

Case Report

Destructive Synovial Hemangioma of the Hip Resembling Pigmented Villonodular Synovitis

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Abstract: We report a case of synovial hemangioma of the hip causing joint destruction severe enough to warrant arthroplasty in a 32-year-old man who presented with a 2-year history of hip pain and worsening limp. Plain radiographs demonstrated advanced degenerative changes of the hip articular surface. Magnetic resonance imaging revealed changes consistent with pigmented villonodular synovitis throughout the joint and erosion of the acetabular bone. A total hip arthroplasty was performed, and histopathologic examination of the resected tissue demonstrated synovial hemangioma. This case, unique in the literature, demonstrates the destructive capacity and diagnostic pitfalls of synovial hemangioma. **Keywords:** hemangioma, pigmented villonodular synovitis, magnetic resonance imaging, hip arthroplasty.

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Hemangiomas are common lesions, accounting for 7% of all soft tissue tumors [1]. However, they are uncommon in synovial tissue (a thorough literature review has identified only 204 cases). Synovial hemangioma most commonly occurs in the knee [2], less frequently in the elbow [3], ankle, wrist [4], and temporomandibular joints [5]. There is only one published case of synovial hemangioma occurring in the hip joint [6].

Case Report

A 32-year-old man was referred to our department with a 2-year history of variable pain from his left hip and a progressing limp. The pain was felt in the groin, buttock, and laterally over the hip, radiated to the thigh and was made worse by activity, but he did not experience night pain. Pain relief was achieved with nonsteroidal anti-inflammatory drugs. Occasionally, the

pain prevented him reaching his foot, and he had given up his routine activities that included swimming and cycling for 6 weeks before presentation.

His family history was unremarkable, and his medical history included Osgood-Schlatters disease at age 14 and a microdisectomy at 26 years of age. He had no history of trauma to the hip.

Clinical Examination

On examination, he was a slim, 6' 4" male who mobilized well. His left hip had 50° to 130° flexion, 10° abduction, 5° adduction, and minimal rotation. Attempted rotation of the hip when in flexion reproduced the pain. His right hip showed a normal range of motion.

Imaging

Conventional plain films showed cysts and an intra-articular soft tissue mass. T1/T2 magnetic resonance imaging (MRI) of the left hip showed changes consistent with extensive pigmented villonodular synovitis (PVNS) throughout the hip joint and erosion of the superior and medial acetabular bone. There was evidence of advanced osteoarthritic change of the articular surface of the hip and large cystic areas in the femoral neck and the metadiaphysis of the femoral shaft leading to a provisional diagnosis of cystic fibrous dysplasia (Fig. 1).

Based on the clinical and radiologic findings, a hip arthroplasty with removal of the suspected PVNS and curettage of the fibrous dysplasia in the femur was planned.

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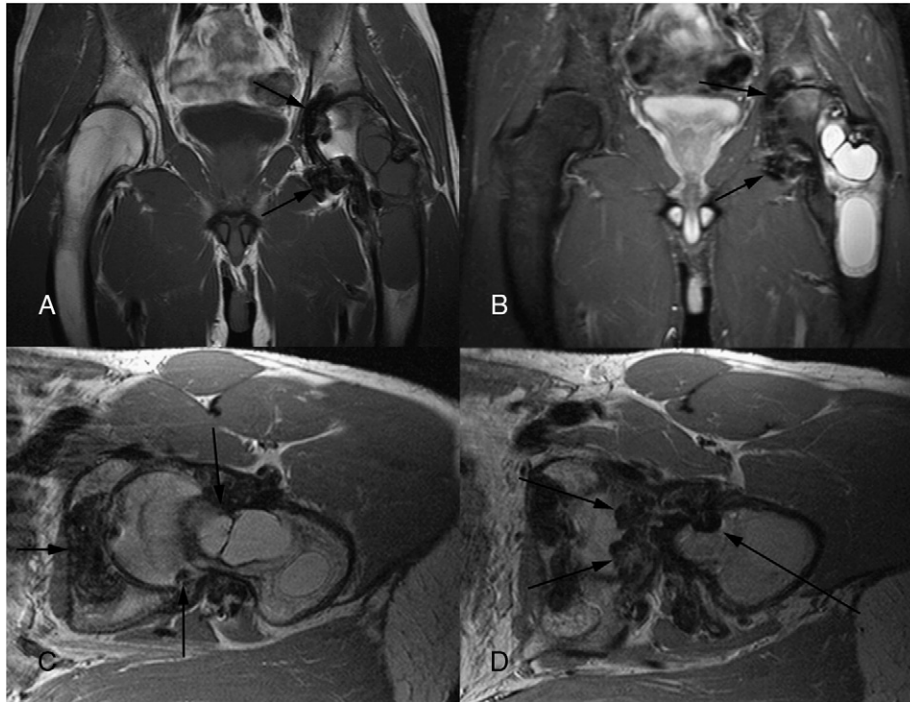


