Extraskeletal Mesenchymal Chondrosarcoma of Shoulder: An Extremely Rare Case

Salman Ghaffari[1], Ali Farsavian[1], Seyed Mohamad Mehdi Daneshpoor[1], Masoud Shayesteh Azar[1]

What to Learn from this Article?

We should be careful in open reduction and internal fixation of fracture and also should consider in mind the diagnosis of pseudoaneurysm during plate removal, especially in patients who referred by pain and swelling.

Abstract

Introduction: Extraskeletal chondrosarcoma (EMC) is a rare, aggressive neoplasm which has been seen in the soft tissue area. This soft tissue sarcoma is classified to myxoid and mesenchymal based on histologic criteria. The mesenchymal subtype has a poor prognosis. In approximately 50% of patient with EMC, we could observe soft tissue lesion and stippled calcification in the conventional radiography.

Case Report: In the current paper, we introduced a 47-year-old Iranian male patient having painless, mobile, nontender, and firm mass in left shoulder. We did not find neurovascular disturbance at the upper extremity, and the patient had a full range of motion in the left shoulder. The tumor was treated with wide resection and followed by radiation therapy.

Conclusion: Complete wide resection of mesenchymal chondrosarcoma could be enough as an initial treatment and chemotherapy reserved for patients that have unresectable masses. Apparently, the main key in the treatment is the surgical resection, and this process is the most important method in their management.

Keywords: Mesenchymal chondrosarcoma, Conventional chondrosarcoma, Extraskeletal malignant cartilaginous tumors

Introduction

Extraskeletal chondrosarcoma (EMC) is a rare tumor, which originates in the soft tissue compartment and is classified as myxoid and mesenchymal based on histologic criteria. Myxoid type is more common than the other. In general, mesenchymal subtype included <10% of all types of chondrosarcomas and is more common in the bone [1, 2, 3]. Recent studies have shown that the extraskeletal involvement is to be near 30-40% of these malignancies [1, 4].

At the first time, EMC was described by Lightenstein and Bernstein, in 1959, as a low-grade soft tissue sarcoma, but it had worse prognosis and was too rare [5]. Pathologically speaking, histologic criteria both in bone and soft tissue are the same and specific. To be noted, EMC includes small round blue cells with islands of benign appearing cartilage [1, 5].

Most of these tumors are located in the orbit, cranial, and spinal meningeal coverings and lower limbs, especially in the thigh. The recent researches have shown that involvement of the upper extremity, particularly in

Access this article online

Website: www.jocr.co.in

DOI: 2250-0685.560

Author's Photo Gallery

Dr. Salman Ghaffari

Dr. Seyed Mohamad Mehdi Daneshpoor

[1]Department of Orthopaedics, Orthopaedic Research Center, Mazandaran University of Medical Science, Sari, Iran.

Address of Correspondence
Dr. Masoud Shayesteh Azar,
Department of Orthopaedics, Orthopaedic Research Center, Mazandaran University of Medical Science, Sari, Iran.
E-mail: dr.daneshpoor@gmail.com

Journal of Orthopaedic Case Reports | pISSN 2250-0685 | eISSN 2321-3817 | Available on www.jocr.co.in | doi: 10.13107/jocr.2250-0685.560

the shoulder is extremely rare. We could distinguish EMC from other chondrosarcomas in several features. EMC displays a female preference and occurs in soft tissue area in patients more than 40 years old [6, 7].

In approximately 50% of patient with EMC, we could observe soft tissue lesion and stippled calcification in the conventional radiography [8].

In this paper, we reported a 47-year-old Iranian male patient with painless mass in left shoulder, who had a tumor quite in this location 20 years ago which was diagnosed with an EMC.

Case Report

A 47-year-old Iranian male patient referred to our outpatient clinic in the Imam Khomeini Hospital, Sari, with a palpable and painless mass in his left shoulder.

The patient did not complain about pain or weakness in this location. On examination, the mass was small, mobile, firm, and nontender located in the soft tissue of left shoulder. The skin covering the mass was intact, and there was no ecchymosis and even erythema (Fig. 1). The shoulder range of motion had no functional impairment, and the patient was able to do the routine tasks with no neurovascular disturbance. Medically speaking, important clinical signs and symptoms such as fever, weight loss, and respiratory symptoms were not detected.

There was no visible fluctuation in this field, and it was estimated 0.5-1 cm in length and width. Tumor consistency was not similar to bone, and it seemed to be a soft tissue mass. The patient denied any history of trauma and also no significant family history was identified. Interestingly, the patient was operated by general surgeon 6 times because of relapses in a way the last one was 1.5 years ago. At the first time, the patient was referred to the outpatient clinic because of gradual mass enlargement. The patient was operated by wrong method as transverse approach in the lateral border of the shoulder (Fig. 1).

