

Scapula chondrosarcoma

A case report

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Abstract

Rationale: Chondrosarcoma is a malignant mesenchymal tumor originating from cartilage. The pelvis, ribs, femur, and humerus are the most frequently affected sites, and scapula involvement is relatively rare. The aim of the present study was to report a case of chondrosarcoma in the scapula.

Patient concerns: A 42-year-old woman presented with a 3-month history of a painful mass in the right scapula.

Diagnoses and intervention: The patient underwent tumor resection. The post-operative pathological diagnosis was scapula chondrosarcoma.

Outcomes: Following resection, the patient continued to receive routine follow-up care. There was no recurrence or tumor metastasis at a follow-up of 5 years.

Conclusions: Surgery remains the primary therapy for chondrosarcoma. One of the greatest challenges in the management of chondrosarcoma is to accurately assess tumor grade before surgical intervention. Chemotherapy and radiotherapy have been applied without success. Chemo- and radioresistance have been examined beyond classic phenotypic properties to identify more efficient therapeutic strategies. Therefore, development of future novel therapies is contingent upon elucidating the molecular mechanisms of chondrosarcoma.

Abbreviations: CT = computed tomography, ECT = emission computed tomography, FDG = fluorodeoxyglucose, MRI = magnetic resonance imaging, PET = Positron emission tomography, SUV = standardized uptake value.

Keywords: chondrosarcoma, scapula, tumor

1. Introduction

Sarcomas are uncommon mesenchymatous tumors comprising <1% of all cancer types that are present in the cartilage, endothelium, muscles, and support structures and are divided into two groups: bone and soft-tissue sarcoma.^[1] Chondrosarcoma is the second most common primary bone malignancy after osteosarcoma and is a slow-growing malignant tumor characterized by the formation of cartilage, not bone, by tumor cells.^[2] Chondrosarcomas originate from embryogenic cartilaginous rests and may arise in any region where cartilage is present, which has been extensively reported in the literature.^[3] Involvement of the scapula is relatively rare. Adults over 30 years

of age are more likely to suffer from it. Surgical resection is the key treatment for chondrosarcoma because this tumor is resistant to chemotherapy and radiotherapy.^[4] Prognosis depends largely on tumor grade. Widely recognized criteria for grading are those described by Evans et al.^[5] Currently, the clinical course, histogenesis, cytogenetics, and prognosis of chondrosarcoma remain largely unknown. Treatment for this tumor relies on the outcome of surgical resection, and the prognosis for patients with chondrosarcomas is good when tumors are completely resected.

Therefore, the objectives of this case were to report on a relatively rare case of chondrosarcoma of the scapula in a middle-age woman with typical pain and tumefaction and to attract public concern on this subject.

2. Ethical statement and consent

The institutional review board and ethic committee of Plastic Surgery Hospital, Chinese Academy of Medical Sciences and Peking Union Medical College approved the ethical, methodological, and protocol aspects of this investigation. The ethical approval number was 2019.06. We confirm that all methods in the present study were carried out in accordance with the relevant guidelines and regulations. Informed written consent was obtained from the patient for publication of this case report and accompanying images.

3. Case presentation

A 42-year-old female presented to our hospital with complaints of a growth of a tumor on her right scapula persisting for 3 months. This was her first episode of such tumor which had

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The authors declare that they have no competing interests.

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gradually increased in volume, beginning with pain. Physical examination revealed a large 5.0 × 5.0 cm soft-tissue mass with areas of maceration. The mass was firm and tender on palpation. There was no dilatation of the superficial vein or swelling of the ipsilateral limb.

Further checks were performed after admission. The results of laboratory tests, including complete blood count, electrolytes, biochemical profile, and coagulation function, were all normal. Subsequent computerized tomography (CT) scan of the right shoulder showed irregularities with cystic low density areas, and the lesion had expansive developing tendency. The shoulder joint and space appeared normal, with no interruption of bone continuity. Emission computed tomography (ECT) scanning indicated a clear bone image, while anomalous concentration of radioactivity was observed in the right scapula (Fig. 1). The chest radiograph exhibited a normal appearance. However, incisional biopsy was not performed before the surgery. The operation proceeded with a posterior approach to the right shoulder joint. A horizontal incision was first performed, and the soft tissue was separated until the upper end of the scapula appeared. The bone wall was destroyed; thus, bone was curetted. The intraoperative pathological diagnosis was cartilage derived lesion. Subsequently bone grafting was performed. Macroscopically, the tumor presented as a pale amyloid tissue, and histologically, it was characterized by the presence of large amounts of chondrocytes and the nuclear atypia was mild with normal cartilage lacuna. Besides, there was no mitotic activity and no mucoid degeneration. Anatomopathological examination indicated that this was a chondrosarcoma, grade IIa.^[6] Safe margins were confirmed negative by multiple border frozen sections. The patient's vital signs and other monitored parameters were normal after returning to the ward, and she was discharged 7 days after

