
CASE REPORT

Dedifferentiated Chondrosarcoma in the Scapula

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ABSTRACT

Dedifferentiated chondrosarcoma is a rare but highly malignant form of chondrosarcoma with grave prognosis. Early and accurate diagnosis followed by radical surgery seems the only treatment to improve survival. We report a case of dedifferentiated chondrosarcoma in the scapula of a 55-year-old woman presenting with non-specific left shoulder pain. Radiography, computed tomography, and magnetic resonance imaging showed an expansile osteolytic lesion with cortical erosion and internal punctate calcification involving coracoid and glenoid processes. Soft tissue components of the lesion appeared to have invaded the rotator cuff muscles. Computed tomography-guided biopsy confirmed the diagnosis and the tumour was excised with the shoulder joint. Histopathology of the specimen demonstrated intramedullary tumour showing conventional chondrosarcoma admixed with a malignant fibrous histiocytoma-like spindle cell sarcoma. The features were compatible with dedifferentiated chondrosarcoma arising from a conventional chondrosarcoma. Having reviewed the clinical and radiological features of dedifferentiated chondrosarcoma, we stress the importance of radiological imaging features to identify tumour bimorphism suggestive of the diagnosis, and select the site to biopsy.

Key Words: Bone neoplasms; Cell differentiation; Chondrosarcoma; Prognosis; Scapula

中文摘要

肩胛骨的去分化型軟骨肉瘤

譚枝慧、覃天笙、李仲啓、張文鳳

去分化型軟骨肉瘤是一種罕見的高惡性度腫瘤，預後差。盡早為病人作出準確的診斷，繼而進行根治性手術似乎是唯一可改善存活期的方法。本文報告一名55歲肩胛骨去分化型軟骨肉瘤女性患者，病發時有左肩胛骨疼痛。放射檢查、CT及磁共振影像均發現膨脹性溶骨灶，並有皮質中斷及牽涉鳥口突起及關節盂的內部小點狀鈣化灶。而病灶的軟組織成分似乎經已侵佔肩袖肌。經CT引導下穿刺確診，並把腫瘤及肩關節一併切除。病變組織學顯示髓內腫瘤切除樣本有一般軟骨肉瘤的特徵，並呈類似惡性纖維組織細胞瘤的梭細胞癌。這些症狀都符合由傳統軟骨肉瘤演變成去分化型軟骨肉瘤的特徵。回顧去分化型軟骨肉瘤的臨床及放射學特徵，可以知道認識雙相腫瘤的放射學徵狀有助確診病情及選擇穿刺的位置。

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Submitted: 13 Jun 2011; Accepted: 19 Jul 2011.

INTRODUCTION

Dedifferentiated chondrosarcoma (DDCS) is an uncommon subtype,¹ which is considered to be the most malignant form of chondrosarcoma with a notoriously grave prognosis, despite radical operative treatment. The five-year survival ranged from 10.5 to 24% in the recent retrospective study by Grimer et al.² Median survival rates ranged from 5 to 18 months. We report on a 55-year-old Chinese woman with such a scapular chondrosarcoma.

CASE REPORT

A 55-year-old woman with a history of medically treated thyrotoxicosis presented to our orthopaedic department in April 2007, because of sudden-onset left shoulder pain for one month. She had no feverishness or other constitutional symptoms. Physical examination revealed exaggeration of pain with movement but no mass lesion. Initial radiographs in April 2007 showed an osteolytic lesion with a wide zone of transition involving the left coracoid and glenoid processes with bony expansion and cortical erosion (Figure 1a). Internal amorphous and punctate calcification was also present. Contrast-enhanced computed tomography (CT) revealed an irregular expansile destructive apparently osteolytic lesion with irregular internal punctate calcification, cortical erosions, and an enhancing soft tissue component (Figure 1b). In T1-weighted magnetic resonance images (MRI), the lesion was low to intermediate in signal intensity; T2-weighted fat-saturated images showed high-signal intensity with low-signal septations and small irregular punctate area of low-signal compatible with calcification (Figure 2). T1-weighted post-contrast images showed mild gadolinium enhancement of the lesion's osseous part and intense enhancement of the soft tissue component, which appeared to have invaded the subscapularis, supraspinatus, and infraspinatus muscles. No invasion into the axillary neurovascular bundle was evident. A whole body bone scan involving Technicium 99m bisphosphonate showed markedly increased tracer uptake in the left coracoid process extending to the glenoid region, where there was moderately increased uptake. CT-guided biopsy of the lesion confirmed the diagnosis of DDCS. CT thorax did not reveal any lung metastasis. The tumour was excised with the shoulder joint. A free fibula graft for shoulder arthrodesis was performed with a cancellous bone graft (harvested from iliac crest) to fill the gap between clavicle and the fibula graft.

Macroscopically, the surgical specimen appeared to be a chondroid tumour expanding the coracoid process with destruction of anterior and posterior cortex (Figure 3). Its pinkish white fleshy tissue arose from the periphery of the tumour and seemed to have invaded into extra-osseous soft tissue. Microscopically, the intramedullary component of the tumour showed conventional chondrosarcoma (grade 1-2), admixed with a malignant fibrous histiocytoma-like spindle cell sarcoma (Figure 4). The latter appeared to have invaded through the cortex into adjacent muscle. Its resection margin was clear. The overall picture was compatible with a DDCS arising from a conventional chondrosarcoma.

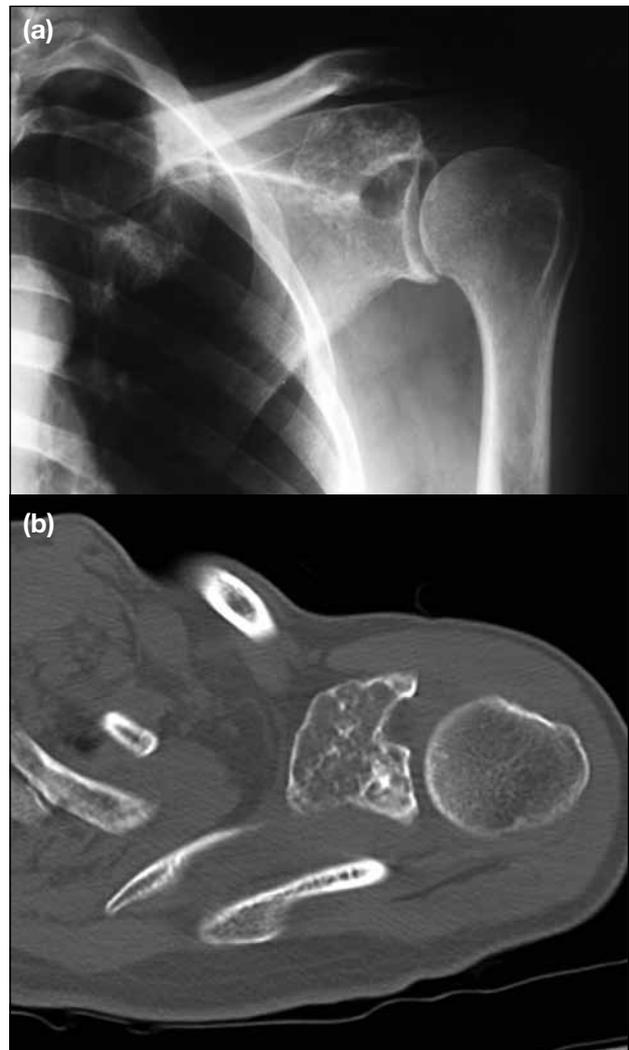


Figure 1. (a) Radiograph and (b) axial computed tomographic image in a bone window of the left shoulder showing an osteolytic lesion with a wide zone of transition involving the coracoid and glenoid processes leading to bony expansion. The images also reveal internal amorphous punctate calcifications and cortical erosion.

