Extended Pigmented Villonodular Synovitis of the Hand

Sinovitis villonodular pigmentada extendida de la mano

Carlos Irisarri[1] Javier Yañez Calvo[2]

[1] Consultant in Hand Surgery, Hospital Vithas Nuestra Señora de Fátima, Vigo, Spain
[2] Complejo Hospitalario Universitario de Vigo, Spain

Address for correspondence Carlos Irisarri, MD, PhD, Consultor de Cirugía de la Mano, Hospital Vithas Nuestra Señora de Fátima, Vía Norte, 48, 36206, Vigo, Pontevedra, Spain (e-mail: irisarri@iies.es).

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Abstract

We present the case of a 36-year-old woman with pigmented villonodular synovitis (PVNS) of the right hand. The magnetic resonance imaging (MRI) showed that the lesion had spread over an unusually wide area. Despite this, after one single operation, there has been no relapse after 5 years, with both the functional and cosmetic results remaining satisfactory.

Keywords
- pigmented villonodular synovitis
- hand

Resumen

Se presenta un caso de sinovitis villonodular pigmentada, que afecta a la mano derecha de una mujer de 36 años. Como mostró la resonancia magnética, la tumoración presentaba una inusual gran extensión. Pese a ello, tras una única cirugía, no se ha detectado recidiva del tumor a los 5 años de la cirugía, y tanto el resultado estético como el funcional continuaran siendo satisfactorios tras dicho período.

Palabras clave
- sinovitis villonodular pigmentada
- mano

Introduction

Chassaignac[1] first reported giant cell tumors of the tendon sheath in 1852, and he considered the lesions to be sarcomata ('cancer de la gaine des tendons'). The early literature was confusing, as many different terms were used to label it, such as giant-cell tumor, and benign synovioma. Masson[2] named these lesions xanthoma, subdividing them into a nodular mass form originating in the fibrous tissue of the tendon sheaths, and a second more extended type. His paper includes diagrams of a case in which the tumor was occupied the whole of the ulnar and radial bursae, and extended through the second and third metacarpals to the dorsum. He reported that the tumor was made up of different sized yellowy-orange lobes, surrounded by a thin capsule.

In 1941, Jaffe et al[3] introduced the term pigmented villonodular synovitis (PVNS), reviewing twenty cases involving joints, tendon sheaths and bursae. They observed that the 'nodular' and 'diffuse' lesions were histologically similar. The benign course led them to conclude that the lesions were not in fact tumors, but an inflammatory response to an unknown agent. On the other hand, Rao and Vigorita[4] supported the concept that PVSN was a neoplastic proliferation of synovial fibroblasts and histiocytes. Jones et al[5] also maintained that PVSN originated from histiocytes. Abdul-Karim et al[6] analyzed seven PVSN cases through flow cytometry, and the DNA pattern in several of the studied cases study, along with the presence of specific chromosomal abnormalities, led the authors to attribute to PVNS a neoplastic nature.

O'Connell et al,[7] in an effort to clarify the immunohistochemical profile of PVNS, examined the pathological tissue of 12 PVNS samples fixed in formalin, and their results obtained suggested that PVNS are synovial cell tumors. Alguacil-Garcia et al[8] also considered PVNS to be proliferative lesions of the synovial cells.

The histological examination is characterized by the proliferation of either the fibroblastic or the histiocytic mesenchymal cells, or both, beneath the synovial or tenosynovial lining

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cells. The appearance of "foam cell" type macrophages and iron deposits seem to be secondary changes, and are usually seen in the periphery of expanding tumor nodules.

Fisk[9] attributed PSVN to repeating minor trauma, which is untenable in most cases. In summary, despite all these efforts to clarify the pathogenesis, the true etiological agent is still unknown.

In 1968, Byers et al[10] classified the giant cell tumors (GCT) within nodular types, which are commonly seen in the finger tendon sheaths (GCTTS). They also classified PVNS as a second diffuse type, commonly seen in large joints (especially the knee), and only rarely in the wrist or the hand.

Pigmented villonodular synovitis usually affects young adults, with approximately equal numbers of men and women. In the nodular forms, fingers lesions are the most common, with the diffuse form usually affecting the knee, where occasionally both knees are involved.

