Pigmented villonodular synovitis: a recurrent case with atypical location and extra-articular extension

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ABSTRACT

Pigmented Villonodular Synovitis (PVNS) remains a diagnostic challenge because of its non-specific presentation and subtle radiological findings. This uncommon entity is characterized by a benign synovial membrane proliferation of joints, tendon sheaths and bursas. The authors report a case of recurrent PVNS affecting an uncommon location (the first left metatarsophalangeal joint), which extended well beyond the joint margins on recurrence, in a patient with non-specific complaints of joint pain and swelling.

Keywords: Pigmented Villonodular Synovitis; Recurrent; Synovial membrane.

INTRODUCTION

PVNS is an uncommon entity characterized by a benign synovial membrane proliferation of joints, tendon sheaths and bursas[1,2]. There are two distinct clinical forms of this disorder: localized (LPVNS) and diffuse (DPVNS) pigmented villonodular synovitis. In the diffuse form, the entire synovium of an affected joint or structure is involved, exhibiting coarse villi, diffuse nodularity, often an heavily pigmentation (ranging from dark yellow to chocolate brown) and is accompanied with joint effusion and soft tissue mass[1-3]. The localized form involves only a portion of the synovial surface, tends not to be as darkly pigmented and has less villous proliferation but can have nodules, small tumefactions or pedunculated masses[1-3]. PVNS typically affects large joints in young adult patients, remaining confined within a synovium-lined joint and rarely extending beyond the joint capsule. This is a rare disease with an annual worldwide incidence of 1.8 patients per 1 million[1,2,4-9].

The authors report a case of recurrent PVNS affecting an uncommon location (the first metatarsophalangeal joint) associated with extra-articular extension on recurrence. Atypical PVNS presentations are challenging and may contribute to additional diagnostic and treatment delay.

CASE REPORT

A 60-year-old woman presented with a one-year history of progressive pain and swelling of the plantar surface of the first metatarsophalangeal (MTP) joint of the left foot. She complained of pain while walking and even at rest and referred no clinical benefit from non-steroidal anti-inflammatory drugs. She excluded previous trauma and denied other osteoarticular or systemic complaints. Six years before she had been submitted to the surgical resection of a nodular lesion of the first left MTP joint, whose histology revealed localized PVNS, with a good clinical outcome.

Physical examination of the left foot revealed a moderate swelling and redness of the left plantar forefoot, especially at the level of the first MTP and first joint space (Figure 1) which was tender to palpation but without restriction of joint motion.

The laboratory findings were unremarkable. The foot x-ray showed a soft tissue oedema surrounding the first MTP joint, but no joint or bone abnormalities. The foot MRI identified an extensive mass surrounding the first MTP joint and extending proximally to the first cuneo-metatarsal joint, with 6.5 cm of longitudinal diameter and 2.5 cm of transversal diameter (Figure 2)

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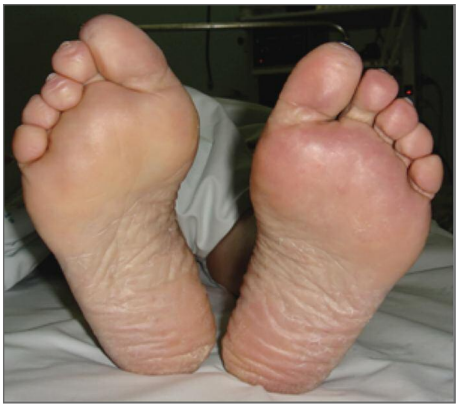

FIGURE 1. Plantar swelling of the left forefoot, particularly evident at the level of the 1st MTP joint and interdigital space

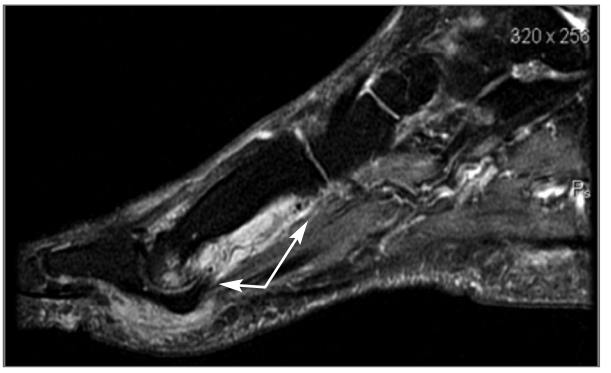

FIGURE 2. MRI of the left foot (sagital T1-weighted image): plantar soft tissue lesion involving the 1st MTP joint, extending from the proximal hallux phalange to the cuneo-metatarsal joint of the 1st toe (arrows)

