

Case report

Giant Osteosarcoma of Femur: a Well Documented Case with Management Difficulties in Yaounde (Cameroon).

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ABSTRACT

Osteosarcomas are primary malignant bone tumours in which mesenchymal cells produce an osteoid matrix. It is generally the most common malignant bone neoplasm. The purpose of this article was to present the difficulties bound to the management of an osteosarcoma of the thighbone in Yaounde. A 49-year-old woman referred to the University Hospital Center of Yaounde for further management of the complete and large deformation of the right thigh. A biopsy realized 14 years rather concluded in an osteoma. The patient had refused an amputation proposed 5 years after this first diagnosis, in spite of the loss of shape of the limb. The main clinical findings were hard swelling and large deformation of the right thigh. Review of the X-ray revealed multiples larges and lytic expansible lesions of third lower femur. A microscopic analysis of the specimens stemming from the part of amputation showed areas of osteoid formation surrounded by hyper cellular stroma, confirming the diagnosis of osteoblastic osteosarcoma. The patient was then recommended for oncologic treatment. It thus seems clearly that in the absence of multidisciplinary confrontation, the management is not only delayed and extended but burdens considerably the prognosis of patients.

Key words: Bone, Osteoma, Osteosarcoma, Management, Prognosis.

RESUME

Les ostéosarcomes sont des tumeurs malignes osseuses primitives caractérisées par la production d'une matrice ostéoïde par des cellules mésenchymateuses. Ils représentent les tumeurs osseuses les plus fréquentes. Le but de cet article était de présenter les difficultés liées à la prise en charge d'un ostéosarcome du fémur à Yaoundé. Il s'agit d'une patiente de 49 ans référée au Centre Hospitalier et Universitaire de Yaoundé pour la prise en charge d'une tumeur déformatrice de la cuisse droite. Une biopsie réalisée 14 ans plus tôt concluait à un ostéome. La patiente avait refusé une amputation proposée 5 ans après ce premier diagnostic, au vu de l'évolution déformatrice du membre. Le nouvel examen clinique mettait en évidence une tuméfaction indurée et déformante de la cuisse droite. Les images radiographiques montraient de larges multiples lésions osseuses lytiques du tiers inférieur du fémur. L'analyse histopathologique des spécimens issus de la pièce d'amputation montrait une tumeur ostéofomatrice à stroma mésenchymateux hyper cellulaire, confirmant le diagnostic d'ostéosarcome ostéoblastique. La patiente bénéficie actuellement d'une chimiothérapie adjuvante. En l'absence de confrontation pluridisciplinaire, la prise en charge médicale est non seulement retardée et prolongée mais grève considérablement le pronostic vital des patients.

Mots clés: Os, Ostéome, Ostéosarcome, Management, Pronostic.

INTRODUCTION

Sarcomas are neoplasms arising from connective tissue elements of the body. Approximately 80% arise in soft tissue, while the remainder originate in bone. Bone sarcomas are characterized by their location in bone and sometimes produce osteoid or immature bone [1]. The exact time frame for the malignant transformation of benign lesion of bone is not known [2]. This presentation aimed at showing the slow evolution of an initially benign bone tumour into a huge malignant osteosarcoma.

HISTORY

A 49 years old woman presented with a history of swelling and discomfort in the right thigh since 1996. She was evaluated elsewhere for his complaints and a biopsy was performed and concluded as an osteoma of bone. The patient was subsequently referred to the University Hospital Center for further management. On examination, the complete and large deformation

of the right thigh was seen (Figure 1 - A). Review of the X-ray revealed multiples larges and lytic expansible lesions of third lower femur. A magnetic resonance imaging (MRI) was performed that showed three larges fusiform lesions involving the right thigh with extension to the adjacent muscle planes. The lesion exhibited cloudlike areas of low signal intensity on both T1- and T2- weighted images. With this variegated clinico-radiologic picture, the patient was taken for amputation of the leg. On gross examination (Figure 1 – B and C), three lesions measuring 12.5x12 cm for the postero-external one which present a necrotic area in the center, 9x8 cm for the medial one and 8x7 cm for the posterior one was seen in the thigh. On serial sectioning, an unencapsulated grayish-white lesion was identified near an along the right femur, extending up to surrounding muscle plane. Three lymph nodes are collected near of the femoral vascular pedicle.

