Localised pigmented villonodular synovitis of the knee

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Abstract

Localised pigmented villonodular synovitis (PVNS) of the knee is a rare, idiopathic condition presenting with symptoms that can be confused with various other intra-articular pathologies. The condition is usually monoarticular, the knee being most commonly affected. If totally excised, complete cure can be achieved and recurrence is very rarely reported. In this report, we present a patient with PVNS of the knee who underwent arthroscopic treatment.

At short-term follow-up, he had complete recovery of symptoms with no apparent recurrence.

Key words: Knee, Pigmented villonodular synovitis, Localised form

Introduction

Although there has been a great deal of speculation regarding the etiology of pigmented villonodular synovitis (PVNS), the cause still remains unknown. It is apparent that the lesion is not a neoplasm and probably represents an inflammatory process. The forms of PVNS are classified as follows: localised, diffuse and mixed. It is a rare disorder, with an estimated annual incidence of 1.8 cases per million population, only one quarter being of the localised form.

Case Report

A 23-year-old male presented with pain in the right knee joint for one year with swelling and intermittent locking of three months duration. There was no history of fever, trauma, weight loss or similar swellings elsewhere. On physical examination, he had a diffuse swelling of 4 x 3 cm over the superomedial aspect of the right knee. Colour and texture of the overlying skin were normal. On palpation there was an ill-defined tender irregular swelling of firm consistency. There was no synovial thickening. It was not fixed to the overlying skin and reduced in size on tensing the quadriceps. The range of movements of the joint was full and free. X-ray and MRI of the knee are shown in (Figures. 1, 2) An arthroscopic examination was performed. A yellowish-pink coloured nodule embedded in the synovium posteromedial to the infrapatellar fat pad was detected and was totally excised. (Figure. 3, 4) Gross examination of the resected specimen revealed a polypoid 3.5 x 2.0 x 1.0 cm firm gray soft tissue mass with small brown and yellow areas. Microscopic examination showed a fibrocollagenous stroma containing sheets of small ovoid cells with indistinct cell margins interspersed with several multinucleated giant cells, cleft like spaces and focal aggregates of...
The nomenclature used for PVNS in early reports in the literature is often confusing, terms such as xanthoma or giant-cell tumor, myeloxanthoma, villous arthritis, benign synovioma being used. Jaffe et al., introduced the term pigmented villonodular synovitis. Fisk attributed the changes of PVNS to repeated minor trauma of synovial fringes, with consequent hydrarthrosis. Young and Hudeck produced changes that they regarded as similar to those of PVNS by repeated injection of blood into the knees of dogs. It has been suggested, however, that these changes are not really comparable with those of PVNS, but more closely resemble those seen in haemophilia.

PVNS can be classified into 3 forms - localised, diffuse, and mixed (which represents a transition between the localised and diffuse forms). The disease is typically a monoarticular arthritis of the knee joint that affects adults in the third or fourth decades of life. The most common location of PVNS in the knee is the meniscocapsular junction. Other sites occurring with notable frequency are the intercondylar notch, tibial eminence, and peripatellar areas. Rarely, PVNS may be localised in the

Discussion

The nomenclature used for PVNS in early reports in the

Haemosiderophages. The patient is totally free of symptoms 18 months after surgery and there has been no evidence of recurrence.

Figure 2a, 2b. MRI showing presence of intra-articular nodular mass of low signal intensity on T1, T2-weighted & proton-density-weighted images.

Figure 3. Arthroscopic excision of the tumour.

Figure 4. Photograph showing the resected specimen.

Figure 5. Histopathology: Flat synovial cells, polygonal mononuclear cells, giant cells, lipid and haemosiderin deposits are seen.
posterior compartment of the knee\textsuperscript{15} or in the patellar fat pad.\textsuperscript{16} It is a rare disorder, with an estimated annual incidence of 1.8 cases per million populations, only one quarter being of the localised variety.\textsuperscript{3,4}

Initial symptoms are generally mild or moderate episodes of knee pain and swelling. The period between the onset of symptoms and diagnosis may be quite long. In the localised form, a mass is usually palpable in the knee. This mass causes restriction of motion and locking or ‘popping’ sensations in the knee.\textsuperscript{17-19} Standard radiographic findings are rare and subtle in PVNS. Nodular soft-tissue tumefaction without any calcification is the main radiologic finding.\textsuperscript{20} Today, MRI is an excellent clinical tool for the evaluation of intra-articular tumours of the knee joint. In the case of PVNS, it is an effective means for the detection and definition of the size, position and extent of this discrete, nodular lesion.\textsuperscript{21}

The common pathologic features of PVNS are villous or nodular proliferation of synovial tissue and pigmentation caused by the presence of hemosiderin and lipid. Depending on the presence of intraneural hemorrhage or lipid accumulation, the degree of pigmentation ranges from a barely detectable yellow colour to dark brown. On microscopic examination, there is synovial cell proliferation and subsynovial infiltration by histiocytes, fibroblasts and hemosiderin or lipid-laden macrophages (foamy cells). Although occasional mitotic figures may be present, abnormal mitoses characteristic of malignancy is never seen.\textsuperscript{22} The main steps of treatment are arthroscopic extirpation for the localised form and an additional synovectomy for the mixed form.\textsuperscript{2} The best results are obtained in cases of localised PVNS with almost no recurrence. The differential diagnosis includes loose bodies, synovial sarcoma, rheumatoid arthritis, osteoarthritis, chondromalacia patellae and meniscopathies.\textsuperscript{21}

Our patient did not have a previous trauma, which is contradictory to Fisk’s explanation of traumatic pathogenesis.\textsuperscript{2} The length of time our patient experienced symptoms before seeking help is one year thus explaining the mild symptomatology of this disorder. He had a palpable mass and, MRI was performed to evaluate it. In the localised form of PVNS, patients initially have nonspecific complaints. In a patient with a palpable mass in the knee joint or in a patient with intractable pain and/or synovitis, PVNS has to be considered in the differential diagnosis.

There are several reasons to prefer arthroscopic surgery to arthrotomy in cases of suspected PVNS. Arthroscopy is less invasive than classical arthroscopy, it allows a more accurate diagnosis, and facilitates the discovery of small localised forms of PVNS. It also allows for the specification of the size and the site of a localised mass. Arthroscopy permits the evaluation and biopsy examination from the remaining synovium in order to classify the form of PVNS. Synovectomy might be recommended for mixed and diffuse forms of PVNS.

Numerous papers report complete cure with total excision in the localised form of PVNS,\textsuperscript{1,17,23} except for Panagiotapoulos et al.,\textsuperscript{25} who presented a case with local recurrence 17 years after excision.

We believe that arthroscopy has assumed a dominant role in the diagnosis, treatment, and follow-up in patients with synovial disease of knee.
Case Report

Jiss et al.: Localised pigmented villonodular synovitis of the knee


Source of funding: Nil; Conflict of interest: Nil

Cite this article as: