

Villonodular synovitis of the lumbar spine: Case report of a rare pathology

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Abstract

Lumbar pigmented villonodular synovitis is rarely localized on the lumbar spine. Awareness of pigmented villonodular synovitis is important despite its rarity because it may mimic several other lesions of this location.

KEYWORDS

lumbar spine, neurosurgery, orthopedics, villonodular synovitis

1 | INTRODUCTION

Pigmented villonodular synovitis (PVNS) is a rare proliferative disorder of an uncertain origin belonging to benign synovial dystrophies, characterized by villous or nodular hyperplasia of the synovium.^{1,2} There are mainly two types of PVNS: the first affects the synovium of a joint diffusely, whereas the second localized form focally involves the synovium about a tendon sheath.^{1,3} The diffuse form is usually monoarticular, affecting large joints such as knees, hips, ankles, shoulders, and elbows.^{1,4} But PVNS rarely involves the axial skeletal system.⁵⁻⁷ Within the spine, lumbar localizations are the less encountered. Thus, their description in the literature is only restricted to some case reports.^{1,7} In this paper, we report a new case of a PVNS of the lumbar spine and try to review different epidemiologic, clinic, radiologic, and therapeutic features related to this pathologic entity.

2 | CASE DESCRIPTION

We report the case of a 57-year-old woman, known to be suffering from diabetes, and operated during the childhood for

an appendicitis. She presented for right sciatica without any defined systematization. These pains began since one year and progressively worsened, with association to intermittent claudication and decrease of the walking range. Nevertheless, the patient did not complain from any sphincteric disorders. On physical examination, the patient had a slight motor deficit of the right lower limb, associated with absent patellar and achilean reflexes, as well as a stiffness of the lumbar paravertebral muscles. Otherwise, there were neither sensitive deficit, nor lumbar skin abnormalities. Spine MRI (Figure 1) showed a right paramedical tumor on the level of L4-L5, which seemed to be intradural and extramedullary. The tumor was compressing the right L4 root as well as the cauda equine, which was congruent to the clinical symptomatology of the patient. This was associated with a grade I L5-S1 listhesis with bilateral foraminal stenosis. The decision was to operate the patient for both removal of the tumor and stabilization of the lumbar spine. The first stage of surgery consisted on an osteosynthesis by pedicular screws ranging from L3 to S1, skipping the right L4 screw because the tumor was situated at that level. After spinolaminectomy, the second stage focused on the removal of the tumor. In fact, the lesion was greenish, extradural, seeming to originate from the L4 vertebral body,

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FIGURE 1 A and B, Sagittal section of a lumbar spine MRI on T2-WI (A) showing a tumor on the level of L4-L5. This was associated with a grade I L5-S1 listhesis with bilateral foraminal stenosis. Axial section of a lumbar spine MRI on T2-WI (B) showing that the tumor is right paramedical, intradural, extramedullary, and compressing the right L4 root as well as the cauda equine

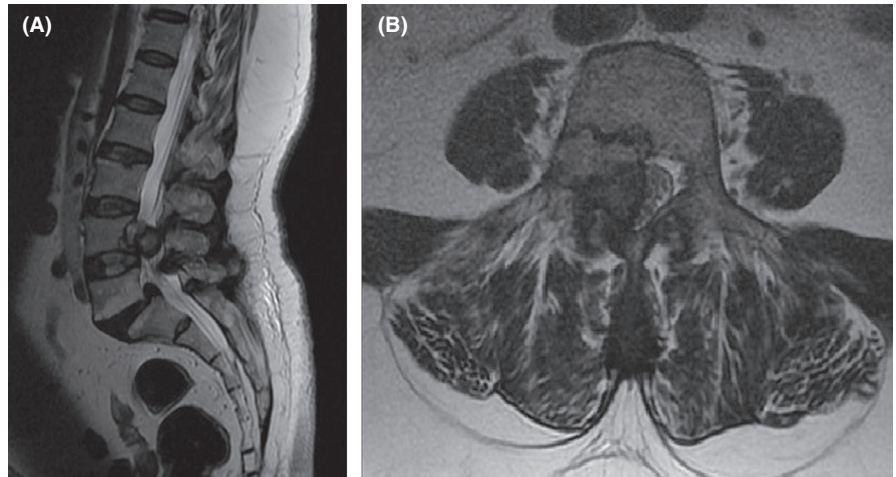


FIGURE 2 A and B, Pathologic examination (A) showing villous appearance of the synovial with elongated fringes (Hematoxylin Eosin $\times 10$). On higher magnification ($\times 40$) (B), the connective tissue seems to contain an inflammatory infiltrate with lymphocytes and macrophages