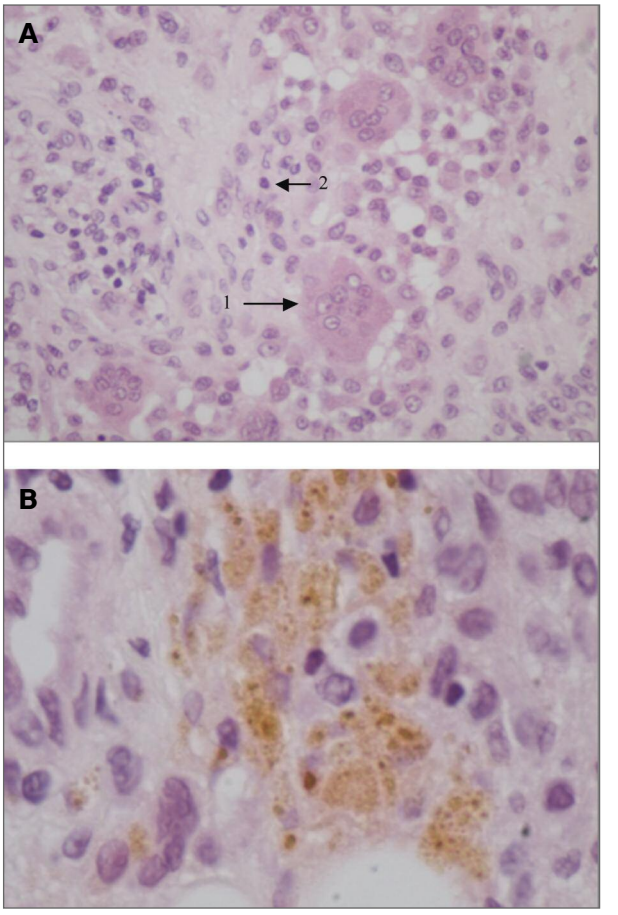

FIGURE 3. Histology of the lesion: **A)** Giant multinucleated cells (arrow 1) and histiocitic cells (arrow 2) in a synovial stroma. **B)** Hemosiderin deposition (brown) in a synovial stroma. H&E, original magnification x400

with intermediate signal on T1 and T2-weighted images. A CT-scan guided biopsy of the plantar soft tissue mass, at the level of the first left MTP joint, was performed and the histopathological study revealed the presence of histiocitic and giant multinucleated cells associated with hemossiderin deposits, in a synovial stroma, typical of PVNS (Figure 3).

The patient was submitted to surgery. A marked proliferation of the entire synovial membrane of the first MTP joint was found, associated with an extra-articular yellow-brownish mass located on its plantar aspect coinciding with the MRI images (Figure 4). A total synovectomy of the joint associated with a complete removal of the surrounding mass was performed. At 1-year follow-up there were no signs of clinical recurrence and foot MRI excluded the presence of joint or soft tissue lesions.

DISCUSSION

PVNS is an uncommon entity and the case we report has several peculiarities, which deserve attention. The monoarticular distribution is typical but the disease usually affects large joints[1-3,7,9,10], the knee being the most commonly involved joint, followed by the hip, ankle and shoulder (representing 80, 15, 5 and 2 % of the cases, respectively)[1,11-13]. More uncommon PVNS locations are the temporo-mandibular joint (52 cases reported)[14], wrist (25 cases reported)[15-21] and elbow (21 cases reported)[22]. Metatarsophalangeal joint involvement is even quite rare, with just 10 cases reported in the literature[23-31]. This shows that the hallux is not commonly affected in PVNS and this may delay appropriate diagnosis and treatment.

PVNS generally affects young adults in their third

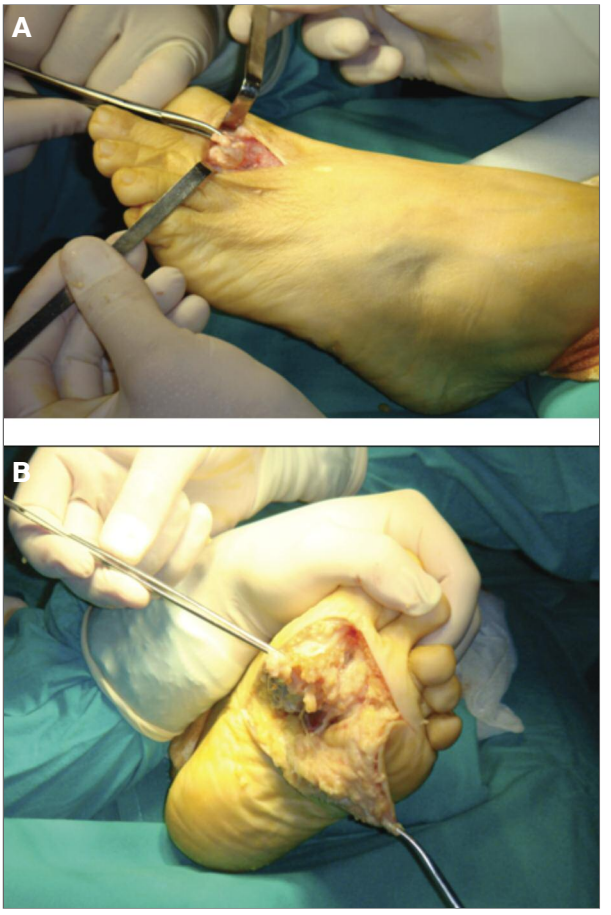

FIGURE 4. Surgical resection of the lesion. **A)** Total synovectomy of the 1st MTP joint. Presence of a yellow-brownish colour proliferative synovial membrane. **B)** Removal of the extra-articular plantar component of the synovial membrane lesion.

and fourth decades[1,2,4,9,32] and although sex distribution is not known, a slight female preponderance was found in large series[4]. Again, the presentation of a monoarthritis in a 60-year-old female may not elicit the possibility of PVNS, resulting in delayed diagnosis.