Fig. 1. Magnetic resonance imaging of left hip: coronal T1 (A) (TR, 551 ms; TE, 20 ms) and short TI inversion recovery (STIR) (B) (TR, 5000 ms; TE, 19 ms; TI, 150 ms) MR images of the pelvis showing low-signal intraarticular soft tissue eroding the acetabulum and medial femoral neck (arrows). Axial oblique intermediate (proton-density) weighted MR images of the left hip (TR, 4480 ms; TE, 14 ms). At midacetabular level (C), low-signal tissue can be seen within the joint, deeply eroding the acetabular fossa, the medial wall of the acetabulum, and femoral head/neck (arrows); at the inferior aspect of the joint (D), there is further erosion of the acetabulum and inferomedial femoral neck (arrows). Rounded areas of fluid signal intensity extending from the femoral neck into the femoral shaft are visible on all images, consistent with cysts. TR indicates repetition time; TE, echo time; TI, inversion time.

Surgical Procedure

A large amount of intraarticular synovial tissue was resected, together with the femoral head. There was extensive erosion of the acetabular floor. The proximal femur was curetted and sent together with the synovium and the femoral head for histologic analysis. A ceramic-on-ceramic uncemented total hip arthroplasty was used.

Histologic Evaluation

On gross inspection, the synovial tissue ($8 \times 7 \times 3$ cm) removed was hemorrhagic and tan colored (Fig. 2A). The femoral head showed a severely roughened articular cartilage, and the cut surface revealed an intramedullary cyst (2.5×2 cm) and numerous subchondral fibrous areas (Fig. 2B). The tissue removed from the femoral canal ($8 \times 6 \times 4$ cm) was soft, glistening membranous fragments that were tan colored with areas of hemorrhage ($8 \times 6 \times 4$ cm).

Microscopically, the synovium was diffusely hyperplastic with a papillary architecture. There were numerous randomly placed variably sized blood-filled vessels lined by flat endothelium with areas of extensive perivascular hyalinization (Fig. 2C). In addition, there was extensive deposition of free and macrophage-ingested hemosiderin pigment. These features are characteristic of a diffuse hemosidic synovitis secondary to synovial hemangioma.

Microscopy showed deep fissuring and loss of the articular cartilage and subchondral and intramedullary cyst formation (Fig. 2D). This confirmed the gross appearance of severe degenerative joint disease of the femoral head.

Fragments removed from the medullary cavity of the femur represented fibrofatty tissue without evidence of fibrous dysplasia or malignancy (Fig. 2E). The extent of bony destruction is greater than that typically seen with degenerative joint disease, and whether the extensive cavitation seen was due to the synovial hemangioma is not clear. Final postoperative radiographs are presented in Fig. 3.