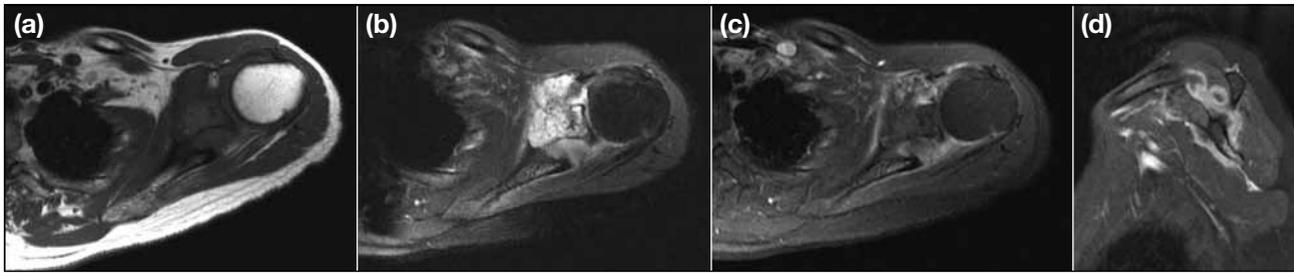


Figure 2. Magnetic resonance images of the lesion involving left coracoid and glenoid process. (a) A T1-weighted axial image shows the low-to-intermediate signal of the lesion. (b) A T2-weighted fat-saturated axial image of the lesion shows the high-signal intensity with low-signal septations and a small irregular punctate area of low signal compatible with calcification. (c) A T1-weighted fat-saturated post-contrast axial image shows mild gadolinium enhancement of the osseous part of the lesion and intense enhancement of the soft tissue component. (d) A T1-weighted fat-saturated post-contrast coronal image demonstrates soft tissue components of the lesion invading the subscapularis, supraspinatus, and infraspinatus muscles.

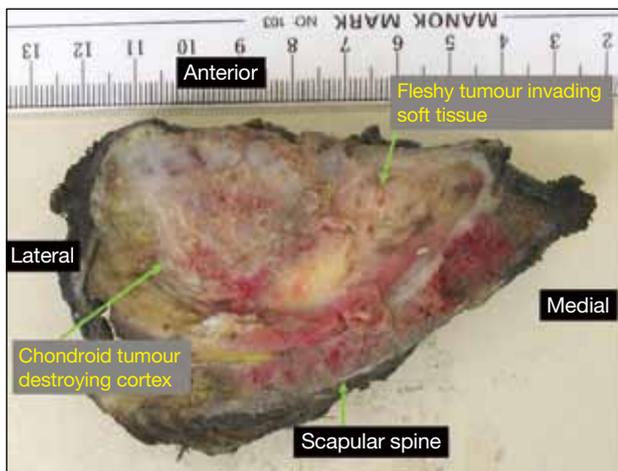


Figure 3. Gross pathology in coronal section of the specimen shows chondroid tumour appearing to expand the coracoid process with destruction of the anterior and posterior cortex. Pinkish white fleshy tissue appears to arise from the periphery of the tumour and invade into extra-osseous soft tissue.

Radiotherapy was given for local disease control. Postoperative chemotherapy was not offered, as such treatment has toxicity and confers limited survival benefit. The patient was found to have multiple pulmonary metastases one year after initial presentation. About four months later, the patient was diagnosed to have brain metastasis and received whole brain radiotherapy. Subsequently, the patient received conservative treatment and succumbed by the end of 2008.

DISCUSSION

DDCS is a special subtype observed in 11% of such tumours. It was first described by Dahlin and Beabout in 1971,³ and typically affects subjects in their fifth to sixth decade, but has no sexual predilection.^{1,4-7}