A surrounding mass extending much beyond the joint margins is quite odd and is the most remarkable feature of the case. A few reports of PVNS with extra-articular extension are described in the literature, particularly in the knee and hip joints[33-38]. Some authors defend that a rapid proliferation of the synovial tissue increases the intra-articular pressure, weakening the joint capsule, which gives way to the synovium[34,36]. Others enhance the possibility of secondary cellular seeding when a diseased joint is surgically approached[35]. In the last case, the synovial cells could simply migrate into the disrupted tissue planes or be displaced to surrounding locations, during the surgical procedure. Although uncommon, distant metastases have also been reported in some cases[39,40]. Our patient had a previous first left MTP surgery, what had probably contributed not only to the extra-articular expansion of a recurrent synovial lesion but also to the local spread of damaged cells into the surrounding tissue.

Other aspects of our case were closer to the typical features of PVNS as reported in text books and journal articles. The condition manifested itself by nonspecific clinical symptoms with an insidious onset and a long-standing course. The average duration of the symptoms before diagnosis has been reported to be between 10 and 26 months (range 2-72 months)[1]. Localized tenderness, pain, locking, giving away, diminished range of motion, palpable mass, swelling, stiffness, snapping or instability are described as the most frequent clinical findings[1,2,4,9,32]. Recurrence after surgery is not unusual, especially in localized PVNS[6].

Routine haematological, biochemical and immunological blood tests were, as usual, inconspicuous[3,8]. When joint effusion is present, the aspirated synovial fluid is typically xanthochromic or serosanguinous[1,2,8,9].

Plain radiographs are generally normal, revealing abnormalities in only 15% of the cases[1,2], which consist of increased density of the synovium, radiolucent cystic defects and bone erosions[1,2,6,10]. Bone erosions are mainly found in hip PVNS and have been attributed to an increase in the intra-articular pressure, due to the hyperplastic synovial growth inside a strong capsule[10]. In pigmented villonodular tenosynovitis surrounding bone erosions are found in about 25% of the cases[7]. Osteopenia in the juxta-articular bone can also be identified[3]. Calcifications are rare and more consistent with synovial sarcoma[8]. Loss of joint space is, generally, a late finding in the course of the disease[10]. So, the absence of any joint or bone abnormalities in our case represents the most common case in PVNS.

We chose not to perform bone scintigraphy, ultrasound or CT scan because they have limited value in this condition[7]. On the contrary, MRI allows the identification of features which are considered highly suggestive of PVNS: low signal on T1 and T2-weighted images (because of hemosiderin deposits), synovial hyperplasia, bone erosions, preservation of bone density and, commonly, the presence of a joint effusion[1,3,9,10]. These signal features correlate with the varying amounts of hemosiderin in the lesion[1-3]. When

hemosiderin deposition is minimal, the characteristic low signal may not be apparent, as in our patient[1,10].

The final diagnosis of PVNS is established on the basis of histological examination of synovial tissue[6]. The findings in our case are typical of PVNS: subsynovial mononuclear histiocytic reaction with occasional multinucleated giant cells, sheets of small ovoid or spindle-shaped cells, macrophage plasma cells and rich vascular plexus, with common occurrence of mitotic features[1,2]. Extracellular hemosiderin-like deposits are another common finding[2].

The optimal therapeutic approach for PVNS is debatable[6]. Simple marginal excision (even arthroscopically) is regarded as the adequate therapy for localized PVNS while total or subtotal synovectomy is usually advised as the therapy of choice in the diffuse type[4,6-8]. Relapses are, generally, treated with resynovectomy[6]. All associated bone lesions should be treated with careful curettage followed by bone graft if necessary[6,10]. Some nonsurgical therapies, such as steroid injections, 90Y synoviorthesis and external beam radiation are reported in the literature as having considerable benefit in selected patients[7]. Our patient presented a recurrent case of PVNS, with a diffuse synovial membrane proliferation, which made total synovectomy the therapy of choice.

The risk of recurrence of both types of PVNS seems to depend on the margins obtained at surgery[6]. The literature reports rates of recurrence ranging from 0% to 48% for localized PVNS and from 0% to 46% for the diffuse type[6]. MRI is a useful tool in controlling the preclinical recurrence of the lesion[2]. At 1-year follow-up our patient was free of symptoms and the left foot MRI excluded local recurrence.

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