surgery in stable condition. There was no recurrence or tumor metastasis at a follow-up of 5 years.

4. Discussion

Chondrosarcoma accounts for 20–25% of all bone sarcomas, most commonly involving not only pelvis, but may also the diaphysis regions of long bones.^[7] Chondrosarcoma has an indolent natural history, the clinical appearance of it is usually nonspecific. Pain is a common sign, which worsens as the disease progresses. Few patients are admitted to the hospital following appearance of a mass and patients typically experience gradually increasing tumefaction and pain. The clinical behavior of chondrosarcoma is greatly variable and is linked to histologic grading. The etiology of chondrosarcomas remains unclear, and their clinical management is controversial. For this reason, physical examination and clinical history taking are fundamental steps in diagnostic reasoning.

CT is one of the primary diagnostic imaging modalities, which is used to detect the characteristics of this lesion, and showing expansive, osteolytic destruction, soft tissue mass, calcification, and irregularities with cystic low density areas.^[8,9] These typical CT findings of chondrosarcoma were consistent with our report. The possible reasons are:

- (1) cartilage matrix formed by tumor possessed high water content, which would present calcification in peripheral;
- (2) tumor invasion could appear irregular cystic shadows.

We arranged CT and ECT for the patient which demonstrated irregular shapes of the scapula and anomalous concentration of radioactivity. On one hand, ECT has high sensitivity to chondrosarcoma which indicates that radioactivity could aggregate