"Extended" PVNS located in the hand and/or wrist is rare. Swelling is always present in cases of long duration as well as differing degrees of joint stiffness. Bony changes are often present, and include cortical erosions and/or intra-osseous cystic lesions, which during the initial stages are difficult to identify. Masson[2] pointed out the 'involvement of the carpal bone at the base of the thumb', although no X-rays were included in his paper.

Moser and Madewell[11] reported a case with a 3-year follow-up that showed a slow progressive enlargement with penetration in distal radius and triquetral bone. Schajowicz and Blumenfeld[12] reported a PVNS in a 61-year-old man, with 20 years of evolution, where extensive penetration of the tumor in the distal radius was observed. Enzinger and Weiss[13] reported a case affecting an adolescent patient, with progressive distal radius and triquetral bone involvement after a 3-year follow-up. Patel and Zinberg[14] also published the case of PVNS in a woman, with involvement of the hamate bone, as well as the fourth and fifth metacarpal bases.

Case Report

A 36-year-old woman attended our clinic, with a voluminous soft tissue swelling in her right hand, not stuck to the skin, which had been gradually worsening over several months, with progressive discomfort and functional impairment. The patient did not refer to any relevant traumatic background.

In the X-ray (►**Fig. 1**) no bony erosions were appreciated, but on the magnetic resonance imaging (MRI) different tumor masses were observed (►**Figs. 2–4**) both in the palmar and dorsal area, displacing the extensor tendons.

Based on the clinical data and obtained images, we suspected a synovial non-malignant tumor, and so we decided to operate without prior biopsy. The operation was performed with anesthesia through brachial plexus blockage, and we made a dorsal approach. The excision of the multi-nodular masses, which were surrounded by a fine pseudo-capsule, was relatively simple. The extensor tendons were still preserved. Next, the complete removal of the solid bigger nodular mass located in the palmar-radial area adjacent to scaphotrapezial joint was easily performed because it was surrounded by a thick pseudo-capsule.

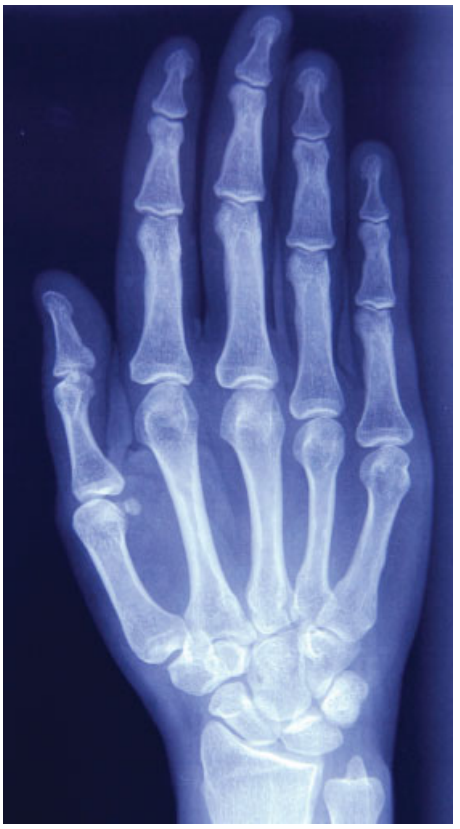

Fig. 1 Preoperative posteroanterior X-ray. No osseous pressure erosions or intra-osseous cystic lesions are visualized.

Much more difficult was the excision in the palm, which was complicated by the need to spare the neurovascular bundles ►**Fig. 5**. The flexor tendons of the middle finger were fragile, and two longitudinal and one horizontal yellowish colored masses surrounded by a very fine pseudo-capsule were completely excised. Later, however, it was very difficult to establish the border between the free tissue and the tumor

