Pigmented Villonodular Synovitis : Dorsum Foot

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MJAFI 2007; 63 : 294-296

Key Words: Pigmented Villonodular Synovitis; Hemosiderin macrophages

Introduction

Pigmented villonodular synovitis (PVNS) is a relatively uncommon neoplastic disorder of the synovium. The disorder results in increased proliferation of synovium causing villous or nodular changes of synovial lined joints, bursae and tendon sheaths. The initial clinical symptoms are indistinguishable from common pathologies (i.e. tenosynovitis, osteochondral defects, or trigonum injury and tendon tears). Surgical treatment includes synovectomy and tenosynovectomy. PVNS is locally aggressive and tends to recur if incompletely excised [1].

Case Report

A 39 year old serving soldier presented with an insidious gradual progressing painless soft tissue swelling over the dorsum of right forefoot of nine years duration. He underwent the excision six years back in a service hospital and diagnosed as a case of giant cell lesion as per the discharge slip. The swelling recurred within six months of initial surgery, increased progressively over the last six years. Clinical examination revealed a 6x4 cm swelling over the dorsum of foot, overlying first to fourth metatarsals with extension into the proximal phalanx of the second toe (Fig. 1). The swelling was nontender noncompressible, soft in consistency. Movements of foot were within normal limits. There was no inguinal lymphadenopathy. Radiograph of foot did not reveal any bony abnormality. Fine needle aspiration cytology was suggestive of a benign giant cell lesion (Fig. 2).

The swelling was approached under spinal anaesthesia through longitudinal incision in line with second metatarsal. There was ventral extension of growth between the metatarsals into the proximal phalanx of the second toe. The lesion was closely applied to the extensor tendons (Fig.3). Total excision was done preserving the tendons, vessels and nerves to second toe. Wound was closed over a small suction drain after ensuring adequate haemostasis. Post-operative recovery was uneventful. He had been referred for radiotherapy to a referral hospital. However the patient was lost to follow-up.

The gross pathological findings included a specimen measuring 5x3x2 cm with a soft consistency and grey colour with bosselated surface. The cut surface showed a variegated appearance with alternate dark and grey white areas. On microscopic examination sections of the growth showed an encapsulated tumour made up of predominantly closely packed medium sized polyhedral cells with variable admixture of multinucleated giant cells and hemosiderin laden macrophages. The mitotic rate was not high. There were focal areas of hyalinization and collagenisation seen. Papillary projections were made up of foamy macrophages.

Discussion

Pigmented villonodular synovitis (PVNS) is a proliferative disorder of the synovial lining of the joints of all age groups, commonly seen in young adults and females. Knee, ankle, shoulder and small joints of hand are commonly involved. Bilateral disease is exceptional. PVNS usually presents with joint or tendon sheath swelling and the lesion may occasionally penetrate the underlying bone. The history of previous injury is known in few patients. The diagnosis is based on physical examination, standard laboratory tests, radiography, ultrasonographic /magnetic resonance examination and histopathological examination of synovium obtained by open or arthroscopic biopsy.

On histology cellular nodules of histiocytes like cells

Fig. 1 : Photograph showing swelling dorsum of foot.

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Fig. 2 : FNAC showing multinucleated giant cell

Fig. 3 : Intraoperative view of the growth showing bosselated appearance extending diffusely into soft tissues

with scattered giant cells, siderophages and foam cells, probably related to haemorrhage are seen. Mitoses are rarely seen. Collagen and chronic inflammation suggest an older lesion. In PVNS the synovium usually forms papillary projections. Ultrastructural and immunohistochemistry studies have shown that the tumour cells have features of synovial cells alternating with fibroblasts and histiocytes.

PVNS has been classified into three types: localised nodules, diffuse PVNS involving the entire synovial membrane and a combination of a diffuse involvement of the synovial membrane and an extra-articular presentation. Articular disease localization is 2.5 times more frequent as compared to the non-articular. PVNS is almost always a single lesion. The diffuse-variant tenosynovial giant cell tumour is rare, although it shares histological features with the exclusively intra articular pigmented villonodular synovitis and local tenosynovial giant cell tumour, its differs in being aggressive locally. A comparative study has been done to assess the quantitative evaluation of cell populations involved in the proliferative and inflammatory compartment in both localized and diffuse PVNS. It was found that the number of CD3+ and CD20+ cells was significantly

Fig. 4 : Tumour showing polyhedral cells, giant cells and haemosiderin laden macrophages (H&E stain-400X).

higher in localized PVNS while the number of CD57+ natural killer (NK) cells was significantly higher in diffuse PVNS [2].

