

IMAGES IN RHEUMATOLOGY

## Juvenile presentation of a pigmented villonodular synovitis of the wrist

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### CASE REPORT

A 17-year-old male patient, with no relevant personal history, was referred to the Rheumatology appointment due to swelling of the left wrist. The patient reported the appearance of a painless swelling on the dorsal surface of the left wrist for about 4 years, with progressive growth, without any other associated symptoms. Physical examination showed nodular swelling on the dorsal surface of the left carpus, approximately 3 cm in diameter, painful on palpation, without other inflammatory signs and without limitation of wrist joint amplitudes (Figure 1). The remaining joint examination was unremarkable.

At this moment, the patient had a magnetic resonance of the wrist, which revealed a large lobulated formation with irregular contours on the dorsal surface of the wrist, hypointense in T1 sequence and with intermediate signal in T2 sequence (Figure 2A-B). The main diagnostic hypothesis suggested from these findings was a synovial proliferative process of undetermined nature.

In his first evaluation in Rheumatology Department an ultrasound of the left wrist was performed, revealing an exuberant intercarpal synovial hypertrophy, with more than 3 cm in extension in the axial and coronal



**Figure 1.** Nodular swelling on the dorsal surface of the left carpus at physical examination.



**Figure 2.** MRI of the left wrist in a coronal plane showing a large lobulated formation (white star) with irregular contours on the dorsal surface of the wrist, from the plane of the dorsal surface of the hamate to the plane of the scaphoid joint with the trapezium and trapezoid; the lesion presents an hypointense signal in T1 sequence (A) and an intermediate and heterogeneous signal in T2 (B).

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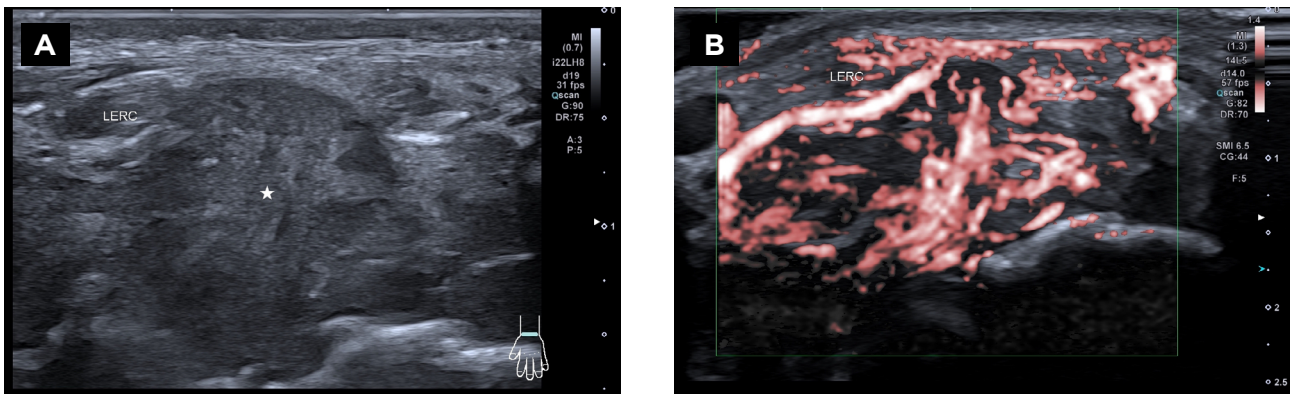
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Submitted: 22/03/2023

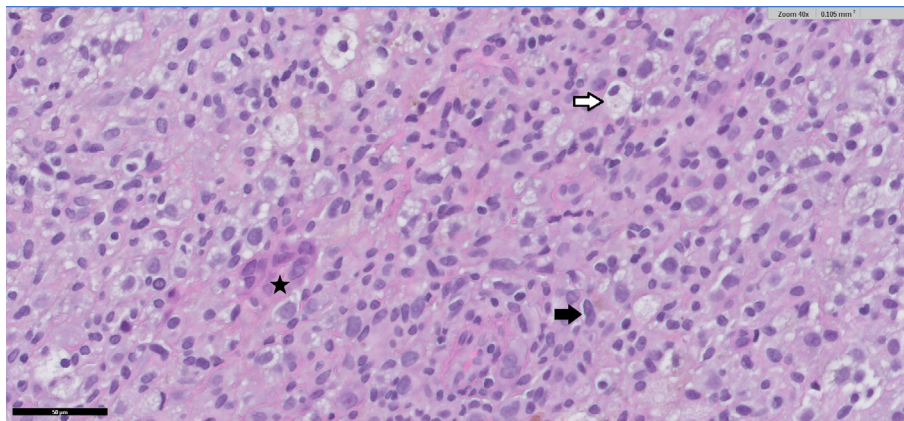
Accepted: 25/06/2023

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**Figure 3.** Ultrasound of the left wrist in a transverse view. Grey scale image (A) showing an exuberant intercarpal synovial hypertrophy (white star) with marked SMI signal (B). LERC: left extensor radialis carpi tendon.



**Figure 4.** Pigmented villonodular synovitis. Tumor with mononuclear cells (black arrow), multinucleated giant cell (black star) and foamy macrophages (white arrow). Hematoxylin and eosin stain 400X.

planes, with marked SMI (superb microvascular image) signal (Figure 3A-B).

The laboratory work up showed no increased inflammatory markers and negative rheumatoid factor, anti-citrullinated peptide antibodies and HLA-B27.

An ultrasound guided synovial biopsy of the left carpal swelling was performed. The microbiological, mycobacteriological and mycological exams of the synovial membrane were negative. Histologically, the tumor displayed mononucleated cells with a histiocytic phenotype, foamy macrophages, sometimes with hemosiderin, and osteoclast-like giant cells (Figure 4). These morphological aspects were consistent with a pigmented villonodular synovitis.

The patient was then referred to the Orthopedic Department, having been submitted to surgery for tumor excision. To date, no relapses of the lesion were reported.

## DISCUSSION

Pigmented villonodular synovitis is a rare benign lesion

that results from the proliferation of the articular, bursal or tendon sheath synovial membrane, predominantly affecting females, with a peak incidence between the 3rd and 5th decades of life, being more rarely described in children<sup>1-3</sup>.

With this case we highlight the importance of considering pigmented villonodular synovitis in the differential diagnosis of joint and peri-articular swelling in juvenile patients, although its uncommon frequency. It also emphasizes the importance of synovial biopsy in the establishment of a definitive diagnosis, making it a central complementary method in the diagnosis of this condition.

## REFERENCES

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