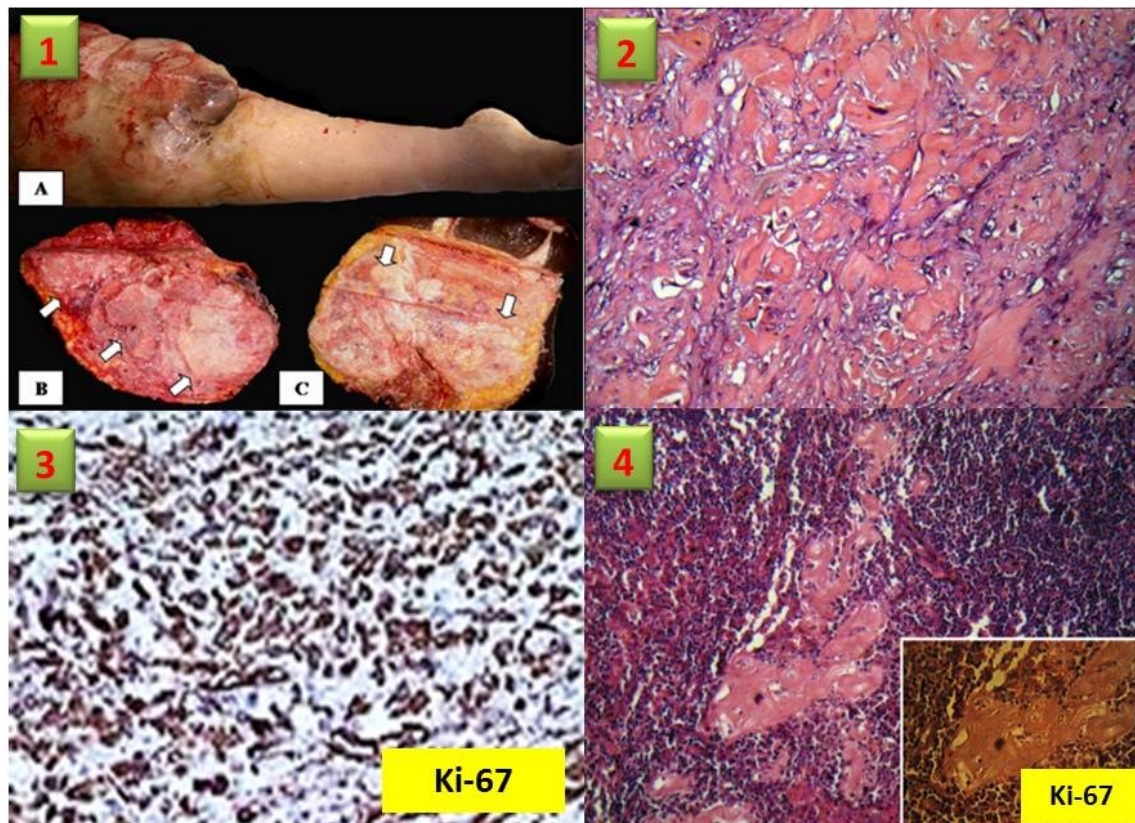


Figure 1. The gross amputation specimen. A shows large tumour nodules, B and C (arrows) invading the soft tissues. **Figure 2.** High-grade tumours showed undifferentiated mesenchymal cells with osteoid matrix deposition (H&E stain, (250)). **Figure 3.** Ki-67 expression after IHC staining. **Figure 4.** Lymph node metastasis of high-grade tumours also showed osteoid matrix deposition and Ki-67 expression (H&E (250) and IHC stain,).

