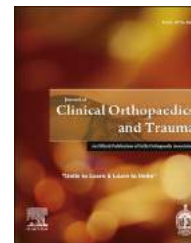


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Case Report

Spindle cell sarcoma of bone arising from a non-ossifying fibroma: A case report

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1. Introduction

The majority of malignant bone lesions, and spindle cell sarcoma of bone is no exception, arise spontaneously. Less frequently, they may be secondary to other conditions like radiation therapy, bone pathologies like Paget's disease, bone infarct, aseptic necrosis and many benign tumors or tumor like bone conditions like metaphyseal fibrous defect (MFD), giant cell tumor (GCT), osteochondroma, chondroma and others.¹

This work describes a very rare case of sarcoma of bone arisen in the same area of a benign lesion, diagnosed occasionally ten years ago, which has not undergone previously medical, radiotherapeutic or surgical treatment.

2. Case history

We report a case of a 45-year old man with a pathologic fracture of the distal left femur because of a spindle cell sarcoma of bone arising in a pre-existent pseudotumoral lesion diagnosed like non-ossifying fibroma (NOF) ten years before. We show that a sarcoma of bone can arise in the same area of a tumor-like lesion.

In 1999 because of a contusion of the knee the patient underwent x-rays and computed tomography scan (CT). The antero-posterior view of X-ray (Fig. 1) showed a round osteolytic lesion in the lateral metaphysis of the distal femur. CT scan (Fig. 2) confirmed an osteolytic lesion surrounded by sclerotic margins, measuring 2–3 cm. The diagnosis was NOF.

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Fig. 1 – Antero-posterior (AP) radiograph of the knee shows a round osteolytic lesion of the distal femur with sclerotic margins (arrow).

In October 2008 the patient began to have pain in the distal part of the left thigh without any trauma. At the beginning the pain was moderated; then it became continuous.

On April 6 2009 he had a pathological fracture of the distal left femur (Fig. 3a) and the day after he underwent surgical treatment with curettage of the lesion, cement and osteosynthesis with plate and screws (Fig. 3b) without any preoperative histological exam.

The final diagnosis was high-grade sarcoma of bone. Therefore he was admitted at our Institute in July 2009 because the surgical treatment was inappropriate (intralesional). CT of lungs and bone scintigraphy showed no evidence of metastatic lesions. Histological slides were reviewed from our pathologist who confirmed the diagnosis of spindle cell sarcoma of bone grade 4.

After 3 cycles of preoperative chemotherapy according with EUROBOSS protocol (cisplatin, adriamycin and ifosfamide), a resection of the distal femur and reconstruction with

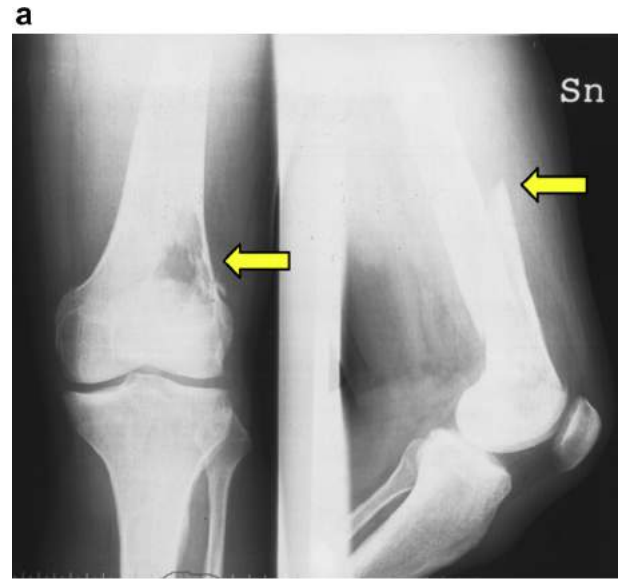


Fig. 3 – a: Antero-posterior (AP) and latero-lateral (LL) radiographs of the knee show a pathological fracture of distal femur (arrows). b: AP and LL radiographs show the curettage and cement of the sarcoma and osteosynthesis with plate and screws.

