CASE REPORT
UDC: 616.988.6:616.717.2:616-089.8

Undifferentiated chondrosarcoma of extraskeletal supraclavicular localization

Aleksandar Milovančev, Dejan Ilinčić, Milorad Bijelović, Miloš Koledin, Dejan Đurić, Dejan Vučković

Institute for Pulmonary Diseases, Sremska Kamenica, Serbia & Montenegro; Address correspondence to: Prof. Dr. Aleksandar Milovančev, Institute for Pulmonary Diseases, Institutski put 4, 21204 Sremska Kamenica, Serbia & Montenegro, The manuscript was received: 12.12.2003, Provisionally accepted: 04.06.2004, Accepted for publication: 04.10.2004

ABSTRACT

We reported a rare extraskeletal localization of undifferentiated chondroscarcoma in a 71-year old patient. The neoplasm with a 20-year history in the left supraclavicular region exhibited a rapid growth in only a few months. Six months after a total tumor excision the patient has been in an excellent general health condition, with no signs of either a local relapse or distant metastases.

KEY WORDS: *Chondrosarcoma; Soft Tissue Neoplasms; Clavicle; Surgery; Treatment Outcome*

INTRODUCTION

Chondrosarcoma makes about 20% of all primary malignant bone tumors (1). It is the second common bone tumor, reaching the highest incidence in the 40-60 yr age group. It develops *de novo*, or (10% of the cases) as a sarcomatous transformation in Ollier or Paget's bone disease (2). The Epidemiology Department of the Institute of Oncology in Sremska Kamenica reports 24 chondrosarcoma cases registered in the region of Vojvodina from 1991 to 2000, with the male : female ration of 1.4 : 1. Extraskeletal chondrosarcomas have also been reported. They are believed to originate from a primitive mesenchymal cell (3). Undifferentiated chondrosarcoma is a neoplasm composed of two coexisting components: 1) a high malignancy grade non-chondroid sarcoma and 2) a low malignancy grade cartilaginous tumor. Undifferentiated chondrosarcoma makes about 11% of all chondrosarcomas and ranges among the most aggressive malignant tumors. It affects older-aged patients, usually older than 50 years (4).

CASE REPORT

A 71-year old male patient was admitted to the Chest Surgery Department of the Institute for Pulmonary Diseases in Sremska Kamenica due to the tumor in the left supraclavicular region. The tumor had had a 20-year long history but it exhibited an exceptionally rapid growth a few months prior to admission. Before it started its intensive growth, the tumor was soft, a few centimeters in diameter, producing no troubles to the patient.

On admission, the patient was in a good general condition, lacking any subjective symptoms. The clinical examination confirmed the tumorous mass in the left supraclavicular region at the neck base. The tumor was irregularly oval in shape, over 10 cm in diameter (Figure 1), of the mixed firm-soft consistency, migrating in regard to the adjacent neck structures. The skin covering the tumor was thinned, with no signs of infiltration or inflammation. The chest X-ray finding was normal.

The diagnosis of sarcoma was established by fine needle biopsy tumor. Having received an adequate preoperative treatment, the patient was submitted to the surgical extirpation of the tumor in total anesthesia (Figure 2).

