

The expression of selected immunocytochemical diagnostic markers in the case of chondrosarcoma with a mesenchymal component

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The case of 52-year-old man is presented, who had suffered from pains in his right brachial region and in whom, upon admission to the Lower Silesia Centre of Oncology, a tumour of 20 cm in diameter and restricted mobility was disclosed in the right brachial region and proximal 1/3 of his right arm. Radiograms of his right humerus disclosed non-uniform restructuring of the osseous tissue, dominated by osteosclerosis in the upper half of his right humerus, while in the surrounding soft tissues of the proximal portion non-uniform shades were seen of calcified appearance. The patient was qualified to surgical biopsy and histopathological examination of the sample disclosed Chondrosarcoma G II. Following amputation of the right upper extremity together with the scapula, the tumour was subjected to histopathological and immunocytochemical examination in the Department of Pathomorphology, Lower Silesia Centre of Oncology. In numerous samples of the tumour dedifferentiated chondrosarcoma was diagnosed with a dominating component of malignant fibrous histiocytoma (MFH), which was confirmed by detecting a high expression of alpha-1-antichymotrypsin (ACT) within the spindle-shaped cell component of the chondrosarcoma. Spindle-shaped elements in dedifferentiated chondrosarcoma (DChSa) may represent patterns of fibrosarcoma, osteosarcoma or the malignant fibrohistiocytoma (MFH) type and they are present as if in the form of restricted fields with no reciprocal infiltrates. Moreover, due to the worse prognosis in the case of the MFH component in dedifferentiated chondrosarcoma as compared to that in classical chondrosarcoma, common evaluation of the material was made by a surgeon, radiologist and histopathologist, accompanied by a minimum panel of immunocytochemical tests (ACT, Vimentyna, Desmina, S-100). This enabled a final diagnosis for all tumour components to be established and prognosis to be made regarding the further fate of the patient.

key words: dedifferentiated chondrosarcoma, dedifferentiated sarcomas

INTRODUCTION

Chondrosarcoma is the second most frequently developing malignant tumour of the bones (or third, if multiple myeloma is also included), accounting for around 10% of malignant bone tumours [2, 4, 9]. It appears mainly in the 4th to 6th decade of life, although around 10% patients are below 20 years of age [2, 3, 9]. It affects males with a slightly higher frequency than females. In more than 70% of cases it develops in the bone of pelvis, the proximal ends of the humerus and femur and in the bones of the trunk [2, 9]. In a proportion of cases, the chondrosarcoma component may form one of element of complex tumours, consisting of, for example, osteochondroma. The tumour has seldom been noted in the small bones of the hands and feet, in the mandible, maxilla, nasal septum or laryngeal cartilages or in the soft tissues [2, 5, 9]. Chondrosarcoma of the bones can be subdivided into certain subtypes depending on clinical, radiological and histological criteria [4, 8, 9].

CASE REPORT

Patient J.R., 52 years of age felt pain in his right shoulder from the beginning of February 2002. On 12th November, 2002 he reported to the Oncological Outpatient Clinic, Lower Silesia Centre of Oncology (DCO), for a surgical biopsy. Upon admission his general condition was extremely good. Examination of his right shoulder and proximal 1/3 of his right arm demonstrated a tumour of poorly defined limits, restricted mobility and 20 cm in diameter. In the right armpit a mobile lymph node, 1 cm in diameter, was disclosed. Radiograms of his right humerus demonstrated a non-uniform restructuring of the osseous tissue with prevalence of osteosclerosis in the upper half of the right humerus. The soft tissues of the proximal portion of the right humerus contained heterogeneous shades of calciferous appearance. On 14th November, 2002 an incision, made in general anaesthesia along the long axis of the right arm exposed a tumour, from which a biopsy was taken for histopathological examination and the patient was released home. Histopathological examination no. 21084-85/02 demonstrated chondrosarcoma GII. The patient was re-admitted to DCO and on 12th December 2002 and was operated on in general anaesthesia. The right upper extremity was amputated, together with the scapula and clavicle, from incisions parallel to the right clavicle from the front and along the long axis of the scapula from the rear, following infiltration of the brachial plexus