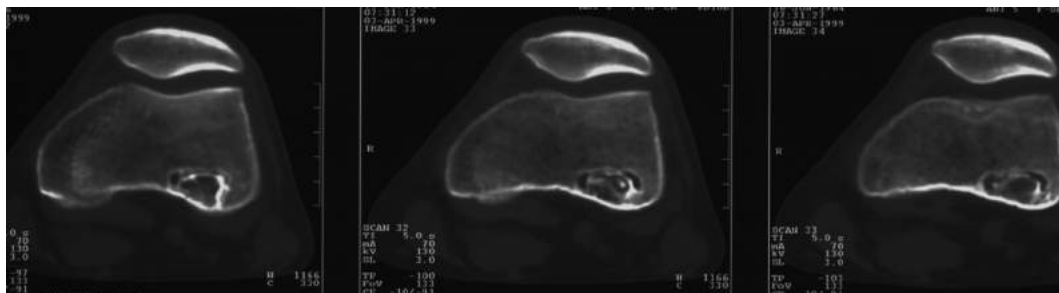


Fig. 2 – CT-scan of the knee shows a round osteolytic lesion of the distal femur with sclerotic margins and no periosteal reaction or cortical destruction.



Fig. 4 – AP radiograph shows reconstruction of the distal femur with modular prosthesis.

modular prosthesis (GMRS) was performed (Fig. 4). Then he underwent postoperative chemotherapy with the same protocol.

The analysis of the specimens confirmed the diagnosis of spindle cell sarcoma of bone grade 4 (Fig. 5 a–c).

During the follow-up the patient showed pulmonary nodules and he underwent metastasectomy on September 2010. At last follow-up on September 2012 the patient was alive with no evidence of disease.

3. Discussion

The first descriptions of malignant transformation of a pseudotumoral lesion were attributed to Hastrup² and Bhagwandeem,³ who reported a case of osteogenic sarcoma of bone arisen in a NOF, previously treated with radiation therapy and curettage and packing with bone allograft respectively.

Then, Braddock et al⁴ and Rockwell et al⁵ described respectively a case of osteosarcoma developing in a patient with multiple enchondromatosis and in a solitary enchondroma.

Later, several authors^{6–8} reported coexistence between sarcoma and MFDs, like fibrous cortical defect (FCD) and NOF, which occur in 35–40% of growing children studied with x-rays. These studies don't have evidence that a sarcoma can arise from a MFD: both FCD and osteosarcoma occur in the same age (second decade of life) and in the same site (distal femur and proximal tibia). Therefore, it's to be expected that

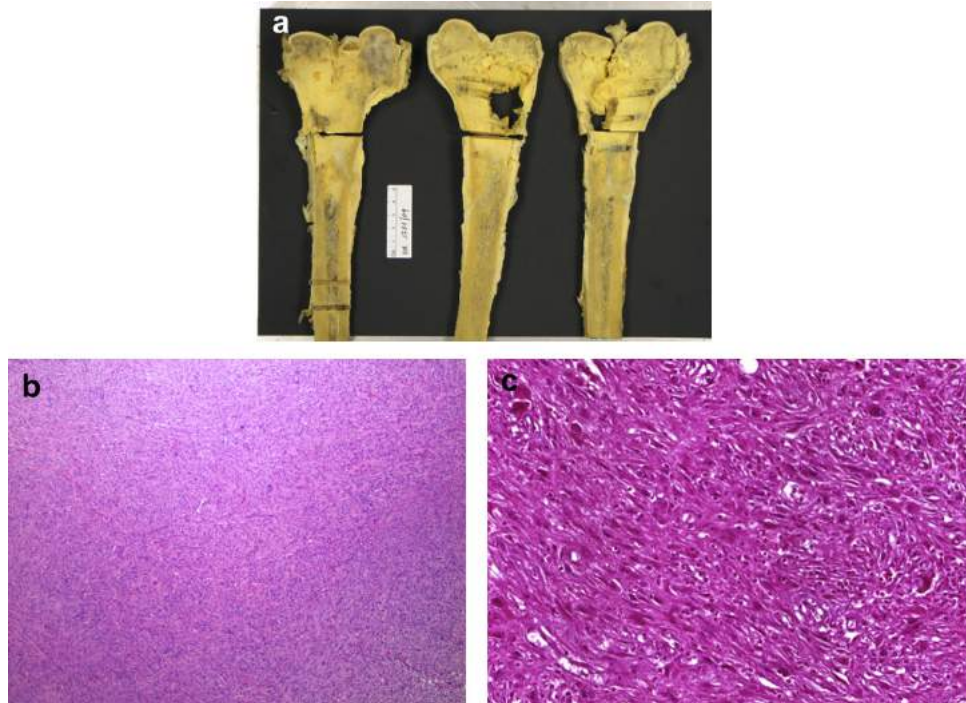


Fig. 5 – a: On gross section of the entire specimen a greyish lesion measuring about 9 cm in the longitudinal axis is present in the metaphysis and epiphysis around the areas of the previous curettage. The lesion also infiltrates the soft tissue around the bone. b, c: Low-power (b) and high-power (c). Proliferation of malignant spindle cells in a storiform pattern diffusely infiltrating the bony trabecule. A histological diagnosis of spindle cells sarcoma grade 4 according to Broder's classification was made. Tumoral necrosis after neo-adjuvant chemotherapy was 57% (grade 2 according to Huvos classification) (Hematoxylin–Eosin, ×).

the simultaneous occurrence of fibroma and either GCT or osteosarcoma in the distal femur could happen.