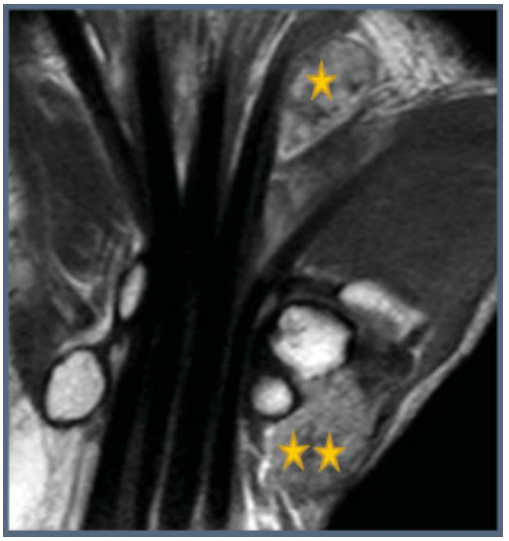

Fig. 2 Magnetic resonance imaging (MRI): palmar mass (marked with a yellow asterisk), and another close to the scaphotrapezial joint (marked with two yellow asterisks).

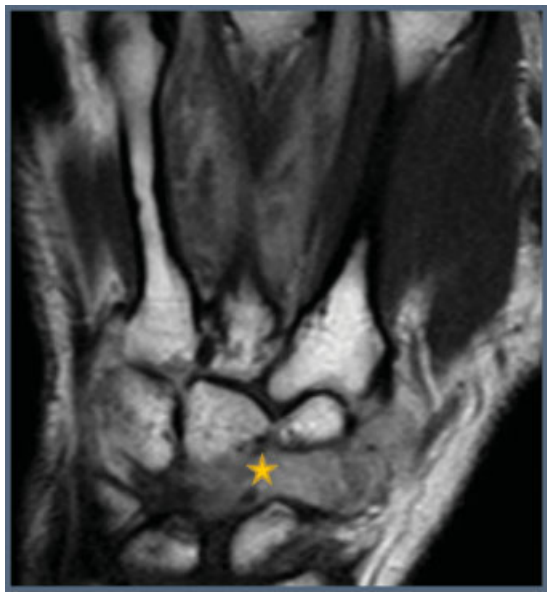

Fig. 3 MRI: horizontal mass (yellow asterisk) at carpal level. Volar intrinsic muscles in the second and third intermetacarpal spaces involved.

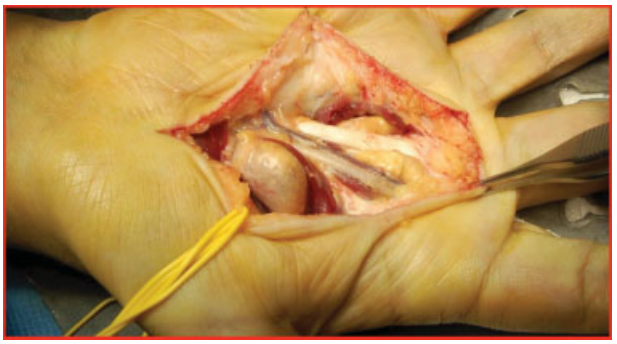

Fig. 5 Intraoperative image of the volar masses.

tissue. Thus, we decided to only excise the more affected areas of these muscles; the flexor tendons were still relatively well preserved. After performing this step, the tourniquet was released, and we made a meticulous coagulation with a bipolar cautery.

The histological examination confirming PVNS was the following:

Macroscopic examination: yellowish-orange colored soft tissue fragments, with well delimited nodules, a thin fibrous capsule and occasional fibrous septa.

Microscopic examination: profuse synoviocyte type cells, with an inconspicuous cytoplasm and round central nuclei. Some of the cells had hemosiderin. A moderate amount of giant multinucleated cells were evident, with mitosis being scarce in some areas but abundant in others.

The post-surgical period was uneventful. A wrist splint, allowing for early finger movement, was maintained for 2 weeks, and for three months the patient had physiotherapy, with progressive recovery of wrist and finger movement.

Postoperative MRI was obtained two and four years after the surgery. No new soft-tissue masses appeared, nor any bone involvement ►**Fig. 6**. At the last visit, five years after the surgery, the patient mentioned that she was able to make her daily tasks satisfactorily, as well as continuing her job as a shop assistant in a clothes shop. She had recovered a full range of flexion and extension movements, both of the wrist and fingers, and only the adduction/abduction of the central long fingers were a little bit limited. Dorsal scar quality had an excellent appearance, and even though the palmar scar showed a discrete retraction in the radial angle of the zig-zag incision, the patient refused our proposal to carry out a Z-plasty correction. ►**Fig. 7**.