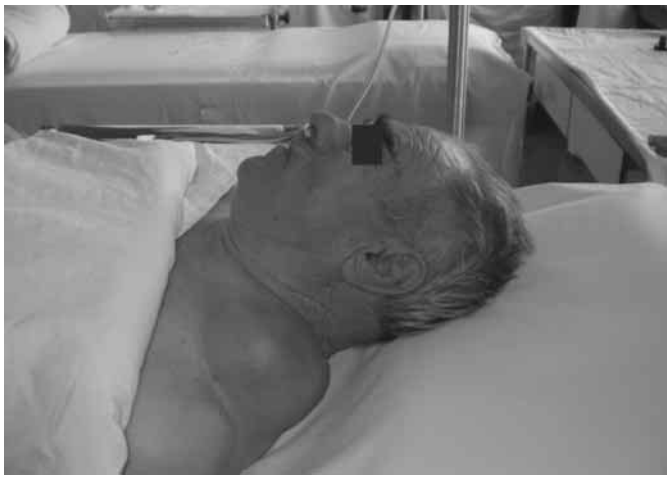

Figure 1. The tumor presentation in the left supraclavicular region

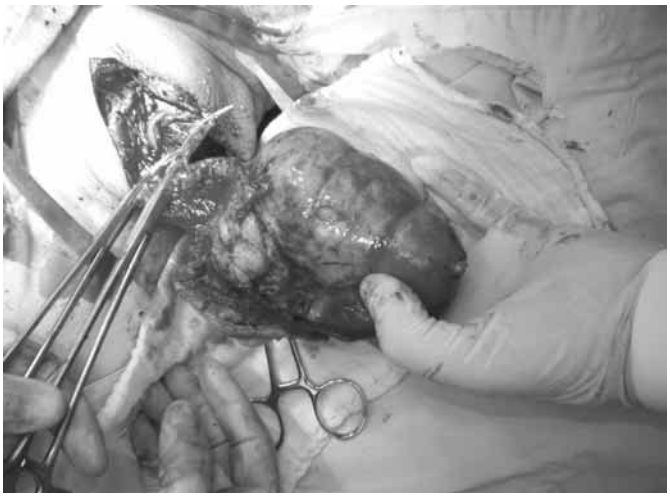

Figure 2. Intraoperative appearance of the tumor

The frozen-section histological analysis established the mesenchymal origin of the tumor, favoring its malignant etiology in the differential diagnosis. A local dissection of the neck was performed. The fat tissue with adjacent tumor-free lymph nodes, as confirmed on the frozen section analysis, was excised.

The definite histological diagnosis was chondrosarcoma undifferentiated in type, established on an ellipsoid skin sample 12x12 cm in size, containing a clearly defined, 10x10x8 cm sized node at the distance of 0.7 cm beneath the skin surface. The inside of the node, to the skin surface, had a firm, crescent-shaped tissue formation in the size of 7x5x3 cm, whitish in color. The remaining tissue had the tissue features. Four histological sections of the material were stained by the HE staining method. Atypical cells, mostly spindle and occasionally round or polygonal in shape, were seen in the surroundings of the whitish formation. These cells were moderately large, with the cytoplasm ranging from sparse to moderately abundant. The spindle-shaped cells produced entangled bundles, which took diverse directions. The space between the tumor cells was involved by numerous blood vessels. A great number of tumor cells were very large, with one or more rather big hyperchromatic nucleoli. Cellular cartilaginous islands with atypical hyperchromatic cells were sporadically scattered between the tumor cells (Figure 3). Tumor was partly necrotic and partly composed of mature fat tissue. The postoperative course and wound healing were normal. Regarding the established tumor type, the consulting oncologist abandoned both the irradiation and chemotherapy treatment. On the control examination six months later, the patient was in an excellent general health condition, free of symptoms (Figure 4).

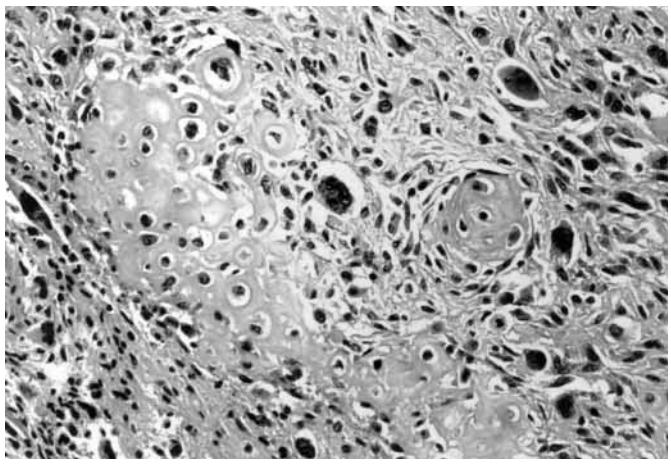

Figure 3. Dedifferentiated chondrosarcoma HE x 250

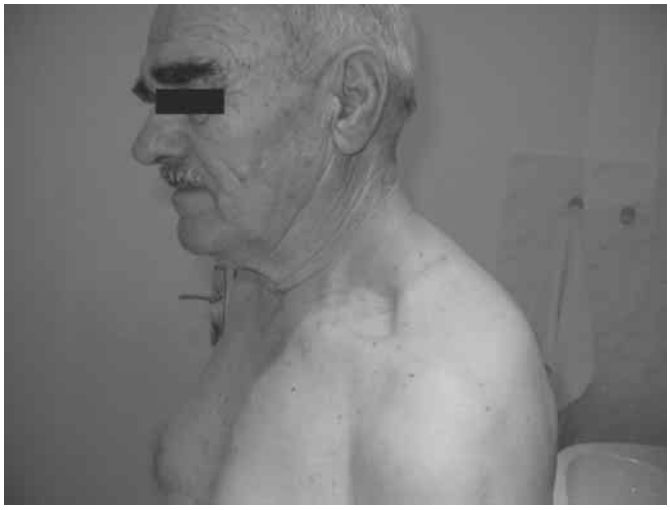

Figure 4. Six months later - no signs of the local relapse

The radiological finding showed signs of the surgery-induced left phrenic nerve paresis, but no local relapse or distant metastases.