In the recent recurrence, chest X-ray showed no obvious lung or bone involvement (Fig. 2 and 3). Magnetic resonance imaging (MRI) illustrated a small extraskeletal nodular lesion with ring enhancement and no bone involvement at subcutaneous fat on lateral aspect of shoulder measuring about 13 mm which could be due to a recurrence of the previous tumor (Fig. 4 and 5). Unfortunately, no previous documentation such as MRI was found. Thus, this caused difficulty for interpretation of the course of the main disease.

First, we decided to operate the tumor as an incisional biopsy because we were not sure about the nature of the lesion. An important note in this approach was limited access to the lesion because of an incorrect previous approach. Pathologic specimen confirmed the diagnosis of chondrosarcoma. Then, he underwent a wide resection of the lesion in the beach-chair position. In surgery, the five dense fibrotic and firm masses measuring from 0.5 to 2 cm and the middle part of deltoid muscle (because of involvement) were excised and sent to pathology ward. Microscopic examination showed infiltrative hypercellular islands of pleomorphic chondrocytes composed of enlarged cells with irregular hyperchromatic nuclei and inconspicuous cytoplasm which was compatible with the diagnosis of chondrosarcoma with free surgical margins (Fig. 6 and 7). We should say that the diagnosis was based on

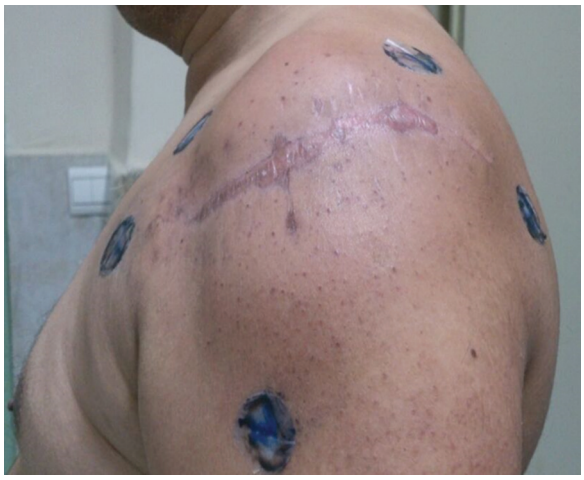

Figure 1: Transverse incision for excision of tumor and irradiation site.

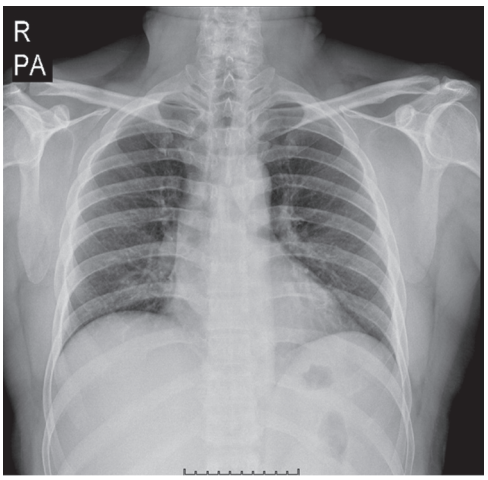

Figure 2: Chest X-ray shows no obvious lung or bone involvement, no calcification, or cortical destruction.

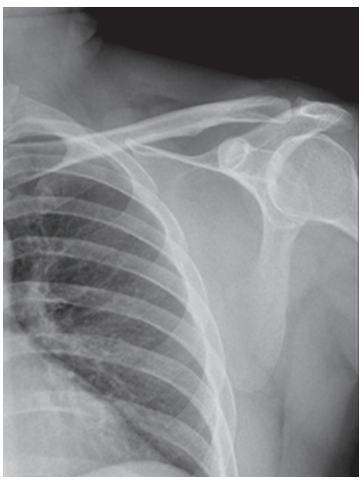

Figure 3: Anteroposterior radiograph of left shoulder reveals no lytic sclerotic lesion, calcification, or cortical destruction.

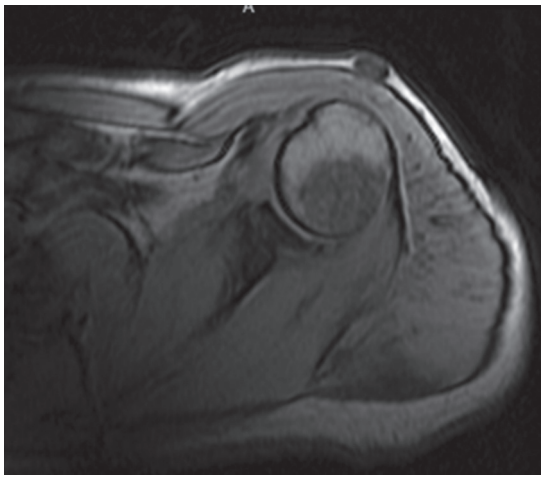

Figure 4: Axial T1-weighted, magnetic resonance imaging of the left shoulder shows an intermediate signal and lobulated mass near the skin.