According to Kryakos et al,⁷ the finding of a malignant bone tumor in intimate association with a MFD is a chance occurrence but to demonstrate a clear relationship between them, some criteria have to be satisfied: the original lesion must have the radiologic features of a MFD which, over time, change to an aggressive appearing pattern with its epicenter corresponding to that of the previous benign lesion. Moreover, no radiation therapy must have been given. So they reviewed and rejected the reports of a malignant transformation of a MFD because they lacked convincing radiological or histopathological evidence of a pre-existent benign fibrous lesion.

Recently Picci et al⁹ reported a series of 12 secondary bone sarcomas arising in benign bone lesions as GCT, aneurysmal bone cyst (ABC), NOF and simple bone cyst treated with curettage and grafting. In order to the long latency of development of sarcoma, the authors speculated an origin “de novo” of the neoplasia, may be because of the use of mesenchymal stem cells in their treatment. Conversely, Takazawa¹⁰ and Abdelwahab¹¹ described a case report of osteosarcoma arisen in the same site of a desmoplastic fibroma, respectively 16 and 11 years later a curettage and bone grafting: they thought that malignant transformation of benign lesion is extremely rare but possible. In these cases is very difficult to understand if it was a sarcoma from the onset because the differential diagnosis between a low-grade fibrosarcoma and a desmoplastic fibroma is a challenge even for experienced pathologists.

According to Huvos et al,¹ the commonest secondary osseous malignant fibrous histiocytoma is due to radiation therapy (15.4%). The latent period between irradiation and the appearance of the bone sarcoma ranged from 4 to 47 years with a mean of 16.5 years. The other forms of secondary malignant bone lesions are due to benign bone pathologies or tumor like bone conditions: while it's known the relationship between Paget's disease and bone sarcoma, is not the same for benign bone conditions.

However, Marui et al¹² described 2 cases of malignant transformation of GCT 10 years after surgery and without radiation therapy. In these cases, histological examination showed the coexistence of zones of high-grade sarcoma and GCT: according to them, these are examples of malignant transformation of benign lesions.

Grote et al¹³ reported a case of spontaneous malignant transformation of conventional GCT of the iliac crest in a 35-year old woman into a high grade osteosarcoma 10 years after the first appearance on x-rays. The tumor remained untreated until sarcomatous transformation occurred because of the patient refused therapy for personal reasons. This is another rare case of secondary malignant GCT developing spontaneously without any previous irradiation: in literature only 7 cases like this are found. It's unknown whether the tumor was not malignant from the onset without being recognized as such or whether it contained malignant areas that might have been overlooked.

Concerning the relationship between NOF and secondary sarcoma of bone, two possibilities can be considered: the first is that NOF occurs incidentally in the same area and isn't

related to the occurrence of sarcoma; the second is that cells of fibroma underwent malignant transformation in the surrounding osteoprogenitor cells. Even the first one seems to be the more likely, we can not exclude the possibility of a malignant transformation of a NOF.⁸

Despite a part of Kryakos' criteria⁷ is not satisfied (absence of histological documentation of the previous lesion), this case is a clear malignant transformation of a pseudotumoral lesion, that is visible on X-rays and CT (Fig. 6a and b). The sarcoma arose in the same site of a NOF in a middle-aged man (is not the typical age of onset of osteosarcoma). As a result the sarcoma completely replaced the previous lesion that is not present on the specimen. We believe that this is the first clinical and radiographic documented case of malignant transformation of a NOF which has not undergone radiation therapy or surgical treatment.

In conclusion the goal of this study is to emphasize that this eventuality is extremely rare but possible and these patients should be followed over time.

