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Case Report

Plexiform Fibrohistiocytoma in the Patella

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Abstract

The patella is an unusual location for any primary or secondary bone tumor, and would rarely be considered a neoplasm in the etiology of anterior knee pain. Plexiform fibrohistiocytic lesions are tumors of very low recurrence. This neoplasm mostly occurs in the second decade of life and its predilection is towards the upper limbs between 40 and 50%, lower limbs between 9-20% Patellar tumors are rare and usually affect young patients. They are often benign, in a proportion of 70%, we present a 17-year-old male case who consults for pain at the level of the right knee of 3 months of evolution and in the studies carried out there is osteolytic image at the level of the kneecap and in the pathological study a plexiform fibrohistiocytoma is diagnosed, total pathelegmy and reconstruction of the extensor mechanism was performed.

Keywords: Plexiform; Fibrohistiocytoma; Patella

Introduction

Plexiform fibrohistiocytic lesions are tumors of very low recurrence. The first case reported was in 1988 by Enzinger and Zang [1-3]. Where 65 cases were analyzed in which they presented as small, low-growth nodules with characteristics of being asymptomatic. This neoplasm mostly occurs in the second decade of life and its predilection is towards the upper limbs between 40 and 50%, lower limbs between 9-20% [2], trunk between 8-15% and head and neck between 10%. These lesions arise from the soft tissue at the level of the dermoepidermal junction and the subcutaneous cellular tissue [1]; Patellar tumors are rare and usually affect young patients. They are often benign, in a proportion of 70% [4], osteosarcoma being the most frequent malignant entity, in 6% of cases. The most common types found in this location are giant cell tumors (GCTs) and chondroblastomas [5]. For this reason, we want to report the following case, since there is currently little literature on these lesions, which originate at the bone level with plexiform fibrohistiocytic characteristics.

Case Report

A 17-year-old male presenting with pain in the right knee of approximately 3 months of evolution, of sudden onset, with mild intensity, without irradiation, which is exacerbated by physical activity and improves at rest, is evaluated in the outpatient clinic

- **Physical Examination:** Right knee: Claudicating gait, slight increase in volume with flushing, warmth and tenderness in the anterior aspect of the knee in relation to the insertion of the quadriceps tendon, limitation to the extension of the knee and deformity in 10º flexion, no palpable masses.

A multidisciplinary assessment, staging, biopsy are performed

- **Blood tests:** No alteration.

Computerized axial tomography

Osteolytic image at the level of the upper middle pole of the patella with rupture of the cortical in superior pole towards the cartilaginous portion that imprints giant cell tumor

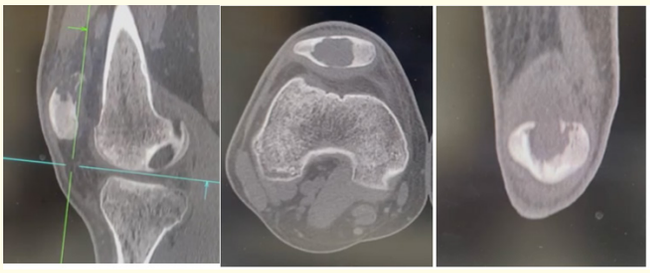

Figure 1

Anatomical pathological findings

- **Macro:** In Pathological Anatomy, the right patella is received from hemipatelectomy, measuring 6x5x3cm with a slice of white skin. The surface is smooth, greyish, firm in shape with a poorly defined, whitish thickening, alternating with areas of hemorrhagic bone tissue. Samples are taken for microscopic study.
- **Micro:** In the microscopic study, an infiltrative tumor growth with the formation of small nodules was observed, consisting of histiocytic cells of the epithelioid type and giant cells of osteoclast type surrounded by spindle cells type fibroblasts. In addition, some areas of hemorrhage and necrosis.
- **Immunohistochemistry:** An IHC study was performed to determine the degree of malignancy. Fibroblast cells expressed positive Actin and Vimentin. Histiocytic cells and giant cells were positive for the CD68 antibody. The tumor tested negative for the S100 antibody. The injury had a Ki67 of 5%.

All this led us to conclude the case as a plexiform fibrohistiocytic tumor, with close follow-up of the patient.