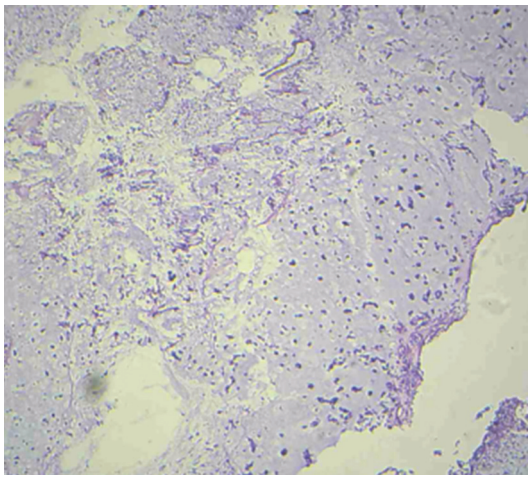

Figure 6: Chondrosarcoma, microscopic examination shows infiltrative hypercellular islands of pleomorphic chondrocytes (H and E, ×4).

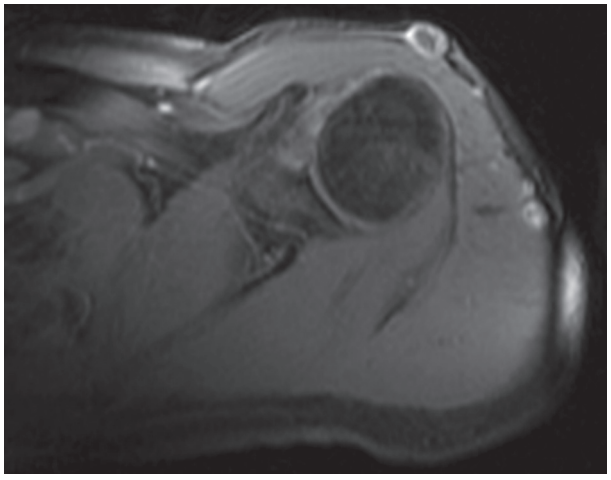

Figure 5: Axial T2-weighted, magnetic resonance imaging of the left shoulder shows a high signal and lobulated soft tissue mass near the skin of left shoulder.

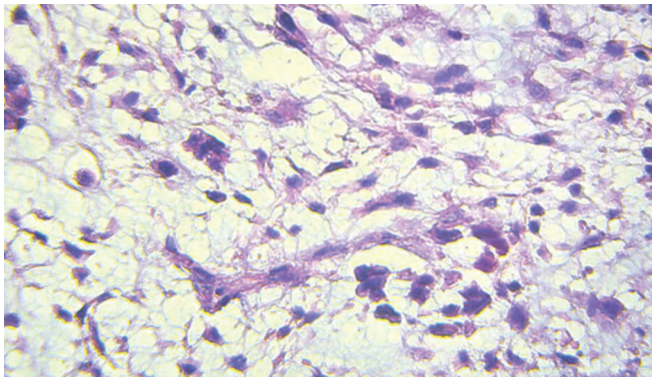

Figure 7: Chondrosarcoma, higher magnification shows enlarged cells with irregular hyperchromatic nuclei and inconspicuous cytoplasm (H and E, ×40).

the characteristic clinical, radiologic, and pathological criteria; moreover, it was confirmed by immunohistochemistry. Hence, after *consulting with the oncologist, radiation therapy was done.* We have followed the patient for 6 months after operation, and until now, no recurrence is recognized.

Discussion

EMC is an aggressive and rare soft tissue sarcoma, which arises from extraskeletal origin in comparison with skeletal type [1, 2, 3, 4]. This cartilaginous malignancy firstly reported by Stout and Verner, in 1953, and histologic properties were quite similar to skeletal ones [5, 9]. These malignant tumors are classified to myxoid and mesenchymal based on histologic criteria. Myxoid type is more common than other. At the first time, EMC was described by Lightenstein and Bernstein, in 1959, as a low-grade soft-tissue sarcoma which was too rare in the upper extremities. The literature review shows that there are just a few cases of this kind of soft tissue tumor; besides, it is very rare in the shoulder area [9, 10].

Overall, mesenchymal chondrosarcomas included <10% of all type of chondrosarcomas and often originates from the skeletal area [5, 6, 9]. In a review literature, only 22% of these arise from extraskeletal areas such as the meninge, brain, and lower extremities and also these malignancies in the soft tissue are rarer than in the bone [6, 10]. Accordingly, the involvement of the shoulder has extremely rare occurrence [7, 8]. EMC could be distinguished from other chondrosarcomas in several features. Despite our case who was male, EMC has a female preference, and this occurs in soft tissue area in patients more than 40 years old such as our case.

