Pigmented villonodular synovitis of the thoracic spine: case report and review of the literature

Sinovitis pigmentada vilonodular da coluna torácica: relato de caso e revisão da literatura

Sinovitis vellonodular pigmentada de la columna torácica: informe de un caso y revisión de la literatura

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RESUMO

Pigmented Villonodular Synovitis (PVNS), a lesion of the synovial tissues, is rarely found in the spine. We present a 73-year-old male with increasing lower extremity weakness and paresthesias. MRI scans revealed disc herniation and spinal cord compression at the T11-T12 and T12-L1 levels. Intraoperative exploration revealed an epidural mass originating in the T12 lamina, compressing the spinal cord at T11-T12. Pathologic examination was consistent with pigmented villonodular synovitis.

KEYWORDS: Synovitis, pigmented villonodular; Spinal cord compression; Laminectomy; Spinal fusion; Thoracic vertebrae

ABSTRACT

Sinovitis pigmentada vilonodular (PVNS) é uma lesão do tecido sinovial e raramente é encontrada na coluna vertebral. Apresentamos o caso de um homem de 73 anos de idade com aumento de fraqueza da extremidade inferior e parestesia. O exame de imagem por ressonância magnética revelou hérnia de disco e compressão no nível T11-T12 e T12-L1. A exploração cirúrgica evidenciou massa epidural orginária em T2 e compressão da medula espinhal no nível de T11-T12. O exame patológico foi compatível com sinovitis pigmentada vilonodular .

DESCRITORES: Sinovite pigmentada vilonodular; Compressão da medula espinal; Laminectomia; Fusão vertebral; Vértebras torácicas

RESUMEN

Sinovitis vellonodular pigmentada (PVNS) es una lesión del tejido sinovial y raramente se encuentra en la columna vertebral. Presentamos el caso de un hombre de 73 años de edad que mostró aumento de la flaqueza de la extremidad inferior y parestesias. El examen de imagen por resonancia magnética indicó una hernia de disco y compresión en el nivel de T11-T12 y T12-L1. La exploración quirúrgica evidenció una masa epidural originaria en T2 y compresión de la médula espinal a nivel de T11-T12. El examen patológico fue compatible con sinovitis vellonodular pigmentada.

DESCRIPTORES: Sinovitis pigmentada vellonodular; Compresión de la medula espinal; Laminectomía; Fusión vertebral; Vértebras torácicas

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Recebido: 19/06/2008 Aprovado: 02/10/08

INTRODUCTION

Pigmented villonodular synovitis (PVNS) is a lesion of synovial membranes of several tissues, including tendon sheaths, joints and bursa. First described by Jaffe et al.[1], it is a villous or nodular proliferation of synovium often located in large joints of the appendicular skeleton, most commonly the knee. Although classically benign, it may enlarge to become locally aggressive.

CASE REPORT

A 73-year-old male was seen initially with a history of previous L4- 5 discectomy and L3-L5 fusion in 1973, as well as L2-4 laminectomy in 2006. His chief complaints at presentation were decreased strength, paresthesias, and diminished sensation in his right lower extremity. He had a previous 15 year history of left lower extremity weakness and foot-drop requiring an ankle-foot orthotic. In the past 1-2 years his ability to ambulate had steadily declined secondary to decreased sensation and motor function in both lower extremities. He had regressed to using a walker for long distances.

On physical examination, the patient had 4-/5 strength in his quadriceps and gastrocnemius muscles bilaterally. He had normal sensation to light touch in his bilateral lower extremities. He was normoreflexic. He walked with a wide- based gait with shuffling of his feet. The remainder of his physical examination was unremarkable.

MRI of the lumbar spine with contrast revealed T12-L2 moderate bilateral foraminal stenosis secondary to disc herniation. At the level of T11-T12 there was right paracentral disc herniation with severe right lateral recess stenosis and moderate central spinal stenosis with compression of the exiting spinal nerve root. There was a small cyst extending from the medial right facet joint compressing the dorsal portion of the spinal cord, further e xacerbating the stenosis at T11-T12 (Figure. 1).

Due to the recent decline in clinical picture and significant MRI findings of spinal stenosis, the patient underwent a T11-L3 laminectomy, T11-T12 and T12-L1 diskectomy, and T11-L3 fusion and fixation.

