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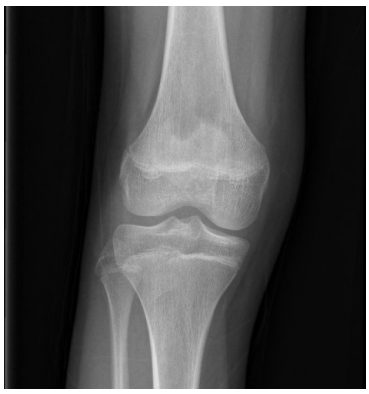

Figure 1. Plain X-ray of the right knee showed a well-circumscribed lytic lesion with sclerotic margin on the superior aspect of the patella.

The patient was a 12-year-old girl with localized pain in the right suprapatellar region for 4 months. Laboratory workup showed normal results (white blood cells = 6470/µL, hemoglobin = 13.4 g/dL, platelet = 210 000/µL, erythrocyte sedimentation rate = 4 mm/h, calcium = 9.4 mg/dL, phosphorus = 4.8 mg/dL, parathyroid hormone = 36 pg/mL, alkaline phosphatase = 828 IU/L, lactate dehydrogenase = 283 IU/L). Plain X-ray of the right knee showed a well-circumscribed lytic lesion with a sclerotic margin (Figure 1). Right knee magnetic resonance imaging (MRI) with and without contrast exhibited a 14 × 12 mm well-defined round lesion on the superior aspect of the patella which was hyperintense on T1W/SPIR images (fat suppressed sequences), isointense on T2W images and showed homogenous enhancement after contrast administration (Figure 2). A thin sclerotic margin and perilesional edema were observed. The previous findings along with internal enhancement suggested radiologic differential diagnoses of benign lesions such as chondroblastoma. Subsequently, surgical resection of the patellar mass was undertaken and histologic evaluation revealed bony trabecula surrounded by large histiocytes, lymphocytes, and plasma cells (Figure 3A). The large histiocytes showed emperipolesis (engulfed lymphocytes within vacuoles) (Figure 3B).

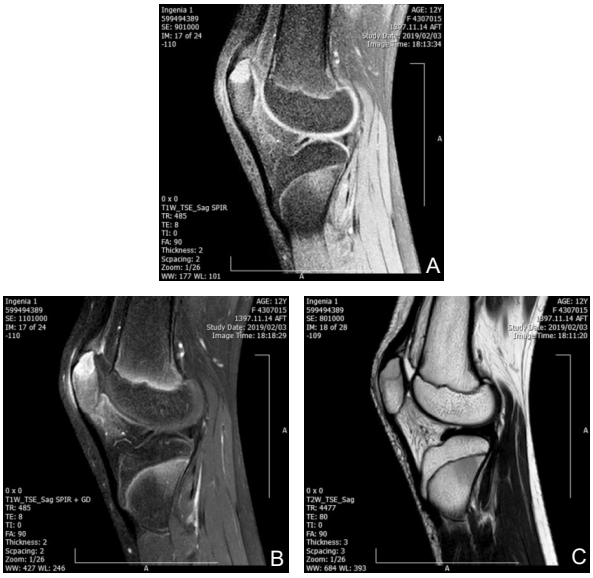

Figure 2. **(A)** MRI T1/SPIR image showed a well-defined hyperintense lesion in the superior part of the right patella. **(B)** MRI T1W image after contrast administration showed homogenous enhancement. **(C)** MRI T2W image showed a well-defined isointense lesion.).

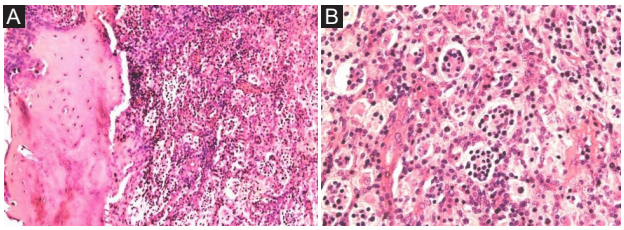

Figure 3. (A) A bone segment surrounded by mononuclear cells mainly composed of histiocytes, lymphocytes and plasma cells (hematoxylin and eosin, magnification x200). (B) Histiocytes showed lymphocytophagocytosis (emperipolesis) (hematoxylin and eosin, magnification x400).

What is your diagnosis?
See the next page for your diagnosis.

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Photoclinic Diagnosis

Rosai-Dorfman Disease Arising in Patella

Rosai-Dorfman disease (RDD) is a rare non-Langerhans cell histiocytosis with a prevalence of 1:200,000 in the United States. RDD is characterized by S100+, CD68+, and CD1a- histiocytes and emperipolesis (not a constant finding). The classical manifestations of RDD are bilateral massive and painless cervical lymphadenopathy. Extra-nodal involvement is a non-classic presentation which can occur in 43% of cases and can lead to cutaneous, central nervous system, head and neck, intrathoracic, genitourinary, gastrointestinal, bone and hematologic manifestations.[1-3]

Bone involvement occurs in less than 10% of RDD patients.[1] According to a recent systematic review on 108 patients with osseous RDD with a mean age of 31 years and a slight female predominance, the cranium, facial bones and tibia were the three most commonly involved bones, followed by the spine/sacrum, femur, pelvis, humerus, tarsal bones, ribs, fingers, clavicle, sternum, scapula, carpal bones, radius and fibula in decreasing order of frequency. The three most common concomitant extra-osseous manifestations were soft tissue, lymph node, and sinus involvement. Most of the bone lesions were lytic on imaging (80%), some were both lytic and sclerotic (18%), and a few were only sclerotic (2%).[4] Bone involvement in RDD is generally associated with a good prognosis, unlike some other extra-nodal types of this disease, especially in lungs and kidneys, which can be fatal.[1,5] There is only one case of RDD with patellar involvement in the background of the disseminated form of RDD (reported in a Tunisian article).[6] The present patient is the first case of osseous RDD with *de novo* patellar involvement and merits attention as a differential diagnosis for lytic-sclerotic patellar lesions.

There are different treatment modalities to RDD according to its clinical presentation. An observational strategy can be selected for uncomplicated lymphadenopathy or asymptomatic cutaneous disease. Surgery could be curative for unifocal disease as in the present case. Corticosteroids could be effective treatments for bone, central nervous system, orbital, and autoimmune hemolytic anemia-related disease. Chemotherapy with cladribine or methotrexate/6-mercaptopurine or vinca alkaloids can be therapeutic in refractory, disseminated or life-threatening diseases. Other treatment modalities like immunotherapy, targeted therapy and radiotherapy are also helpful in certain situations.[6]

Authors' Contribution

MS involved in pathology interpretation, image preparation and drafting the manuscript. BP involved in patient clinical management and drafting the manuscript. NP involved in interpreting radiologic images and editing manuscript.

Conflict of Interest Disclosures

The authors declare that they have no conflict of interest..

Ethical Statement

An informed consent was obtained from the patient's parents for publication of this photoclinic.

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