In approximately 50% of patient with EMC, we could observe soft-tissue lesion and stippled calcification in the conventional radiography. In our case, we did not see this calcification, and the only point was slight soft tissue swelling in the radiography [6, 7, 8].

The prognosis of this tumor is worse because of the rapid growth, high tendency to metastasis, and more malignant behavior depending on the size, grade, and location of the lesion. The most common site for metastasis of these tumors are lungs, and it has poor prognosis especially pulmonary, bone, and lymph nodes metastases are seen at the time of diagnosis [4, 5, 8, 10].

Chondrosarcoma with soft tissue extension should be treated as high-grade tumor and wide or radical resection or amputation with the aim of complete excision are recommended, but there is no comprehensive and universal information about the effectiveness of chemotherapy and radiotherapy on soft tissue chondrosarcoma [4, 6, 9, 10]. There are some guidelines in this field, but they are appropriate and specific for the skeletal origin. In general, chemotherapy or radiotherapy has not been proved to be effective in soft tissue sarcoma [3, 5]. At the first visit, we decided to operate the tumor as an incisional biopsy because we are not sure about this diagnosis. After confirming the diagnosis of chondrosarcoma, the patient underwent a wide resection. As a final treatment, radiation therapy prescribed based on oncologist consult.

An important point in this patient was frequent recurrences and different kinds of operations that had been done. Interestingly, no evidence of distant metastasis was identified. Now, the patient is free of symptom. Medically speaking, we could say that the previous transverse incision in the shoulder was wrong and made limited access to total tumor excision, and so the delayed suitable treatment took place (Fig. 7).

Conclusion

The current study described a case of mesenchymal chondrosarcoma with frequent recurrences that was treated in our department. Microscopic examination revealed recurrence of the previous cartilaginous tumor. The main reason could be incomplete resection of the previous tumor. In this point, we should remark that the initial operation should be done by an experienced surgeon. This surgeon should be familiar with the tumor nature and final treatment. Finally, complete wide resection of mesenchymal chondrosarcoma could be enough as an initial treatment and chemotherapy reserved for patients that have unresectable masses. Apparently, the main key in the treatment is the surgical resection, and this process is the most important method in their management.

Clinical Message

Treatment of chondrosarcoma with soft tissue extension could be challenging because there are no comprehensive and universal guidelines in this field. On the other hand, our goal should be complete excision of this tumor.

References

1. Wu KK, Collon DJ, Guise ER. Extra-osseous chondrosarcoma. Report of five cases and review of the literature. J Bone Joint Surg Am 1980;62(2):189-194.
2. Anderson JT, Lucas GL. Extraskeletal mesenchymal chondrosarcoma of the forearm: A case report. J Hand Surg Am 2007;32(3):389-392.
3. Bertoni F, Picci P, Bacchini P, Capanna R, Innao V, Bacci G, *et al.* Mesenchymal chondrosarcoma of bone and soft tissues. Cancer 1983;52:533-541.
4. Seo CY, Jung ST, Byun JW. Extraskeletal mesenchymal chondrosarcoma in the axillary region: Reports of two cases. Korean J Pathol 2012;46(5):483-488.
5. Lightenstein L, Bernstein D. Unusual benign and malignant chondroid tumors of bone. A survey of some mesenchymal cartilage tumors and malignant chondroblastic tumors, including a few multicentric ones, as well as many atypical benign chondroblastomas and chondromyxoid fibromas. Cancer 1959;12:1142-1157.
6. Nakashima Y, Unni KK, Shives TC, Swee RG, Dahlin DC. Mesenchymal chondrosarcoma of bone and soft tissue. A review of 111 cases. Cancer 1986;57(12):2444-2453.
7. Louvet C, de Gramont A, Krulik M, Jagueux M, Hubert D, Brissaud P, *et al.* Extraskeletal mesenchymal chondrosarcoma: Case report and review of the literature. J Clin Oncol 1985;3:858-863.
8. Weiss SW, Goldblum JR. Enzinger and Weiss's Soft Tissue Tumors. St. Louis: Mosby; 2001. p. 1380-1385.
9. Stout AP, Verner EW. Chondrosarcoma of the extraskeletal soft tissues. Cancer 1953;6(3):581-590.
10. Nguyen DV, Muda AS, Yaacob Y. Mesenchymal chondrosarcoma: A case report. Malays J Med Sci 2013;20(3):71-77.

Conflict of Interest: Nil
Source of Support: None

How to Cite this Article

Ghaffari S, Farsavian A, Daneshpoor SMM, Azar MS. Extraskeletal Mesenchymal Chondrosarcoma of Shoulder: An Extremely Rare Case. Journal of Orthopaedic Case Reports 2016 Sep-Oct;6(4): 35-38.