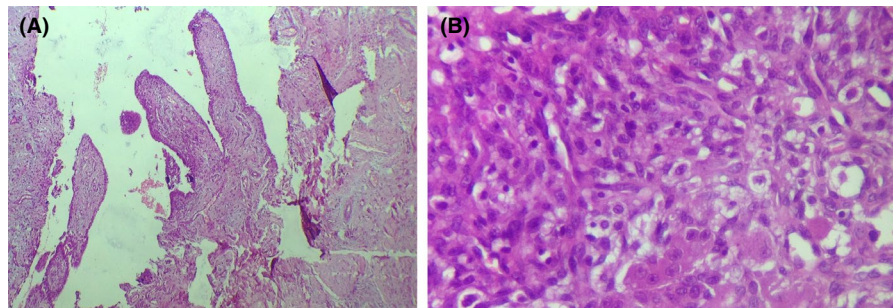
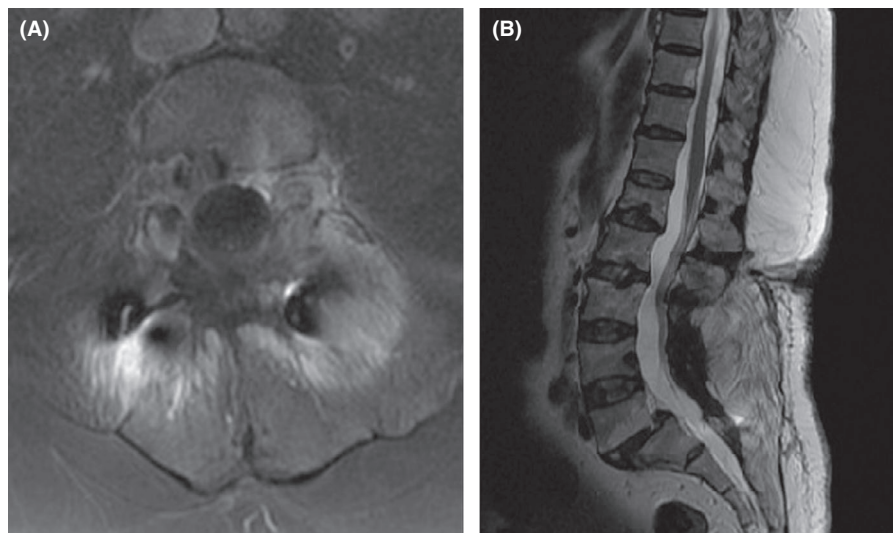


FIGURE 3 A and B, Sagittal (A) section of a lumbar spine MRI on T2-WI and axial (B) section of a lumbar spine MRI on T1-WI with injection of gadolinium showing a small recurrent or residual lesion



extending laterally toward the right pedicle, which was totally destroyed. Underneath the pedicle, the right L4 root was also invaded by the tumor as it seemed to be swollen with yellowish aspect. The facet joint was only partially invaded. The tumor was gradually removed by fragmentation, and the right L4 root was sacrificed. Surgical exeresis was incomplete because of the persistence of some tumor tissue on the posterior wall of the vertebral body, whose access was made hard by the interposition of the cauda equine and bleeding.

Postoperative, the patient reported right sided sciatica, but no major complications were noticed. She was discharged on the 5th day after surgery. Pathologic examination concluded to a pigmented villonodular synovitis (Figure 2). On the follow-up, the patient reported a regress of the lumbosciatic pain, but kept right L5 paresthesia partially improved by gabapentin. A control MRI (Figure 3) was performed 1 year and a half after surgery. It showed a small lesion (recurrence or relic) without any contact with intracanalicular nervous elements.

3 | DISCUSSION

Pigmented villonodular synovitis is an uncommon condition, distinguished by synovial proliferation associated with a hemosiderin deposition in the affected joint.⁷ Calcifications are not featured to this pathology.⁸ PVNS is mainly associated with monoarticular manifestations of lower extremities. In opposite, spinal localizations are very rare.^{5,9} According to Motamedi's¹ study, which is one of the very few who studied spinal PVNS through multiple cases study, as most of the published papers are about case reports: 53% of the spinal PVNS were cervical, 27% thoracic, and 20% lumbar.

The etiology of the PVNS is still under debate. Some authors proposed neoplasms, traumas, inflammation, and metabolic derangement as the *primum movens* for the onset of the derangement.⁹ The high cellularity of the lesion, the tendency for recurrence, and some cases of local malignant behavior support a possible neoplastic process.¹⁰ However, in our case, there was no evidence about any associated tumoral process even after follow-up. The presence of inflammatory cells: histiocytes, macrophages, may suggest an underlying inflammatory process.¹¹ Post-traumatic hemorrhage is also mentioned, but there would be no statistical correlation between traumas and PVNS, as is the case for our patient.^{12,13} In one case, trisomy 7 was documented suggesting that chromosomic aberrations may lead to PVNS, thus the interest to orientate researches toward genetics.¹⁴

The main clinical complaints are mostly represented pain, associated or not to neurologic deficits. Our patient experienced both sciatica and motor deficit. These symptoms produce the compression or even an invasion of the intracanalicular nervous elements, namely the cauda equine or the medulla and roots.^{7,14}

Radiological features concerning PVNS have mainly been reported in detail for PVNS located in the extremities.⁹ On spine located PVNS, Computed tomography usually shows hyperdense enhancing soft-tissue mass that may at first sight raise the diagnosis of disk herniation or bone erosions.¹⁵ Currently, the gold standard for diagnosis of PVNS is magnetic resonance imaging (MRI). It classically shows a mixed signal of the tumor on T2-weighted images, related to hemosiderin deposition.¹⁶ This investigation is also able to reveal the extent of the lesion, mainly any conflicts with nervous structures.¹⁷ Nevertheless, radiologic diagnosis of PVNS is not obvious, and several differential diagnoses may be discussed including primary bone lesions, extradural and mesenchymal neoplasms such as osteoblastomas, schwannomas, large cell lymphomas, fibrohistiocytic tumors and hypertrophic synovitis.^{2,9,16}

When discussing pathological features, giant cell tumors (GCT) can be classified into two forms: localized (GCT

of the tendon sheath, or nodular tenosynovitis) and diffuse (diffuse-type giant cell tumor or PVNS). Localized GCT principally affects the small joints and presents as a solitary slow-growing tumor. PVNS is a more aggressive form affecting both small and large joints.

Localized GCT are commonly slow-growing, painless tumors, fixed to deep structures, and eroding the bone. They always present as a soft-tissue swelling in the hand or foot, adjacent to a small joint. Spinal localizations are extremely rare. On the opposite, PVNS cause often intermittently tender and painful joint. Although considered to be a benign condition, the diffuse form is more aggressive, with a high recurrence rate after surgery of 25%.^{11,14,18}

The aim of the treatment for patients suffering of spinal PVNS is gross total excision of the tumor with functional preservation, and to lower the recurrence rate, which varies between 18% and 50% for all spinal cases.¹⁶⁻¹⁸ Several management options were discussed including surgery, radiotherapy, and radioisotope infusions.^{2,17} Considering the high tendency for local recurrence of PVNS, with very rare cases of metastases, the ideal treatment would be complete operative excision referring to most of the published therapeutic experiences.^{2,7-9,16} Preoperative, several surgical approaches were described, including or not osteosynthesis depending of the extend and the location of the lumbar PVNS. All papers converge toward the evidence of attempting the most complete removal of the tumor.^{2,5,7,10,16} Regarding the high recurrence rates, some authors propose systematic postoperative radiotherapy.^{2,16} Others consider adjuvant treatment just in case of incomplete removal of the mass, as only about 9% of the patients presented tumor recurrence after a total excision.¹⁶ In our experience, the patient had no postoperative radiotherapy despite that he had an incomplete tumor excision. He remained asymptomatic a year after surgery, and control MRI showed no further progression of the mass.

4 | CONCLUSIONS

Pigmented villonodular synovitis involving the spine is rare. Awareness of this lesion is important because it may mimic several other neoplastic and non-neoplastic lesions of this location. Standard treatment for spinal PVNS is surgical gross total removal. There is still no consensus regarding the efficacy of radiotherapy as adjuvant treatment. More case studies of spinal PVNS are needed to clarify treatment following surgery.

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CONFLICT OF INTEREST

Author and co-authors declare having no conflicts of interests.

AUTHOR CONTRIBUTIONS

GG: wrote the article. AB: made the bibliographic research and provided radiologic iconography. AZ: provided pathologic iconography and features. AD: provided radiologic iconography and radiologic interpretation. IZ: corrected the manuscript. MB: corrected the manuscript and made synchronization between different authors.

ETHICAL APPROVAL

We confirm that the manuscript has been read and approved by all named authors and that there are no other persons who satisfied the criteria for authorship but are not listed. We further confirm that the order of authors listed in the manuscript has been approved by all of us. We confirm that we have given due consideration to the protection of intellectual property associated with this work and that there are no impediments to publication, including the timing of publication, with respect to intellectual property. In so doing, we confirm that we have followed the regulations of our institutions concerning intellectual property.

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