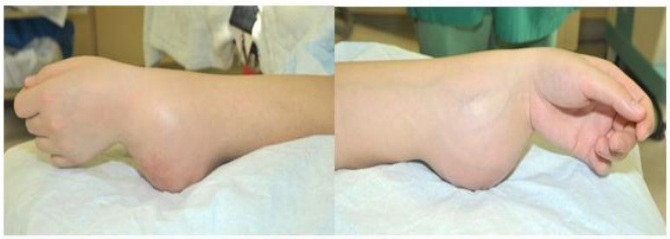


Image 4: X-Ray findings of the patient's left ulna during the first presentation in our OPD during May 2015.



Images 5 and 6: Clinical images of the patient's left ulna 3 months after the first presentation in our OPD.

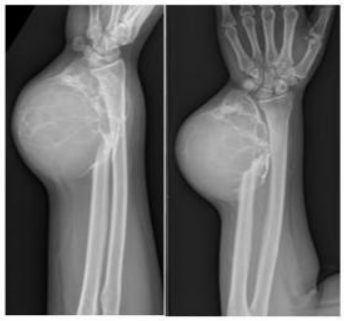


Image 7: Radiological findings of the patient's left ulna 3 months after the first presentation in our OPD.

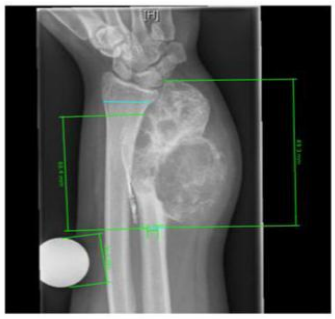
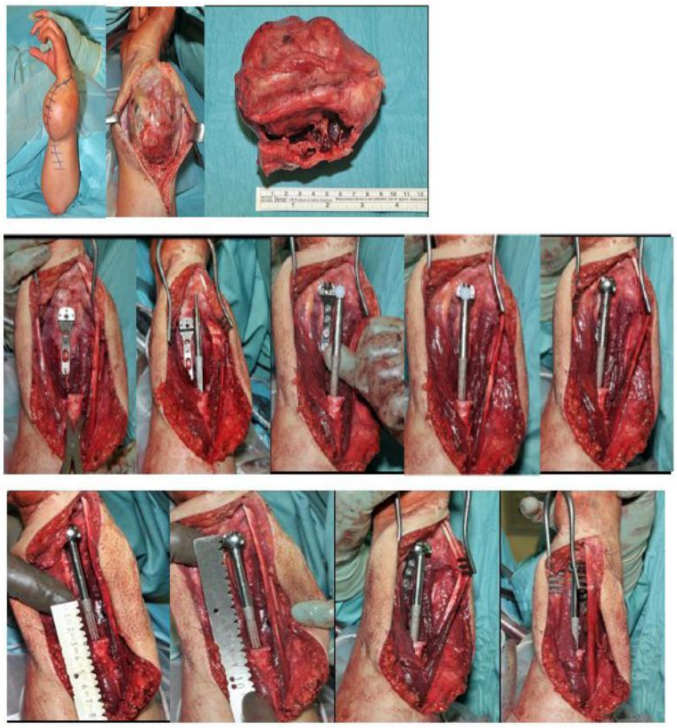
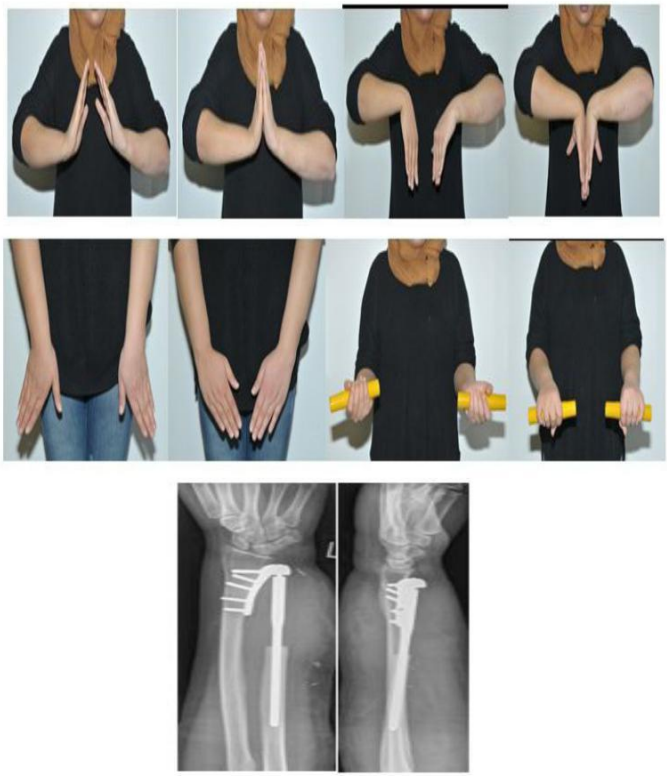


Image 8: Pre-operative planning of the surgical resection of the tumor.