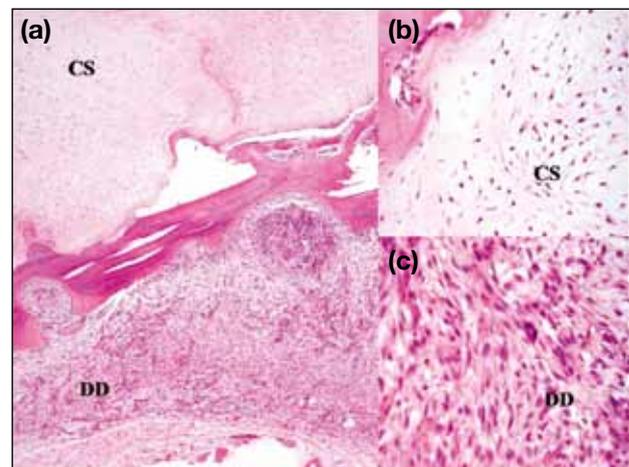


Figure 4. The microscopy sections are stained with haematoxylin and eosin. The histopathological specimen demonstrates intramedullary components of the tumour showing conventional chondrosarcoma (CS) grade 1-2, admixed with a malignant fibrous histiocytoma-like spindle cell sarcoma. The latter appeared to have invaded through the cortex into adjacent soft tissue. Overall, the picture was that of dedifferentiated (DD) chondrosarcoma arising from a conventional chondrosarcoma. Magnification for (a) x 20, (b) x 200, and (c) x 400.

DDCS has two distinct histological components: one consists of a high-grade non-cartilaginous sarcoma arising from the other component consisting of a pre-existing low-to-intermediate-grade chondrosarcoma. The non-cartilaginous tumour can be any mesenchymal tumour of high-grade malignancy, with a differential diagnosis that includes malignant fibrous histiocytoma (MFH), osteogenic sarcoma, fibrosarcoma, leiomyosarcoma and spindle cell tumour. Different case series have reported slightly differing patterns of the dedifferentiated components. Osteogenic sarcoma is by far the commonest dedifferentiated component.^{1,4,6,8} Most studies reported no survival difference between patients with different dedifferentiated components.

Staals et al⁸ indicated poorer survival with the MFH subtype. DDCS was also observed to arise from osteochondroma, other than conventional chondrosarcomas. In that particular series of 18 patients reported by Staals et al,⁸ the survival of DDCS arising from osteochondroma was similar to those associated with conventional chondrosarcomas.

The femur is the commonest site of involvement in DDCS, and is noted in about half (46-55%) of all patients. The pelvis is the second commonest site and involved in 23 to 28% of cases. Pathological fracture is encountered in about 20 to 30% of cases, and is associated with a high rate of amputation. Among all the reported cases, the scapula is a rare site for DDCS noted in about 5% of all cases.⁴⁻⁸

Radiologically, DDCS typically exhibits tumour bimorphism with a chondroid tumour adjacent to a markedly different area, such as a dominant region of increased opacity within the mineralised tumour, a large unmineralised soft tissue mass, a different pattern of mineralisation, or a combination of these findings.¹ The most common radiographic feature of bimorphism is a dominant area of hyperlucency within the chondroid tumour. In CT, a large unmineralised soft tissue mass adjacent to the cartilage tumour or a distinct area devoid of chondroid calcifications can be observed. Similar tumour bimorphism can be demonstrated in MRI, including a large unmineralised soft tissue mass and a dominant area without obvious mineralisation, either alone or in combination with a soft-tissue mass. Such bimorphism is seen in about 35% of radiographs, 48% of CT, and 33% of MRI. The major radiological feature for tumour bimorphism was not clearly demonstrated in our case. Other radiological findings of DDCS are those of an aggressive bony lesion with cortical destruction and a soft tissue component.

The diagnosis and treatment of a scapula tumour may be delayed due to its rarity compared to non-tumourous shoulder conditions (such as rotator cuff disease) that are far more common. In the management of shoulder pain, patients with persistent symptoms unresponsive to conventional treatment should be carefully reviewed with radiographs, to facilitate early detection of underlying bone tumours. Cross-sectional imaging (CT or MRI) should be performed early for further characterisation. In particular, chondrosarcoma should be considered in patients with a primary scapular tumour. In the series of coracoid process tumours of

the scapula reported by Ogoose et al,⁹ chondrosarcoma encountered in eight patients and DDCS three of the 18 cases. In this clinical context, the radiologist should look for features of tumour bimorphism, which is the cardinal radiological feature in about half the cases of DDCS. In addition, when performing image-guided biopsy, tumour bimorphism should be appreciated so as to avoid a sampling error. Both the juxta-chondroid lytic component as well as adjacent soft tissue should be included to maximising the chance of sampling the dedifferentiated component.