DISCUSSION

Chondrosarcoma is resistant to both irradiation and chemotherapy (5), so a radical large-scale surgical resection is the therapy of choice regardless the tumor localization. However, the surgical treatment is usually ineffective due to a local relapse of the tumor, occurring in 50% of the patients (6). Although the benefits from chemotherapy have not been confirmed yet, it is sometimes applied for high malignancy tumors with distant metastases. Chondrosarcoma makes 5%-10% of all neck and head tumors (7). Due to the low incidence rate of regional lymph node metastases (8%-10%), the neck dissection is not necessary, except when lymphadenomegally of the neck lymph nodes is present (8).

The prognosis depends on the tumor localization and its malignancy grade. The overall 5- and 10-year survival rates are 80% and 70% respectively. Malignancy grade III and undifferentiated tumors are exceptionally aggressive, with a higher incidence of local relapses and distant metastases. Grade III tumors have the 5-year survival rate of 50% (9). The extraskeletal localization features mesenchymal chondrosarcoma which was for the first time described as I unique pathological entity in 1956 by Lichtenstein and Bernstein (10). These tumors make less than 2% of all chondrosarcoma cases (11), having the highest incidence in the 20-30 yr age group and the 5-year survival of about 50% (12,13). The extraskeletal localization of undifferentiated chondrosarcoma is very rare. Undifferentiated chondrosarcoma has a very low survival rate. Since metastases can develop long after the initial surgery, these tumors should be followed-up much longer than five years (14,15). Most of the patients die within the first two postoperative years, due to a rapid metastatic dissemination of the tumor. The 5-year survival rate with this tumor type is less than 10%.

REFERENCES

1. Fechner RE, Mills SE. Cartilagenous Lesions. In: Atlas of Tumor Pathology, Tumors of Bones and Joints. Third Series. Washington DC.: AFIP; 1993. p. 101-15.
2. Dahlin DC. Bone tumors: General aspects and data on 6,221 cases, 3rd ed. Springfield: Charles C Thomas; 1978.
3. Forest M. Chondrosarcoma: variants. In: Forest M, Tomeno B, Vanel D, editors. Orthopaedic Surgical Pathology. Diagnosis of tumors and pseudotumoral lesions of bone and joints. New York: Churchill Livingstone; 1998. p 261-92.
4. McFarland GB, McKinley LM, Reed RS. Dedifferentiation of Low-Grade Chondrosarcoma. Clin Orthopaedics 1977;122:157.
5. Sarcoma Meta-Analysis Collaboration: Adjuvant chemotherapy for localized resectable soft-tissue sarcoma of adults. Meta-analysis of individual data. Lancet 1997;350:1647-54.
6. Wanebo HJ, Benjamin RS, Hadju SI, Healey GB, Lindberg, RD, Weber R. Sarcomas of the head and neck. In: Johnson J, Didolker M, editors. Head and Neck Cancer, Vol. III. Excerpta Medica - Int Cong Prem; 1993.
7. Patel SG, Shaha AR, Shah JP. Soft tissue sarcomas of the head and neck: an update. Am J Otolaryngol 2001;22(1): 2-18.
8. Soft Tissue Sarcoma, Brennan et al. In: Devita V, Hellman S, Rosenberg SA, editors. Cancer Principles and Practice of Oncology. Philadelphia: J. Lippincott; 1997. p. 1738-88.
9. Evans HL, Ayala AG, Romsdahl MM. Prognostic factors in chondrosarcoma of bone: a clinicopathologic analysis with emphasis on histologic grading. Cancer 1977;40(2):818-31.
10. Lichtenstein 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 multricentric ones and chondromyxoid fibromas. Cancer 1959;12:1142-57.
11. Nakashima Y, Unni KK, Shives TC, Swee RG, Dahlin DC. Mesenchymal chondrosarcoma of bone and soft tissue. A review of 111 cases. Cancer 1986;57:2444-53.

12. Dorfman HD, Czerniak B. Malignant cartilage tumors. In: Bone Tumors. St. Louis: Mosby; 1998. p. 353-440.

13. Weiss SW, Goldblum JR. Cartilaginous soft tissue tumors. In: Enzinger and Weiss's Soft Tissue Tumors, 4th edt. St. Louis: Mosby; 2001. p. 1361-88.

14. McCarthy EF, Dorfman HD. Chondrosarcoma of bone with dedifferentiation: a study of eighteen cases. Hum Pathol 1982;13:36-40.

15. Marco RA, Gitelis S, Brebach GT, Healey JH. Cartilage tumors: evaluation and treatment. J Am Acad Orthop Surg 2000;8(5):292-304.

16. Anract P, Tomeno B, Forest M. Dedifferentiated chondrosarcoma. A study of 13 clinical cases and review of the literature. Rev Chir Orthop Reparatrice Appar Mot 1994;80(8):669-80.