Images 9-15: Intraoperative clinical pictures of the cyst resection and the implantation of the Prosthesis.



Images 16-23: Clinical as well as X-Ray findings during the patient's 1 year fol-low up showing major improvement of the ROM and no radiological signs for loosening of the prosthesis.

5. Discussion

Even though Aneurysmal Bone Cysts (ABCs) were first described for over 80 years, the actual nature, pathogenesis, as well as the optimal therapy of choice of ABCs remains to be unclear. These lesions were initially described by Jaffe and Lichtenstein in the year 1942 as:" Solitary unicameral bone cysts" [1], only to be after several years (1950 and 1957) further defined by the same authors to have first described them making this disease entity the Jaffe-Lichtenstein disease [2-3].

5.1. Epidemiology

ABCs are a rare entity with a general population incidence of 0.14 per 100,000 per year and thus constituting 1-2% of all primary tumors, with a slight female predominance (1:1.16 male to female ratio). ABCs generally occur during the second decade of life with almost 75-90% diagnosed before the age of 20. When diagnosed these lesions seem to arise in isolation and are rarely multiple [4-6]. Moreover, these lesions can either appear as the primary pathology or as a secondary lesion coexisting with other lesions such as nonossifying fibroma, chondroblastoma, solitary bone cyst, giant-cell tumor of bone, osteoblastoma, giant-cell repara-tive granuloma, fibrous dysplasia, and fibromyxoma [7-8].

5.2. Localization

ABCs have been demonstrated to arise in vertebra and the flat bones such as in the bones of the pelvis, clavicula, and ribs; as well as in the long bones of both the upper and lower extremities of the body. At the level of the long bones these lesions seem to commonly appear in the shaft region of the bones with a less common predilection to affect the ends. However, when it comes to flat bones these lesions seem to occur more commonly at the bone end or even close to the surface of articulation [2].

5.3. Symptoms

The more classical chief complaint when it comes to ABCs is generally going to be pain that is in turn aggravated by movements. Moreover, when lesions tend to be at regions with not enough soft tissue coverage patients will present because of an increasing painful swelling that has rather developed insidiously over a long period of time. In addition, when these lesions seem to arise at or close to the articulation surface, patients will then present with a painful gradually increasing reduced range of motion [2].

5.4. X-ray Findings

Typically, ABCs will show an osteolytic and eccentric lesion with a classically described expanded and remodeled "blown-out or "ballooned" bony contour which shows a distinct fluid-fluid level of the host bone; these lesions have been typically referred to as "soap bubble" lesions [8-10].This expanded contour appearance is the result of bone production by the periosteum, stimulated directly or indirectly by underlying pathophysiologic change.

5.5. Pathogenesis

As previously described ABCs can arise as a single primary lesion of the bone or secondary to other primary bone lesions, interestingly, the pathogenesis of primary ABCs seems to greatly differ from those that arise secondary to other bone lesions.

5.6. Primary ABCs

ABCs were traditionally believed to develop as reactive lesions due to the elevated venous pressure which in turn results in expanding bone voids that fill up with blood. However, more recently, primary ABCs have been shown to be associated with a gain-of-function chromosomal translocations: t(16;17) (q22;p13). These in turn result in a gain-of-function mutation of TRE17/USP6 (ubiquitin-specific protease USP6 gene). This mu-tation activates the matrix metalloproteinase (MMP) via NF-kB. Once activated, MMPs would degrade the various components of the extra cellular matrix resulting in the rapid expansion and growth of ABC lesions. It is worth mentioning, that despite the oncogenic activation of the USP6 gene in ABCs, these lesions are thought to display no malignant transformation potential [11].

5.7.Secondary ABCs

ABCs can also arise secondary to other bone tumors such as chondroblastomas, giant cell tumor, chondromyxoid fibroma, non-ossifyingfibromas,and orfibrousdysplasia. These constitute around 30% of all cases of ABCs worldwide. Secondary ABCs are in turn not considered to be neoplasms since they show no genetic translocations or mutations at their origin [12].