Discussion

Hemangioma is most commonly reported in children and young adults and has a slight female predilection. It occurs ubiquitously, often as a superficial lesion not only in the head and neck region but also in the heart, bone, liver, and muscle [7]. Synovial hemangiomas may be classified as diffuse or pedunculated, and as synovial, juxtaarticular (not truly involving the synovial membrane), or intermediate. Histologically, hemangioma is classified as capillary, cavernous, arteriovenous, or venous depending on the predominant vessel type present. Cavernous (50%) and capillary (25%) are the most

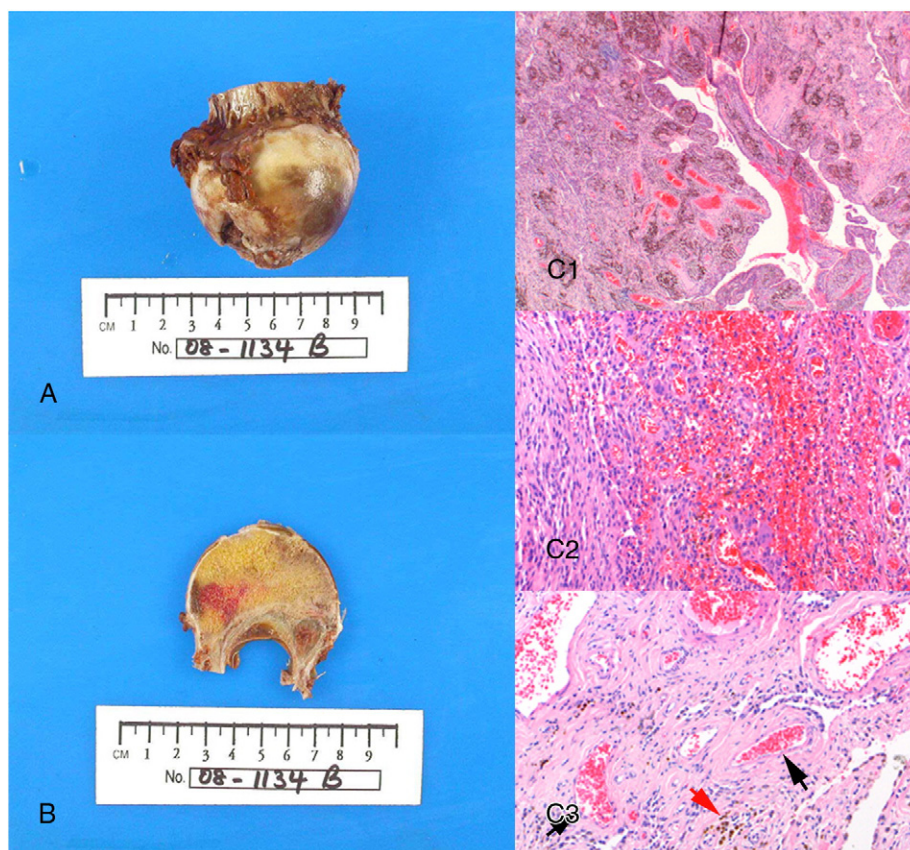


Fig. 2. Gross and histologic appearance of resected specimens. (A) The femoral head together with hemorrhagic tan-colored synovial tissue. (B) Gross image of the cut surface of the femoral head, showing severely roughened articular cartilage. There is an intramedullary cyst and numerous subchondral fibrous areas. (C1-C3) Histopathologic examination of synovial tissue. (C1 and C2) The synovium appears diffusely hyperplastic with a papillary architecture. (C3, black arrows) There are numerous randomly placed variably sized blood-filled vessels lined by flat endothelium with areas of extensive perivascular hyalinization. (C3, red arrow) There was also extensive deposition of free and macrophage-ingested hemosiderin pigment. These features are characteristic of a diffuse hemosidic synovitis secondary to synovial hemangioma.

common forms [8]. Although the etiology of synovial hemangioma is not clear, association with trauma [8] has been suggested. However, it is not certain if it is a neoplastic process or a developmental abnormality.

Synovial hemangioma is difficult to diagnose on clinical examination or with conventional radiography.

