



Case report

Pindborg tumor in the distal femur[☆]



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ARTICLE INFO

Article history:

Received 20 May 2016

Accepted 30 May 2016

Available online 20 February 2017

Keywords:

Femur

Bone neoplasms

Biopsy

ABSTRACT

To describe a case of possible diagnosis of Pindborg tumor on the distal femur.

A 32-years-old female patient, a native of Bolivia, resident in Brazil, arrived to this service for tumor research in the right femur. After biopsies and resection of the lesion, the case was referred to analysis and consultancy in the United States. In a report of review by the Pathology Laboratory, it was characterized the histological appearance and immunohistochemical profile were characteristic of Pindborg tumor. Currently, the patient is being followed-up at the Orthopedics and Traumatology Department of this institution, and presents a good evolution. This study presents the case of a patient with a rare tumor that was investigated for an extensive period and through many tests. Pindborg tumor was suggested as a diagnostic hypothesis due to the characteristics and behavior of the neoplastic lesion. Although this lesion is more commonly observed in odontology, the neoplasia was compatible with the diagnosis. Therefore, despite the fact that this tumor has benign characteristics, long-term monitoring is necessary, given the high rate of tumor recurrence.

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Tumor de Pindborg em fêmur distal

RESUMO

Descrever um caso de possível diagnóstico tumor de Pindborg em fêmur distal.

A paciente de 32 anos, natural da Bolívia, residente no Brasil, veio a esse serviço para investigação de massa tumoral em fêmur direito. Após feitura de biópsias e ressecção da lesão, o caso foi encaminhado para análise e consultoria nos Estados Unidos. Em laudo de revisão do Laboratório de Patologia, foi caracterizado que o aspecto histológico e o perfil

Palavras-chave:

Fêmur

Neoplasias ósseas

Biópsia

[☆] Study conducted at Universidade Católica do Rio Grande do Sul, Hospital São Lucas, Porto Alegre, RS, Brazil.

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<http://dx.doi.org/10.1016/j.rboe.2017.02.002>

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imuno-histoquímico eram característicos de tumor de Pindborg. Atualmente, a paciente é acompanhada no Serviço de Ortopedia e Traumatologia da nossa instituição e apresenta boa evolução. O relato descreve o caso de uma paciente com uma neoplasia rara que foi investigada por muito tempo e com auxílio de muitos exames. O tumor de Pindborg foi sugerido com hipótese diagnóstica devido às características e ao comportamento neoplásico da lesão. A despeito de ser uma lesão mais comumente observada no âmbito odontológico, a neoplasia da paciente mostrou-se compatível com o diagnóstico. Assim, mesmo se tratando de um tumor com características benignas, há a necessidade de acompanhamento por longo tempo, haja vista o alto grau de recorrência do tumor.

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Introduction

Pindborg tumor, also known as calcifying odontogenic epithelial tumor, is a highly rare neoplasm, characterized by local invasiveness and presenting amyloid material. This neoplasm emerges as a painless mass of slow growth, with no predilection for gender. It affects patients between the second and sixth decade of life, mainly in the fourth decade. The vast majority of these tumors are intraosseous masses; only 6% of them are extraosseous. Upon radiological examination, the tumor is usually multilocular and septated or, less commonly, unilocular and radiolucent, and calcifications within the lesion are sometimes observed.

