CASE REPORT

Exceptionally aggressive pigmented villonodular synovitis of the hip unresponsive to radiotherapy

Pigmented villonodular synovitis (PVNS) is a rare benign neoplastic proliferation of synovial tissue which is typically localised and usually responds well to surgery and/or radiotherapy. We present a case of unusually aggressive of PVNS of the hip in a 73-year-old woman.

Pigmented villonodular synovitis (PVNS) commonly affects the large joints of the long bones. The hip is affected in approximately 15% of cases.1 Although it is classified as a benign condition, it can be locally aggressive and highly debilitating owing to pain.2,3 Treatment by surgical synovectomy, especially when the condition affects the knee, and/or external beam radiotherapy, can give good results.2,4

Case report

A 73-year-old Caucasian woman presented with a spontaneous fracture of the left femoral neck which was thought to be pathological (Fig. 1). She had no relevant previous history and underwent hemiarthroplasty. The femoral head and attached synovial tissues were sent for histology, which revealed PVNS causing destruction of bone with no evidence of malignancy (Fig. 2). A bone scan revealed no evidence of bony metastases. No other treatment was given at this stage. She presented again four months later with dislocation of the hemiarthroplasty, which was treated by the insertion of a trabecular metal shell with a modular constrained acetabular liner (Zimmer Inc., Warsaw, Indiana). A further biopsy was taken at operation and the histology again revealed PVNS. She complained of continuing discomfort and was treated with radiotherapy (35 Gy in 15 fractions) two months post-operatively. Unfortunately, this did not control either progression of the disease or her pain, with continuing destruction of the proximal femur and pelvis (Fig. 3). The option of excision arthroplasty was discussed, but the patient declined.

Because of the progressive deterioration of the hip, she was referred to the sarcoma unit at the Royal Orthopaedic Hospital, Birmingham. The diagnosis of PVNS was again confirmed, with the advice that the advanced state of bony destruction precluded further reconstructive surgery.

A whole-body CT scan was arranged to exclude an unrecognised malignant process elsewhere in the body. This revealed lung lesions suspicious for secondary deposits, but with no evidence of deposits elsewhere or a primary tumour. A further open biopsy of the hip was therefore undertaken, which again confirmed the presence of PVNS of an unusually aggressive nature with no evidence of malignancy. Separate microbiology cultures taken from the hip excluded tuberculosis.

A course of imatinib, a monoclonal antibody typically used in the management of chronic myeloid leukaemia, was ineffective in controlling the disease and its symptoms, and...
she was referred for palliative care; including nursing-home care and high-dose opiate analgesia. Further radiological imaging demonstrated extensive bony destruction (Fig. 4).

The patient died as a result of her disease 42 months after diagnosis.

**Discussion**

We report this case as an example of aggressive, locally invasive PVNS. The radiograph at presentation showing involvement of the proximal metaphysis was unusual for PVNS, which typically presents with radiological evidence of lytic changes contained within the joint capsule. Our initial diagnoses of malignancy or infection were excluded by three separate and independently analysed tissue biopsies, coupled with isotope bone scans and microbial cultures.

PVNS is a condition that typically affects young and middle-aged adults, the majority of cases affecting the knee.\(^{5,4,5}\) Although occasionally it is locally aggressive, the capacity for local invasion is usually limited. Progression-free survival after initial surgery has been reported in 65% of cases at 25 years.\(^6\) Radiotherapy, either independently or coupled with surgery, has been reported to achieve control in > 95% of cases.\(^ {3,5}\) Imatinib has been reported as being effective for patients with relapsing PVNS.\(^7\)

To our knowledge, there are no reported cases of patients with histologically proven PVNS who have died or suffered major complications, such as death or amputation as a result of it. We advocate awareness of the potential for atypical behaviour by PVNS in the hip, and consider this case to represent an example of a locally malignant form of this disease.

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**References**