Intraoperatively, the cord was compressed very tightly at T11 and T12 with abnormal-appearing tissue intimately adherent to the dura emanating from the lamina but not visually associated with the facet joints. This soft red-tan epidural mass measured 0.6 x 0.4 x 0.3 cm. During excision, the mass bled briskly. Pathologic analysis of this tissue revealed synovial-type tissue with fibrohistiocytic reaction, multinucleated giant cells, and hemosiderin deposits consistent with pigmente villonodular synovitis (PVNS) (Figure 2).

The patient tolerated the surgery well and noted improved pain and strength in his bilateral lower extremities on postoperative day one. The remainder of his postoperative course was unremarkable.

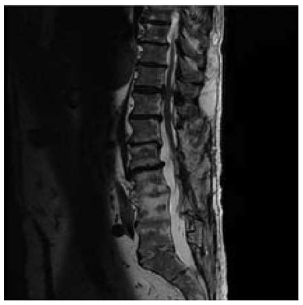

Figure 1
Lateral and axial MRI scans show T11-12 herniated disk and lesion within the lamina

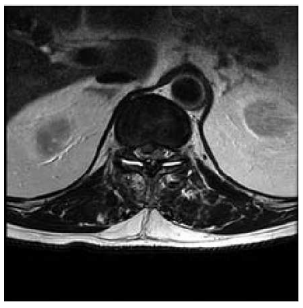

Figure 2
Composite picture showing an epidural pigmented villonodular synovitis in the thoracic spine

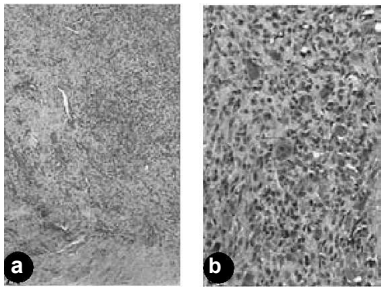

Figure 2 A
(a) Shows a moderately cellular ill defined nodular lesion with focal remote hemorrhage and (b) highlights the fibrohistiocytic changes with multiple multinucleated giant cells

DISCUSSÃO

Pigmented villonodular synovitis is a rare proliferative disorder of synovial tissue with a reported incidence of 1.8 cases per million[2,3]. It is most often located in the appendicular skeleton, though rarely has been found in the axillary skeleton. It frequently causes symptoms of joint pain and swelling as well as catching or stiffness in large joints. The first reported case of its occurrence within the spine was by Kleinman et al. in 1980[4]. There have since been <50 cases reported in the literature to date. Only seven (17%) of these reported cases involved the thoracic spine, making this an extremely rare lesion in this area[5]. Due to its location in the spine, local extension may have potentially debilitating implications. Because of the predilection for expansion and local aggressive behavior, complete resection has become the goal of surgery.

Since first reported, PVNS has been known by a number of different terms, including nodular tenosynovitis, giant cell

synovioma, and fibrous xanthoma of the synovium[6].Most now agree that these terms all represent a single entity with a spectrum of different presentations and anatomical locations. PVNS can be subdivided into two types, one affecting the synovium of a joint surface diffusely and another locally infiltrating the tendon sheath. The diffuse type classically has been found in large joints of the body such as the knee or hip. The localized form has also been referred to as giant cell tumor of the tendon sheath and typically involves the hands or feet. This case represents the diffuse type, involving adjacent vertebrae and extending into the spinal canal. The spinal form of PVNS has been described as arising most often from the synovium of the posterior facet joints. However, in three reported cases, the anterior structures of the spine were the primary location, and it was speculated that the source of these lesions may have been the synovial lining of the accessory joints of the vertebral column[7]. Finn et al. notes that another possible explanation of this finding is anterior growth from the posterior elements with a microscopic or overlooked communication[8]. In our case, though the mass was localized to the posterolateral aspect of the spinal canal, the tumor was not communicating directly with the facet synovium—making its origin somewhat obscure.