Discussion

The patella is an unusual location for any primary or secondary bone tumor, and would rarely be considered a neoplasm in the etiology of anterior knee pain. Therefore, the diagnosis of a tumor within this location is delayed in most patients. Benign lesions are more common, accounting for 70% of tumors involving the kneecap [4]. The most common is benign giant cell tumor (GCCT), followed by chondroblastoma and aneurysmal bone cyst [6]. Other lesions that can affect the kneecap include metastasis, lymphoma, Paget's disease, osteosarcoma, chondrosarcoma, osteomyelitis,

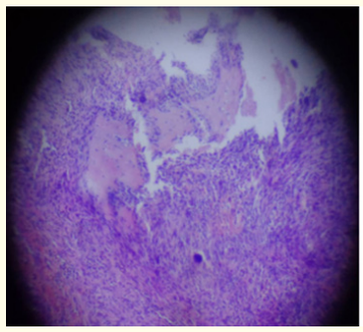

Figure 2: Plexiform fibrohistiocytoma. Panoramic view.

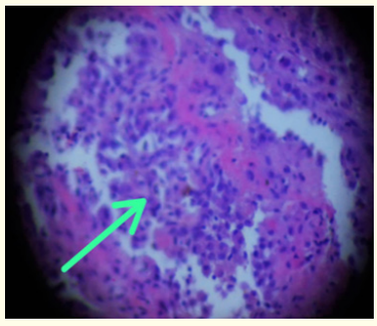

Figure 3: Epithelioid cell nodule.

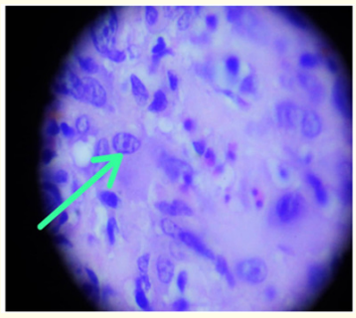

Figure 4: Epithelioid cell.40x.

gout, brown tumor, osteoma, or solitary bone cyst [7,8]. According to a study of 27,403 primary bone tumors conducted by the Bone and Soft Tissue Tumor Committee of the Japanese Orthopaedic Association covering the period from 1972 to 2003, 13,860 of the tumors involved the bones of the lower extremities. Of these, 75 (0.5%) affected the patella, 71 (94%) were benign, representing 22 (31%) of the cases, i.e., the (GCCT) were 0.08% of all patella tumors and 0.15% of the bone tumors of the lower extremities; Campanacci also reported that less than 1% of (TCGB) arise in the kneecap ([9]).Among the differential diagnoses to be taken into account in the face of this tumor, we have: plexiform neurofibroma, plexiform schwannoma, cellular neurotecoma, fibrous hamartoma

of childhood, deep benign fibrous histiocytoma, soft tissue giant cell tumor and myofibromatosis. Fibrohistiocytoma and Soft Tissue Giant Cell Tumor occur mainly in adult patients, ours is an 18-year-old adolescent, in addition, these tumors have a more solid growth than plexiform. Fibrous hamartoma has a primitive cellular myxoid component with the presence of mature adipose and fibrous tissue, in the absence of giant cells. These histological elements were not found in the tumor in question. Myofibromatosis tends to be well defined, a characteristic absent in our case studied, in addition, it presents areas reminiscent of hemangiopericytoma, a detail not found in our study. The labeling study of the S100 protein helped us to rule out plexiform neurofibroma, plexiform schwannoma and cellular neuroteoma [10].

Plexiform fibrohistiocytic lesions are tumors of very low recurrence. The first case reported was in 1988 by Enzinger and Zang [1-3]. where 65 cases were analyzed in which they presented as small, low-growth nodules with characteristics of being asymptomatic. This neoplasm mostly occurs in the second decade of life and its predilection is towards the upper limbs between 40 and 50%, lower limbs between 9-20% [2], trunk between 8-15% and head and neck between 10%. These lesions arise from soft tissue at the level of the dermoepidermal junction and subcutaneous cellular tissue [1].

Conduct

Partial patelectomy of the upper pole and biopsy were performed due to suspicion of Giant Cell Tumor plus reinsertion of the quadriceps tendon, the results of pathological anatomy were obtained, the case was rediscussed and it was decided to complete the patelectomy and perform a mapping of the soft tissues which were free of tumor and reconstruction of the extensor mechanism. And follow-up by outpatient consultation.

Conclusion

Plexiform fibrohistiocytic lesions are tumors of very low recurrence, where only 9% of this type of lesions occur in the lower limbs with bone involvement, since this type of lesions arise from soft tissue at the level of the dermoepidermal junction and the subcutaneous cellular tissue, although the presence of this type of bone tumor in the patella should be considered rarely. Patellar tumors are rare and usually affect young patients. They are often benign, at a rate of 70%.

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