A histological examination of the excised specimen showed that the neoplasm consisted of a spindle-cell proliferation that produced a focal “herringbone” pattern (Figures 2 and 3). On immunohistochemical staining, the tumour cells showed increased Ki-67 expression and were negative for CK (cytokeratin) and EMA (epithelial membrane antigen). After initial and subsequent review of multiple specimens, pathologists of our institution and an outside institution agreed that the final histopathologic diagnosis was a high-grade osteoblastic osteosarcoma with lymph nodes metastasis (Figure 4). The patient was recommended for oncologic treatment where she is currently on follow-up.

DISCUSSION

The exact time frame for the malignant transformation of benign lesion of bone is not known. The typically slow growth of benign osteoma suggests the process may occur over an extended time [3]. The most common histological patterns in transformed benign osteoma include osteosarcoma, malignant fibrous histiocytoma, malignant fibroxanthomas, and high-grade spindle cell tumours [4, 5]. The majority of reported transformations succeeded in osteosarcoma. Without rigorous

scientific approach, we could think that it concerns the presented case.

In epidemiology area, osteosarcomas are the most common bone sarcoma and develop most frequently in 10 to 20-year-olds; their most common location is the distal femur [6]. Osteosarcoma is more likely to represent a second malignancy, frequently related to Paget's disease in adults older than 65 years of age [1]. The clinical presentation of patients with bone sarcoma is highly variable [7]. Patients often present with a mass, typically one that is increasing in size as in the presented case. In general, bone sarcomas are painful but there are exceptions to this general rule. Constitutional symptoms are rare in patients with bone sarcomas, but symptoms such as fever, malaise, and weight loss can be seen, especially in patients with Ewing sarcoma [5]. Despite recent advances in cross-sectional imaging with computed tomography (CT) and magnetic resonance imaging (MRI), radiographs remain valuable for characterizing osseous lesions and enabling accurate differential diagnosis of benign and malignant tumours [8]. Multiple lytic lesions in an adult older than 40 years almost always suggest metastasis or multiple myeloma and the most common cause of a solitary destructive lesion in an adult is metastasis [9]. At the beginning of the affection, there may be some

difficulty on the radiological conclusion on the type of tumour be it benign or malignant [8]. A comprehensive medical history and physical examination are essential at the initial presentation of patients with masses and/or pain suggestive of bone sarcoma. Sarcoma simulators such as hematoma, metastatic disease, or infection can sometimes be ruled out by careful clinical examination, laboratory work-up, and appropriate imaging, but the gold standard for diagnosis is a biopsy [5]. The definition of osteosarcoma is very precise. Osteosarcoma is characterized histologically by malignant tumour cells that directly produce osteoid or immature bone [4]. Importantly it is essential to precise the type and the grade of osteosarcomas. The microscopic pathology of conventional osteosarcoma has traditionally been subdivided into three categories: osteoblastic, chondroblastic and fibroblastic [4, 10]. Histological confusion was sometimes apparent in patients having needle biopsies in which only small samples were obtained. In these cases, the diagnosis was only eventually confirmed following a new biopsy or excision-biopsy of the lesion. Metastatic osteosarcoma is found in approximately 20% of patients at the time of osteosarcoma diagnosis [1]. Osteosarcomas mainly spread hematogenously, and the lungs are the most common initial site of metastases [11]. But in our case, we observed a lymph node metastasis. The differential diagnosis of osteosarcoma includes other bone malignancies that do not form osteoid, such as malignant fibrous histiocytoma, fibrosarcoma, metastatic disease, and Ewing's sarcoma [12]. All these observations suggest many explanations of our case. Given that the initial lesion is not fully documented and the history is not relevant of primary osteoma that got transformed after some time. This really bring serious doubt about is initial benign characters. Perhaps because of the superficial biopsy or the lack of the multidisciplinary approach in the management of this case.

CONCLUSION

Bone sarcomas are neoplasm with variable clinical presentations. A high index of suspicion is required for any unexplained mass with indeterminate imaging findings. Cross-sectional MRI and CT, associated to histopathology have significantly refined the diagnosis and management of bone sarcomas. When faced with a possible sarcoma, the clinician's selection of imaging modalities and histological study has a direct impact on diagnosis, staging, and patient management.

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