PIGMENTED VILLONODULAR SYNOVITIS IN CHILDREN: A MISSED DIAGNOSIS

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Introduction

Pediatric joint pain and swelling is a frequent presentation with numerous possible aetiologies. The clinical profile and relevant laboratory investigations often lead to the correct diagnosis; however some patients present a diagnostic conun-drum. Pigmented villonodular synovitis (PVNS) is a rare entity that may afflict children and be easily missed. This case discusses the aetiopathogenesis of this unusual entity and the role of imaging in its correct diagnosis and management

Case report

A patient aged 1 year 3 months without particular pathological conditions who presents for chronic polyarthralgia evolving over 3 months, inflammatory affecting both knees and both ankles. All evolving in a context of unquantified fever and preservation of general condition. the clinical examination notes a conscious patient hemodynamically stable, aperitic at 36.8 ,FC: 92b/min, FR: 30c/min.With symmetrical bilateral arthritis of both knees and ankles, with limitation of movements without other extra-articular signs.

An ultrasound suggests a large bilateral intra-articular effusion a joint puncture was performed with 3040 predominantly lymphocytic leukocytes with negative culture.

The patient was initially treated for septic arthritis.

Given the lack of improvement, the patient consulted our training with a positive infectious

assessment: VS 186mm at the 2nd hour, CRP at 49 mg/l.

For the immunological assessment C3 and C4: normal, CANCA PANCA: normal.

A joint ultrasound was done again showing bilateral villonodular synovitis of the knees.On MRI: villonodular synovitis.

The patient was initially placed on NSAIDs and given the lack of improvement, the patient was placed on methotrexate 10 mg/week, with very good improvement.

Discussion

PVNS is a rare, benign proliferative disorder of the synovial membrane.
its annual incidence rate is ~1.8 cases per million.PVNS usually presents in adults between 30 and 40 years of ageand is rarely observed in childhood.The main clinical manifestations of PVNS include repeated bleeding, swelling,and sometimes pain in the affected joints. In late-stage PVNS,joint dysfunction occurs because bone and cartilage .MRI has been recognized as the best imaging method for diagnosing PVNS. The differential diagnosis of PVNS includes infection, tumor,hemarthrosis, and rheumatoid arthritis.PVNS is easily misdiagnosed because of its rarity. In extant literature reports,PVNS diagnoses have almost always been delayed.In thelargest report, which involved 237 patients, the median delayfrom initial clinical symptoms to final diagnosis was ~18months.Because PVNS has a much lower incidence inpediatrics, this disease is more easily misdiagnosed in children.PVNS requires radical treatment that combines prostheticarthroplasty and synovectomy.However, frequent recurrence after synovectomy has been observed.

conclusion

Pigmented villonodular synovitis is an exceptional condition in children. The clinical, biological and imaging presentation is not specific and a histology sample is required for certain diagnosis. Because of this lack of specificity, together with the rare occurrence of the disease, late diagnosis is not uncommon, making correct management an even greater challenge in the growing child.