Due to their rarity, only few large case series are published with regard to treatment options. Radical surgery with complete surgical excision remains the primary treatment. Local disease control is satisfactory with complete excision. Some authors consider scapula tumour to be more amendable to surgery, as generous soft tissue resection without limb amputation is often achievable.¹⁰ Besides, brachial plexus and axillary nerves are not easily compromised by scapula tumours. The subscapularis muscle is considered to be an effective barrier to subscapular extension and chest wall invasion. The presence of a pathological fracture is a poor prognostic feature, which is associated with a higher rate of amputation. Other prognostic factors for a poor outcome include older age at presentation, axial location, and involvement of the resection margin.⁴⁻⁸

DDCS is notorious for poor survival. In a case series by Grimer et al,² the overall survival rate was 59% at one year, 38% at two years, and 24% at five years. Median survival for all patients was 1.4 years. In patients with metastasis at diagnosis, the median survival rate dropped to five months, the one-year survival rate was 26% and the two-year survival rate was 10%. Despite adequate local disease control, early distant metastasis is frequently seen, with the lungs being the commonest site. The dismal prognosis is believed to be related to the high-grade mesenchymal tumour component. The effect of chemotherapy and radiotherapy on survival of DDCS is not well-documented.²

CONCLUSION

DDCS is a highly malignant disease with grave prognosis. It is valuable for radiologists to identify the cardinal imaging features of tumour bimorphism, which suggest the diagnosis and aid the site to biopsy. Early and accurate diagnosis with radical surgery is important in the hope of securing better survival in these patients.

REFERENCES

1. Littrell LA, Wenger DE, Wold LE, Bertoni F, Unni KK, White LM, et al. Radiographic, CT, and MR imaging features of dedifferentiated chondrosarcomas: a retrospective review of 174 de novo cases. *Radiographics*. 2004;24:1397-409.
2. Grimer RJ, Gosheger G, Taminiau A, Biau D, Matejovsky Z, Kollender Y, et al. Dedifferentiated chondrosarcoma: prognostic factors and outcome from a European group. *Eur J Cancer*. 2007;43:2060-5.
3. Dahlin DC, Beabout JW. Dedifferentiation of low-grade chondrosarcomas. *Cancer* 1971;28:461-6.
4. Sopta J, Dorđević A, Tulić G, Mijucić V. Dedifferentiated chondrosarcoma: our clinico-pathological experience and dilemmas in 25 cases. *J Cancer Res Clin Oncol*. 2008;134:147-52.
5. Staals EL, Bacchini P, Bertoni F. Dedifferentiated central chondrosarcoma. *Cancer*. 2006;106:2682-91.
6. Bruns J, Fiedler W, Werner M, Delling G. Dedifferentiated chondrosarcoma — a fatal disease. *J Cancer Res Clin Oncol*. 2005;131:333-9.
7. Dickey ID, Rose PS, Fuchs B, Wold LE, Okuno SH, Sim FH, et al. Dedifferentiated chondrosarcoma: the role of chemotherapy with updated outcomes. *J Bone Joint Surg Am*. 2004;86:2412-8.
8. Staals EL, Bacchini P, Mercuri M, Bertoni F. Dedifferentiated chondrosarcomas arising in preexisting osteochondromas. *J Bone Joint Surg Am*. 2007;89:987-93.
9. Ogose A, Sim FH, O'Connor MI, Unni KK. Bone tumors of the coracoid process of the scapula. *Clin Orthop Relat Res*. 1999;358:205-14.
10. Pant R, Yasko AW, Lewis VO, Raymond K, Lin PP. Chondrosarcoma of the scapula: long-term oncologic outcome. *Cancer*. 2005;104:149-58.