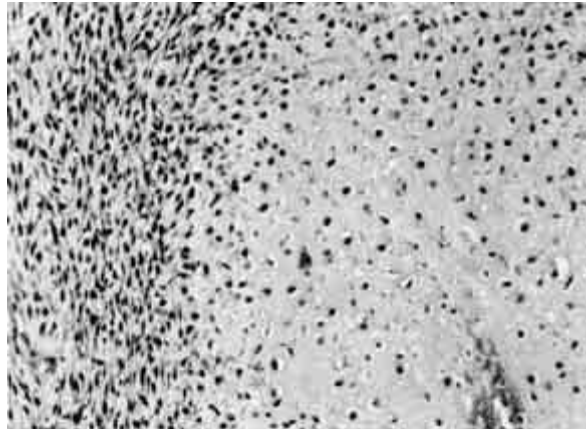


Figure 1. Chondrosarcoma with mesenchymal component. HE; magnification $\times 100$.

with 2% xylocain, ligation and transection of its elements. The general condition of the patient following surgery was good. Histopathological examination of the tumour conducted in the Department of Pathomorphology DCO (no. 23417-19/02) using H+E (Fig. 1) and Van Gieson staining and immunocytochemical tests (ACT, anti-alpha-1-antichymotrypsin, vimentin, desmin, S-100, DAKO) resulted in a diagnosis of dedifferentiated chondrosarcoma with a dominant component of malignant fibrous histiocytoma. Immunocytochemical tests: ACT (+++) (Fig. 2), vimentin (++), desmin (+), S-100 (+).

DISCUSSION

In 1971, Dahlin and Beabout [1] described for the first time a dedifferentiated low grade chondrosarcoma, but recently some authors have questioned the term, arguing that, due to the histological and morphological structure of the tumour, it should be termed a chondrosarcoma with a supplementary

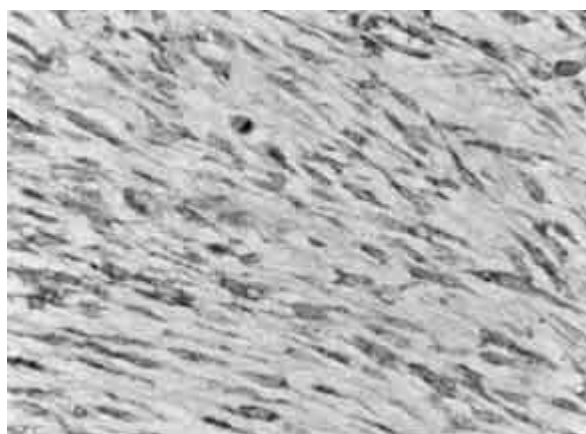


Figure 2. Expression of ACT in mesenchymal component. Immunohistochemical method; magnification $\times 100$.

mesenchymal component. As a rule, the tumour appears in patients older by around 10 years than the most common form of chondrosarcoma, which develops in 4th-6th decade of life/ [2, 3, 9]. It is localised in the same sites and exhibits a similar multi-annual course, with sudden acceleration of tumour growth at the end of the patient's life. In approximately 1/3 of cases the radiological pattern of the tumour resembles that of a conventional chondrosarcoma. The tumour is, however, complex: the chondrosarcoma component of G1 or GII differentiation grade is accompanied by spindle cells of the high-grade spindle cell sarcoma type, most frequently presenting patterns of fibrosarcoma, osteosarcoma or malignant fibrohistiocytoma [5, 7, 9]. Extremely rarely, they may manifest traits of differentiation to rhabdomyoblasts [2, 6, 9]. Characteristic for the tumour structure is a relatively sharp delimitation of the chondrous structure elements, which are well differentiated from the spindle shaped cells of sarcomatous structure. An impression is gained from its appearance of structures positioned side by side, instead of the infiltrations seen in chondroblastic osteosarcoma [5, 8, 9]. This delimitation of the two structures is significant for prognosis. In cases of dedifferentiated chondrosarcoma a 5-year survival amounts to 10% and is significantly worse than in cases of chondroblastic osteosarcoma [9]. Metastases to the lungs develop within 14 months of surgery [5]. Since the radiological as well as clinical patterns are not unequivocal, open surgical biopsy remains the diagnostic procedure of choice [8]. In order to isolate representative material for the studies, preliminary evaluation of the material is indicated, accompanied by analysis of radiological patterns because of the

duality of the tumour structure. In our case a single component only was obtained during biopsy. The final diagnosis, linked to a definitely unfavourable prognosis, was established in preparations obtained from the post-surgery material, isolated from many fields.

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