Case report

A 32-year-old female patient, originally from Bolivia, was admitted to this hospital in August 2009 with a complaint of pain in the right knee for over a year. She denied previous surgeries on the area, as well as using any medications. Patient presented with restricted mobility of the right lower limb and pain upon palpation in the region of the right femoral epicondyle. On the 13th of the same month, she was referred to a surgical procedure – femoral bone biopsy. The specimen, which measured 1.6 cm, was positive for amyloid using Congo red staining. In a scintigraphy, performed on August 19, an irregular radiomarker concentration was observed in the distal third of the right femur. A new biopsy, performed on November 9th, showed bone fragment with dense atypical cell infiltrate and positivity for amyloid using Congo red staining. In the radiological examination, an insulating osteolytic lesion was identified in the inner margin of the distal end of the femur, with septations in its interior. With the suspicion of large cell tumor, patient underwent tumor resection surgery on December 21, 2009. Microscopic examination of the surgical specimen found a tumoral mass partially coated by adipose and muscle tissue, measuring $6.5 \times 4.1 \times 3.4$ cm. When dissected, the lesion was characterized by its whitish, firm, and matte color. Part of the distal portion of the femur and tumor were resected. Patient had a good recovery after surgery. Tissue material was sent to the HSL Pathology Laboratory. Initial diagnosis was unclassified neoplasm; subsequently, an immunohistochemical analysis was performed, which led to a diagnosis compatible with mesenchymal neoplasm with



Fig. 1 – Post-surgery radiograph of the lesion indicating the presence of surgically-inserted orthopedic cement material.

low cell proliferation and amyloid production. The case was referred to consultation in the United States. A surgical follow-up magnetic resonance imaging indicated the presence of surgically-inserted orthopedic cement material, which was also observed on X-rays (Fig. 1), as well as other bone and musculotendinous structures within the limit of normality. In October 2010, a bone biopsy was performed for histological control, which presented no histological changes and was negative for neoplasia. Finally, in a review report of the case made on November 30, 2010, it was indicated that the histological aspect of the neoplasia, as well as its immunohistochemical profile, were compatible with Pindborg tumor, with a positive amyloid profile assessed with Congo red staining. Patient, now aged 38 years, has had a good evolution, with improvement of the mobility of the right lower limb, and is being followed-up at the HSL orthopedics and traumatology clinic. In a follow-up consultation in January 2016, she still presented good limb mobility, with no complaints (Figs. 2 and 3).



Fig. 2 – Post-surgical recovery limb flexion.

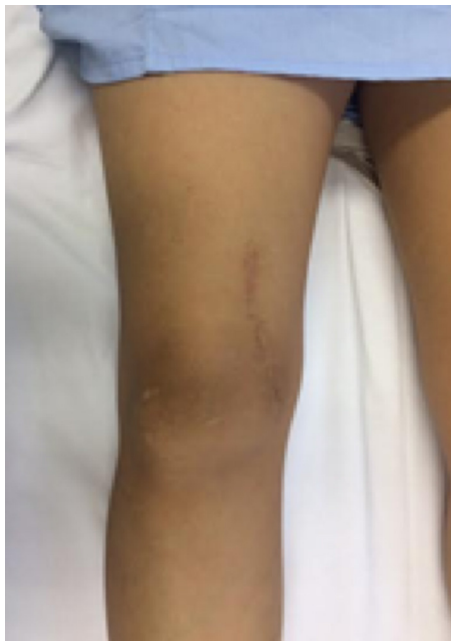


Fig. 3 – Post-surgical recovery limb extension.

Discussion

Described in 1958 by Jens Jorgen Pindborg, the tumor that bears his name is a very rare neoplasm. There are around 200 cases described in the current literature.¹ Description of this neoplasm shares many features with ameloblastoma, but Pindborg tumor is less aggressive and presents a slower growth.² It is known that this type of tumor requires a long follow-up period, because there is a high risk of tumor recurrence if it has been incompletely resected. According to the literature, tumor recurrence rate is around 15%; in these, the frequency of lesions that were treated with curettage is high. In spite of presenting high mitotic activity, a characteristic of malignancy, Pindborg tumor is a benign neoplasia with good prognosis.^{3,4} The histological aspects of this neoplasia include fibrous stroma, islands of polyhedral epithelial cells, homogeneous amyloid content, eosinophilia, and positive reaction to Congo red staining. In some cases, it is possible to find focal areas of clear cells, called rare variants of clear cells, which present a less favorable prognosis for the patient.^{3,5} Treatment for this type of neoplasia consists of surgical removal with sufficient safety margin to prevent recurrences.¹

Conflicts of interest

The authors declare no conflicts of interest.

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