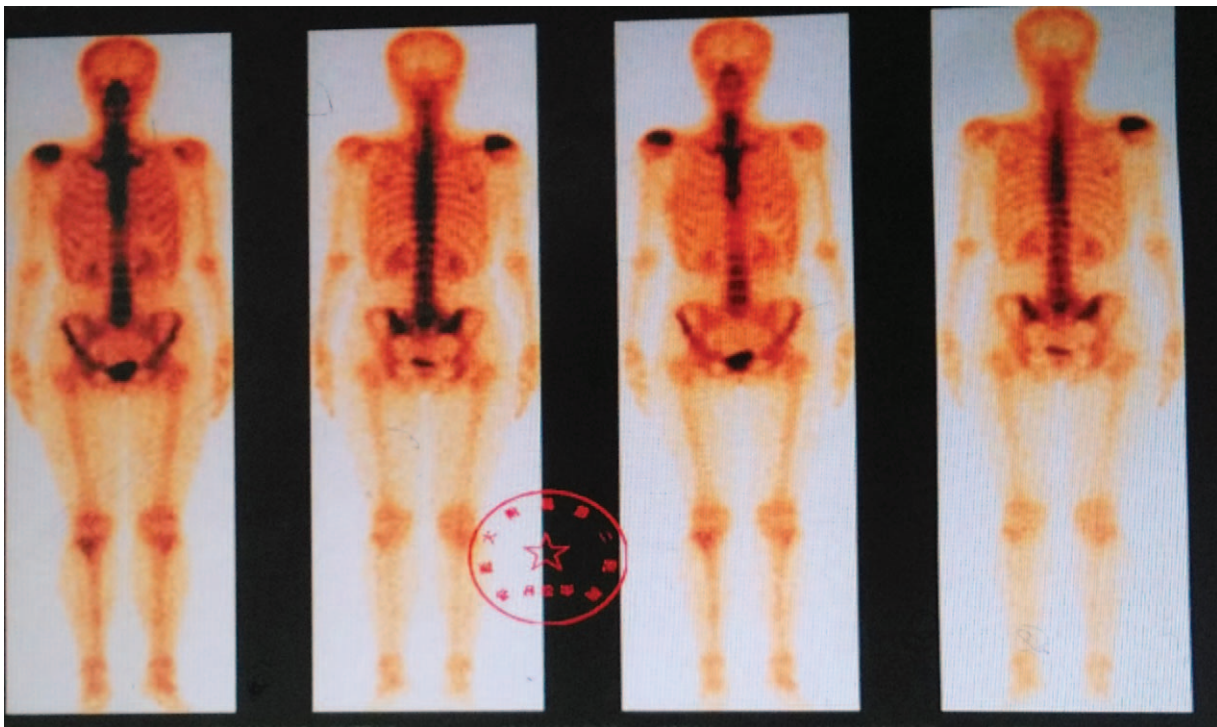


Figure 1. Emission computed tomography (ECT) scanning indicating a clear bone image in which anomalous concentrations of radioactivity were observed in the right scapula.

in tumor locations and tumor imaging can be clearly observed; on another hand, ECT can demonstrate the extension and basic pathological changes of chondrosarcoma accurately, and provide much more important imaging information for surgical operation.^[10] What's more, changes of tumor imaging with ECT is early than the clinical symptom or X-ray, which has an important value of early finding and early diagnosis of chondrosarcoma.^[11] All the hints guided us to the diagnosis of the tumor. MRI is optimal for delineating soft tissue involvement, and due to high water content in the matrix, tumor show very high signal intensity on T2-weighted images, which are an assessment of whether tumor cells have invaded the soft tissue to guide surgical treatment.^[12] Positron emission tomography (PET) scanning with 18-fluorodeoxyglucose (FDG) is unreliable when distinguishing benign from malignant chondrogenic lesions, identifying metastatic disease, or distinguishing recurrent tumors from postoperative changes due to the low metabolic activity of low-grade tumors. However, PET can be useful in identifying high-grade chondrosarcomas that show increased standardized uptake values (SUVs) due to higher glucose metabolism.^[13] It is challenging to form a pre-operative diagnosis for chondrosarcoma due to its atypical symptoms and imaging manifestations; therefore, diagnosis primarily depends on post-operative pathological examination.^[14] During the operation, the anatomopathological examination of the tissue indicated that this was a chondrosarcoma, grade IIa. So we performed a curettage with bone grafting after inactivating the tumor cells with absolute alcohol. Histological examination of Tru-cut biopsy is the only method that can confirm the diagnosis and differentiate this tumor from other diagnoses. Definitive diagnosis can be established by incisional biopsy and histopathological evaluation. Fine needle aspiration biopsy is a safe and cost-effective procedure if the lesion is accessible; if used in accompaniment with radiographic and clinical results, high diagnostic accuracy may be achieved. However, it is difficult and controversial to render the final diagnosis due to low levels of inter- and intraobserver reliability of histological grading. Furthermore, the intrinsic histological heterogeneity of chondrosarcomas makes diagnostic biopsies less reliable. Therefore, emphasis should be placed on imaging studies to complement diagnosis, determine the need for surgery and establish follow-up. It is more valuable to perform an imaging series over time to assess changes and determine tumor behavior rather than performing a single study.

Surgery is the treatment of choice once malignancy is confirmed, and tumors should be completely gross resected whenever possible. Complete resection can cure classic lesions, avoiding the need for adjuvant radiation.^[15] Wide surgical resection is the most effective treatment for chondrosarcoma. For tumors similar to this, local recurrence is more common than distant metastasis, and there is a relatively high association between tumor grade and prognosis. Tumor grade and resectability are the most important prognostic factors for chondrosarcoma. These factors make early diagnosis and complete tumor resection extremely vital for patient prognosis. Radiotherapy for chondrosarcoma is generally utilized as an adjuvant therapy in cases of residual disease rather than as the initial treatment. Chemotherapy as a neoadjuvant treatment inhibits tumor growth and progression. However, it is not beneficial for improving long-term survival or attenuating distant metastasis.

To avoid occurrences of metastatic foci and spreading into neighboring tissues, the first surgical intervention is aimed at resection that is sufficiently wide to prevent local recurrence. In

this regard, it is necessary to set surgical margins of 4 cm on all sides, resulting in cure rates for almost all patients and 10-year survival of 97%.^[16] The incidence of metastasis and disease survival is dependent on histological grade, and local recurrence is determined by the adequacy of surgical margins.^[6] For safety, we separated the soft tissue until the upper end of the scapula appeared in order to deliver a thorough resection. Other reported prognostic factors for disease specific survival include local recurrence, which occurs due to inadequate margins.^[17] Greater tumor size and narrow surgical margins are risk factors for local recurrence. Recurrence is often associated with tumor progression, but what remains unknown is what comprises an adequate surgical margin in high grade chondrosarcoma.^[18] The width of a "wide" margin has never been accurately defined, and discrepancies exist in the practical definition of a wide margin.