Approximately 75% of all biopsy proved soft tissue masses of the foot and ankle are benign tumours or non tumoural lesions representing a variety of histologic types which are easily diagnosed. PVNS can be mistaken for common benign tumours of the foot (e.g. giant cell lesion of tendon sheath, fibromatosis, cavernous haemangioma) and in nonneoplastic soft tissue lesions such as Morton neuroma, ganglion cyst and plantar fasciitis. Another condition called fibroma of tendon sheath (FTS) has been traditionally considered to be two points in a single neoplastic continuum. Both tumours have a lobular appearance. PVNS is more cellular than FTS, it contains conspicuous numbers of osteoclast-like cells and the stroma is not extensively hyalinised. In contrast, FTS is matrix-rich, often with extensive stromal sclerosis and contains giant cells rarely. Immunophenotyping of PVNS shows that both the spindle cell and giant cell components are positive for vimentin, leukocyte common antigen (LCA), CD68, HAM56, AACT and MAC387, suggesting monocyte-macrophage-like features. On the other hand, FTS shows focal staining with HAM56, staining for vimentin and SMA is uniformly intense and diffuse. Sometimes extensively recurrent lesions have been misdiagnosed as fibrosarcoma and synovial sarcoma [3].

Radiologically 50 % of cases might show bone erosion, that is thought to be mediated by matrix metalloproteinases (MMPs). Expression of MMPs in some tumours appears to be stimulated through local production of cytokines and several specific cytokines (tumour necrosis factor(TNF) alpha, interleukin (IL) -1, and IL-6) play an important role in the stimulation of osteoclastic bone resorption. These can be tested by in

situ hybridization for TNF alpha and IL-6 mRNA [4].

The MR imaging appearance of PVNS consists of multiple synovial lesions with low or intermediate signal intensity on T1 weighted images and low signal intensity on T2 weighted and gradient-echo images. PVNS usually have areas of low signal intensity on both T1 and T2 weighted images due to the paramagnetic effect of haemosiderin [5].

Malignant PVNS is a rare lesion whose existence may be debatable. Important histologic features of malignancy are nodular, solid infiltrative pattern of the lesion; large, plump, round or oval cells with deep eosinophilic cytoplasm and indistinct borders; large nuclei with prominent nucleoli and necrotic areas. Atypical mitoses are occasionally seen. It is associated with trisomy 7 and 5 as well as clonal rearrangements involving chromosomes 1, 3, and 15. The malignant nature of this lesion, the histologic architecture similar to that of PVNS and the fibrohistiocytic appearance of the cells suggest that malignant PVNS is an entity [6].

FNAC smears are cellular containing several multinucleate osteoclast-type giant cells and binucleate cells. The non-giant cell population is dispersed principally as single cells that have a cytological appearance mimicking histiocytes and osteoblasts. Anisonucleosis is minimal and nuclear pleomorphism is distinctly absent in both single and multinucleate cells. Mitotic figures are infrequent. Few workers have found the presence of intranuclear cytoplasmic inclusions in the fine needle aspiration smears [7].

Surgical methods such as total or partial synovectomy are applied in the therapy. It is clear that early surgical therapy is the only recommended curative intervention. The decision regarding the surgical approach, arthroscopic versus open, depends on the form of PVNS, the extent of the disease and secondary changes of the joint. Adjuvant radiotherapy in the form of intra articular injections of yttrium 90 (90Y) has been used to control the disease [8]. After finding marked TNF-alpha expression in arthroscopic synovial tissue samples, treatment with an anti-TNF, a monoclonal antibody (infliximab) at a dosage of 5 mg/kg was initiated.

Immunohistologic analysis at follow up identified a marked reduction in macrophage numbers and TNF-alpha expression in the synovium [9].

PVNS should be considered in athletically active patients with persistent pain and swelling with bone erosions on plain radiographs. In the disease, limited to the ankle joint, arthroscopic synovectomy should be the therapy of choice. In advanced diffuse form, total synovectomy and adjuvant radiotherapy forms the treatment of choice.

Conflicts of Interest

None identified

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