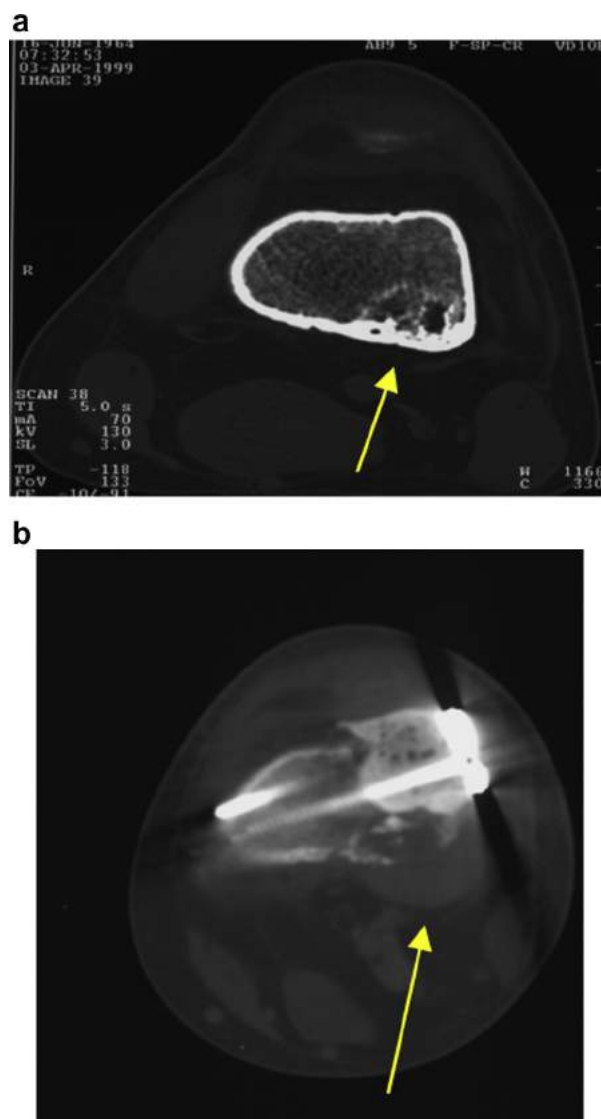


Fig. 6 – a,b: The arrows show that the NOF and the sarcoma are in the same site.

Conflicts of interest

No benefits in any form have been received or will be received from a commercial party related directly or indirectly to the subject of this article.

REFERENCES

1. Huvos AG, Woodard HQ, Heilweil M. Postradiation malignant fibrous histiocytoma of bone. *Am J Surg Pathol.* 1986;10(1):9–18.
2. Hastrup J, Jensen TS. Osteogenic sarcoma arising in a non-osteogenic fibroma of bone. *Acta Pathol Microbiol Scand.* 1965;63:493–499.
3. Bhagwande SB. Malignant transformation of a non-osteogenic fibroma of bone. *J Pathol Bacteriol.* 1966;92:562–564.
4. Braddock GTF, Hadlow VD. Osteosarcoma in enchondromatosis (Ollier's disease). Report of a case. *J Bone Joint Surg.* 1966;48-B:145–149.
5. Rockwell MA, Enneking WF. Osteosarcoma developing in solitary enchondroma of the tibia. *J Bone Joint Surg.* 1971;53(2):341–344.
6. Abdelwahab IF, Klein MJ, Kenan S, et al. Coexistence of primary bone tumors: report of 4 cases of collision tumors. *Can Assoc Radiol J.* 2002;53(5):296–302.
7. Kryakos M, Murphy WA. Concurrence of metaphyseal fibrous defect and osteosarcoma. *Skeletal Radiol.* 1981;6:179–186.
8. Suginoshita T, Kusuzaki K, Nagaoka T, et al. Case report: natural development of osteosarcoma from precancerous lesion. *Anticancer Res.* 2000;20:511–514.
9. Picci P, Sieberova G, Alberghini M, et al. Late sarcoma development after curettage and bone grafting of benign bone tumors. *Eur J Radiol.* 2011;77:19–25.
10. Takazawa K, Tsuchiya H, Yamamoto N, et al. Osteosarcoma arising from desmoplastic fibroma treated 16 years earlier: a case report. *J Orthop Sci.* 2003;8(6):864–868.
11. Abdelwahab IF, Klein MJ, Hermann G, Steiner GC, Yang DC. Osteosarcoma arising in a desmoplastic fibroma of the proximal tibia. *Am J Roentgenol.* 2002; 178(3):613–615.
12. Marui T, Yamamoto T, Yoshihara H, Kurosaka M, Mizuno K, Akamatsu T. De novo malignant transformation of giant cell tumor of bone. *Skeletal Radiol.* 2001;30:104–108.
13. Grote HJ, Braun M, Kalinski N, et al. Spontaneous malignant transformation of conventional giant cell tumor. *Skeletal Radiol.* 2004;33:169–175.