Much of the ambiguity surrounding PVNS stems from its unknown etiology. The list of potential sources include trauma, neoplasia, hyperplasia, metabolic, and hemorrhagic[6,9,10]. Of these etiologies, the two most oft discussed are neoplastic and traumatic. Several authors have suggested a traumatic link, making note of a history of recent trauma in patients[11-15]. Like the current case, this has not been a universal finding and no data supports this assertion, suggesting that other factors may be at play. In addition, experimental attempts to create PVNS lesions in animal models have failed to reproduce this lesion[16]. Recent reports have highlighted the clonal nature of these lesions in cytogenetic studies[17,18]. These studies, coupled with reports of malignant PVNS[19-21], make a neoplastic etiology equally likely. These studies should form the basis of future studies into the possible malignant conversion of benign lesions and the biochemical nature of the spectrum of these lesions.

The pathological evaluation of PVNS demonstrates that it is a villous or nodular proliferation of synovium. Investigation of these lesions underlines its similarity to lesions found in the appendicular skeleton[22]. The lesion is characteristically composed of aggregated round to polygonal mononuclear epithelioid cells with multinucleated giant cells, xanthoma cells, siderophages, lymphocytes, fibrous tissue and hemosiderin deposits in varying frequencies. Pathology of our specimen demonstrated many of these features, and was notable for dense hemosiderin deposits. Not surprisingly, during the process of excision, higher than expected blood loss occurred. Caution should be exercised in excising these masses as bleeding may be brisk. Adding weight to the theory of neoplastic origin for PVNS, mitoses are not infrequently observed. There is, however, not a biochemical marker or feature which alone classifies the lesion as PVNS and the aforementioned characteristics may manifest themselves to different extents. Examination of epidemiologic data gathered on PVNS of the spine demonstrates a wide spectrum of presentation. The most common reported presentation is pain. However, radicular symptoms, progressive weakness, parasthesias, or incidental findings are not uncommon. The most frequent site of presentation is the cervical spine (52%), followed by lumbar (29%) and, finally, thoracic (17%)[5].All ages are equally affected and the mean age of presentation is 38[5]. Our patient is unusual in that he is 73 years old. Though earlier reports suggested that PVNS of the spine is equally distributed among the sexes[23-25], Motamedi, et al. recently reviewed 15 cases and found a predilection for women (64%)[26].

Imaging of spinal PVNS demonstrates osseous erosion of bone with sclerotic margins[27]. Plain radiographs are less diagnostic (45% of lesions) than CT scans (67%) or MRI (67%) for identifying the facet joints as the site of origin of these lesions[24,26]. Motamedi's study emphasized that patient age, a solitary noncystic lesion centered in the posterior elements, lack of mineralization and low-to-intermediate signal intensity on all MRI images suggest the diagnosis[26]. The differential diagnosis of these lesions include neoplasms of the posterior osseous elements, chordoma, chondrosarcoma, osteoblastoma, aneurysmal bone cyst, metastases, multiple myeloma, lymphoma and giant cell tumor[28]. By examination of specific imaging features, the differential can be significantly narrowed. Chondrosarcoma, chordoma and osteoblastoma are usually calcified on CT scan. An aneurysmal bone cyst is cystic in nature, with high signal intensity on T2-weighted MRI imaging. Multiple myeloma and lymphoma are commonly found in older individuals and should be included in the differential diagnosis for our patient. Giant cell tumor usually affects the anterior structures and is associated with the vertebral body. Chordoma and chondrosarcoma have a high signal intensity on T2-weighted MRI. Taking into account this differential in the face of a positive finding on MRI, one can determine the relative likelihoods preoperatively.

The treatment of choice in symptomatic patients is gross-total resection. Other management options include radiation therapy and radioisotope infusion[28], though the role of these modalities has been ill-defined. Giannini et al. calculated a recurrence rate of 18% with gross-total resection of spine PVNS[29]. This figure is similar to that calculated for lesions resected in the appendicular skeleton[25]. Furlong et al. reported that recurrence after gross-total resection may be correlated with diffuse growth pattern, diminished giant cell component, epidural involvement and soft-tissue spread beyond the bone, and patient age less than 30 years5. Regardless of risk factors for recurrence, the consensus remains that close follow-up with frequent imaging is necessary given its rare reported incidence and lack of data defining the natural history of these lesions.

Acknowledgement

The authors thank Karen K. Anderson, B.Sc., for her editorial assistance in preparing this manuscript.

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