The current clinical challenge is to prevent recurrence and to offer new treatment options for patients. A promising new option for improving cancer therapies has appeared that is based on targeting the tumor microenvironment.^[19] Donia et al^[20] conducted research of the structure-activity relationship of hypoxia-activated prodrugs for proteoglycan-targeted chemotherapy in chondrosarcoma and obtained good results. In another study, Peterse et al^[21] found that targeting glutaminolysis with CB-839, metformin, phenformin, or chloroquine is a potential therapeutic strategy for a subset of high-grade chondrosarcomas, irrespective of the presence or absence of IDH1/2 mutation. In addition, combination therapy of mTOR and MEK inhibitors could be an another effective therapeutic approach.^[22,23] Yang et al^[24] support the application of immune checkpoint blockade in chondrosarcoma using PD-L1/PD-L2 expression and Ki-67.

Chondrosarcoma usually invades and destroys adjacent hard tissues, involving and enveloping neighboring soft tissues as well. On pathology slides, chondrosarcoma is usually identified by the presence of malignant chondroid tissue, presenting with hypercellularity, binucleate cells, atypical nuclei, multiple cells in lacunae, and myxoid changes in the hyaline cartilage matrix. Chondrosarcoma usually presents as a malignant tumor composed of fully developed cartilage without tumor osteoids, being directly formed from a sarcomatous stroma. Myxoid changes, calcifications, and ossifications may be seen.^[25] Chondrosarcoma is histologically graded into classes 1, 2 and 3 according to its architectural and cytological atypia.^[26] This grading system is of great importance because it reflects a prognosis based on tumor biology distinct from its location or stage of presentation. While, currently, we cannot completely depend on grading to predict patient outcome, histological assessment sometimes has significant interobserver variability.

Surgery for chondrosarcoma exhibits a 10-year survival rate between 30% and 80%, depending on the grade.^[27] Risk factors, such as grading, metastatic disease, age, and location, significantly influence overall survival.^[28] In a recent study,^[29] CRP was exploited for clinical prediction of this disease. Owing to the possibility of local recurrence and metastasis, resected patients should undergo routine lifelong surveillance, consisting of clinical and radiological examination every 3–6 months for the first 5 years and annually thereafter for a minimum of 15 years. Our patient was diagnosed as grade IIa, so we required her to take a radiological examination every 6 months in our hospital for a healthy concern. And every time when she got the reports, we would take some other tests, which lasted for 5 years with a relaxed result. And we will keep an eye on her healthy conditions

in the following time. Because chondrosarcoma is often slow growing, local recurrence and pulmonary metastasis may not be detected until years, or even decades, after the primary procedure. Local recurrence is the primary cause of death. And lungs, liver, and brain are the common sites of metastasis in this tumor. Other factors influencing prognosis are stage at diagnosis, histopathological grade of the lesion (poor prognosis for high-grade tumors), location, and particular histological subtypes. Long-term follow-up is recommended due to high recurrence rate and distant metastasis. Evans et al reported that 5-year survival rates of grades I, II, and III are 90%, 81%, and 43%, respectively.^[6] Therefore, long-term regular follow-up is needed to ensure that any recurrence or metastasis is diagnosed early and treatment is promptly initiated.

Considering its nature, we arranged a series of tests for the patient when she came to our department. And finally, it was confirmed as malignancy so we performed a thorough resection and scheduled a close follow-up. Fortunately, 5 years have elapsed since the surgery, there have not been any recurrence or metastasis. We will continue to pay a serious attention to follow-up care.

5. Conclusion

Overall, chondrosarcoma is the second most common primary bone tumor and the most common in adults. The primary therapy for chondrosarcoma remains surgery. One of the greatest challenges in the management of chondrosarcoma is to accurately assess tumor grade of the tumor before surgical intervention. Chemotherapy and radiotherapy have been used without success. Chemo- and radioresistance need to be elucidated beyond classic phenotypic properties to identify efficient therapeutic strategies. Therefore, development of future novel therapies is contingent upon an improved understanding the molecular mechanisms of this disease.

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Author contributions

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