Discussion

There have been few reports of extended PVNS involving the wrist and/or the carpometacarpal area, as compared with the localized nodular form of the fingers. Jones et al[5] reviewed the clinical records patients seen at the Mayo Clinic during the ten-year period between 1954 and 1963, and four cases were found in the wrist, another four in the proximal part of the hand, and ninety-one GCT in the fingers.

In most of the series, PVNS tends to occur in young patients, with slightly more females affected than males (Enzinger and Weiss[13]). Typically symptoms have of long evolution, and include soft-tissue swelling, tenderness and limitation of movement.

In the 81 patients reported by Rao and Vigorita,[4] only 1, a 58-year-old man, presented PVNS in the wrist, which was located on the ulnar-volar side. Three years after the excision, he was free from relapse. Moreover, Schajowicz and Blumenfeld[12] reported a patient who underwent two successive operations due to the size of the tumor. The result was considered satisfactory 16 months after the excision. Mathews[15] reported a case involving the entire ulnar bursa, and he recommended a complete tenosynovectomy.

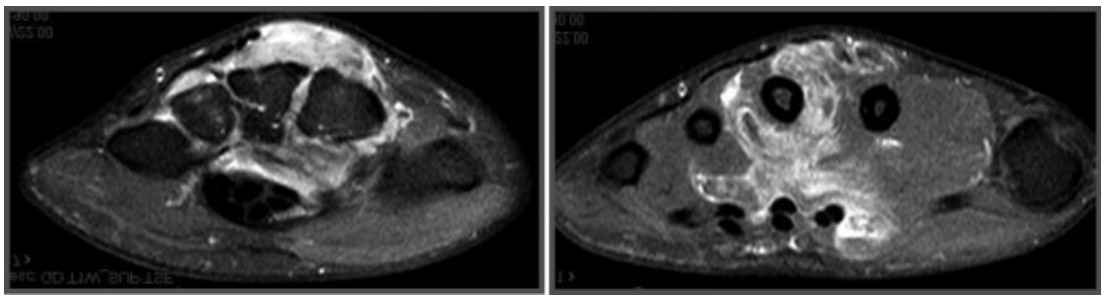

Fig. 4 (**A**) Axial MRI at the carpal level. (**B**) Axial MRI at the metacarpal level.

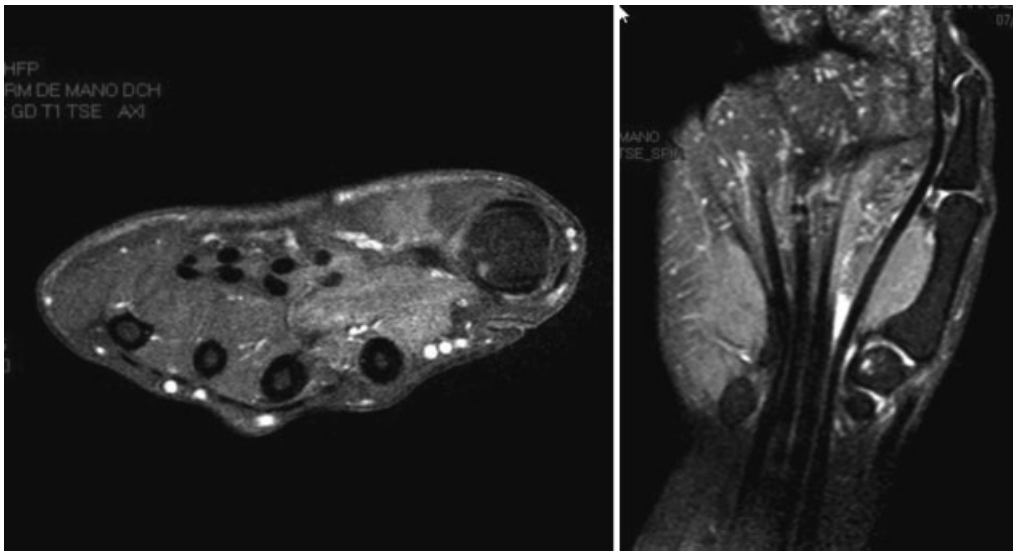

Fig. 6 In MRI scar tissue can be found, but no recurrence of the tumor.