5.8. Therapy

As described previously, even though ABCs have been described over 80 years ago the treatment of choice is still unclear and var-ies based on the lesion's location and accessibility. Current man-agement options include surgical as well as minimally invasive non-surgical techniques with variable success, recurrence, as well as complication rates.

5.9. **En-bloc excision**

This is the complete resection of the tumor and lesion. Even though the technique shows the lowest rates of lesion recurrence, it is however associated with a higher morbidity rate (postopera-tive pain, limb length discrepancies, muscle weakness, and de-creased ranges of motion). This is why this technique is reserved to recurrent lesions refractory to less invasive treatment and those lesions in locations in which function is not compromised with such a resection [13].

5.10. Intra-lesional curettage and bone graft

This technique was originally described and utilized by the Jaffe and Lichtenstein the scientists to have initially described these lesions. These suggested means of therapy was curettage and de-fect reconstruction with bone graft. Although still considered to be the main therapeutic option by many, this techniques shows a relatively high recurrence rate ranging between 10-44% [1]. The fact that this initially suggested treatment showed staggering re-currence rates several twists and modifications have been sug-gested to further improve this traditional technique.

5.11. Intra-lesional curettage and High speed burr:

In this technique, after performing the initial intralesional resec-tion of an ABC lesion, a high-speed burr would be used to aug-ment the initial curettage by mechanically disrupting the lesion to the level of the circumscribing bone. Several series have shown control rates ranging between 82 and 90% [14-15].

5.12. Intra-lesional curettage and cement

In this approach, following the initial curettage, polymethylmeth-acrylate (PMMA) cement is introduced in the defect region in-stead of the traditional bone graft in order to provide immediate stabilization for the resultant cavity. In addition, it can act as a recurrence reducing adjuvant through its exothermic effect as the cement hardens. This technique has shown mixed success rates with some series suggesting a recurrence rate of 17% while oth-ers have shown a recurrence rate comparable to the use of bone grafts [16-17].

5.13. Intra-lesional curettage and Phenol application

Phenol, also known as carbolic acid, is produced in mass quanti-ties from petroleum, and it is a precursor to various materials including plastics, pharmaceuticals, and analgesics. In the treat-ment of ABCs, phenols have been used to "sterilize" or wash the lesion, removing remaining neoplastic cells following curettage. Several retrospective studies have shown recurrence rates of up to 7% compared to curettage alone [18].

It is worth mentioning that other non-surgical alternatives have been introduced and employed in the treatment of ABCs some of the more commonly used non-surgical options include.

5.14. Arterial embolization

Selective Arterial Embolization (SAE) can be used as an adjuvant therapy after surgical treatment of ABCs or can even be used as the primary choice of treatment. This approach seems to be fa-vored in cases of ABCs that are difficult for the surgeon to ac-cess or are at a high risk of hemorrhage. Some series have shown a control rates of up to 94% even though some cases required multiple embolizations. This technique is not free of complica-tions. The most frequently reported complications include skin necrosis as well as transient paresis. This approach shows many limitations especially lack identifiable feeding vessels or may be perfused by vessels that also feed nearby vital tissues and organs [19].

5.15. Sclerotherapy

In this approach endothelium of vessels are damaged using Et-hibloc alcoholic solution which in turn triggers the coagulation

cascade resulting in vessel thrombosis. By inducing sclerosis of the ABC's vascular network, local control of the lesion can be achieved. Even though some series have suggested successful control rates of up to 92% (including patients who required multiple sessions), unfortunately this technique has shown a high complications including aseptic bone necrosis, pulmonary embolism, deep venous thrombosis, and cerebellar infarct leading to death [20]. Another sclerosant is Polidocanol (hydroxypolyaethoxydodecan) which is traditionally used by dermatologists for the treatment of varicose veins. Several series have shown response rates of up to 84.5% after multiple injections [21].

6. Conclusion

In conclusion it is important to reemphasize that ABCs are rare bone tumors that affect a younger patient population and show a predisposition to affecting the long bones of the body, and that even though this entity has been first described 80 years ago by Jaffe and Lichtenstein we do not have definitive answers to the questions regarding the actual pathogenesis of ABCs. According-ly, establishing a gold standard therapy is more challenging than expected with variable success rates as well as recurrence rates for both surgical as well as minimally invasive therapies mak-ing ABCs a challenging entity to handle. Accordingly we recom-mend an intensive discussion of all possible therapeutic options with the patient and reaching an agreement that satisfies all par-ties involved.

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