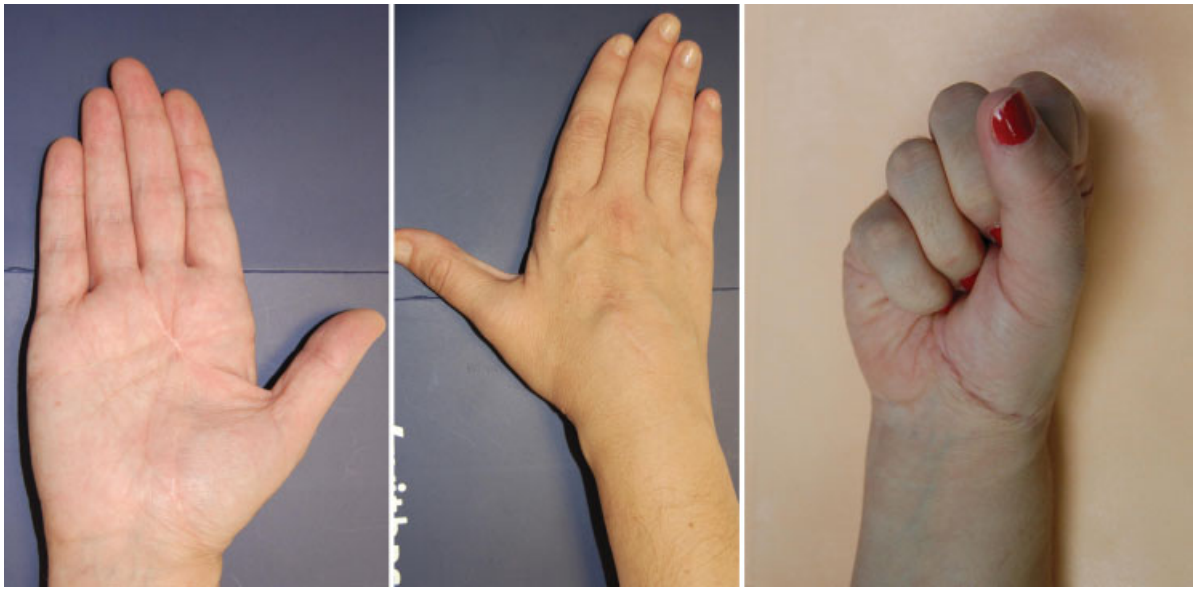

Fig. 7 Clinical aspect and function of the hand five years from surgery.

Histological studies are controversial, but most experts (Alguacil-Garcia et al,[8] Jaffe[16]) consider PVNS to be reactive and borderline proliferative lesions of the synovial cells. As Enzinger and Weiss[12] pointed out, from a practical point of view, these lesions should be regarded as locally aggressive, but non-metastasizing lesions. For this reason, wide excision appears to be the best choice, but a radical one may not be necessary.

With respect to the treatment, experts agree to perform an extensive synovectomy, but even performed in an aggressive way, high recurrence rates have been mentioned, especially in the knee joint. In the past, combined radiotherapy was widely recommended, but this frequently caused joint stiffness. Fortunately, in some cases satisfactory long-term results were obtained, even if the PVNS was not been completely excised. Reported PVNS cases located in the hand and wrist are very scarce, which makes it impossible to carry out a valid statistical study on post-operative recurrences.

In our literature review, we have insufficient data to ascertain either the recurrence rate or the risk factors such as age, gender, size, and location. It is probable that intraosseous invasion (not bone indentation due to tumor pressure) may be associated with a higher risk of recurrence. In any case, experience of PVNS is limited because of the low numbers of reported cases, so it is impossible to draw any definite conclusions. Even so, careful long-term follow-up is mandatory in all patients treated for extended PVNS.

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