In the knee, the lesion often manifests as a palpable swelling and joint pain, but because of the nonspecific symptoms and failure of plain radiographs to show abnormality, the diagnosis is often delayed for many years [4,6,8,9]. During this time, physiotherapy and conservative treatments are often attempted but without

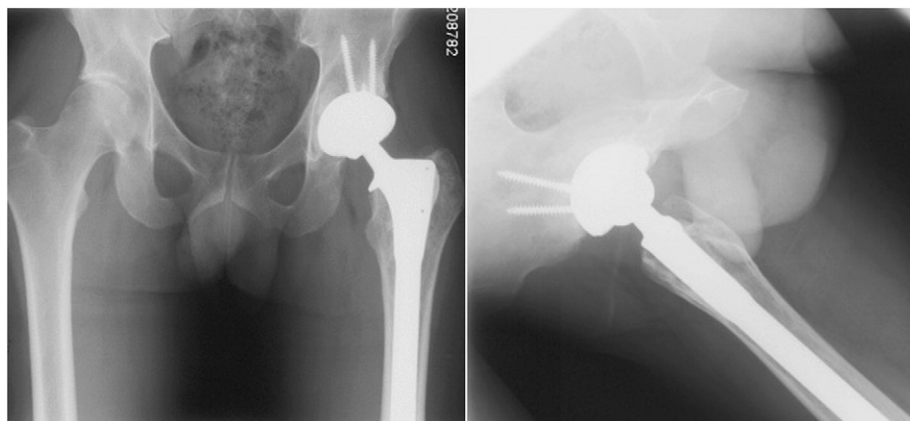


Fig. 3. Final follow-up radiographs (taken 12 months postoperatively).

success [10]. Resection is the treatment of choice (arthroscopic/open). Pedunculated lesions are more easily removed, so arthroscopic surgery is recommended. Diffuse lesions require an open approach to ensure complete removal. Recurrence rates have been reported to be higher in diffuse lesions [2]. Embolization treatment of synovial hemangioma has been reported very rarely [11], and we can find no information on its use before surgical removal to reduce recurrence rates. However, given complete removal of the tumor tissue, it is thought that recurrence ought not to occur [12]. In this patient, a total hip arthroplasty rather than a surface replacement was favored despite his young age due to the level of bone and cartilage destruction seen.

Before widespread availability of MRI, accurate preoperative diagnosis of synovial hemangioma was rarely possible [2,4,13]. However, as MRI often exhibits a characteristic appearance of synovial hemangioma, preoperative diagnosis is now often possible. T2-weighted MRI reveals a high signal intensity (SI) lobulated lesion with low SI linear structures running through it. The high SI regions are likely to represent areas of blood pooling and the lower SI structures fibrous septa [4,13]. T1-weighted images demonstrate a lesion that is often slightly hyperintense but occasionally isointense to skeletal muscle and much less intense than the surrounding fat [13,14]. There is often evidence of hemangioma with identical imaging characteristics in the adjacent extrasynovial/pararticular tissues. Contrast enhancement with gadolinium can result in increased signal intensity of the synovium, indicating synovitis

[14]. Associated effusions are not common. However, even with the resolution provided by MRI small lesions may not be seen, in such cases, arthroscopy can aid diagnosis [6]. Dynamic contrast-enhanced MR angiography, although not more useful diagnostically than conventional MRI, allows identification of high flow areas, helping therefore in surgical planning [15]. These can also be identified by the presence of signal voids, indicating rapidly flowing blood. In addition, bone erosion is reported in relatively few cases of synovial hemangioma [16-18], although in an 8-patient series, 4 had evidence of erosion on plain films [4].

Despite the accuracy of MRI for diagnosing hemangioma, a diagnosis of PVNS was suggested in this patient. This was based on the identification of a large amount of hemosiderin within the hip joint on MRI, the age of the patient, the degree of bone destruction, and the failure to identify MRI features typical of a hemangioma such as a high SI lobular lesion divided by low SI septa. Although intraarticular hemosiderin deposition occurs not only in synovial hemangiomas but also nonspecifically in any other cause of hemarthrosis, the large amount observed in this case without predisposing factors or identifiable hemangioma was thought most consistent with PVNS. A clinical and diagnostic comparison of synovial hemangioma and PVNS is given in Table 1.

Furthermore, the diagnosis of PVNS was preferred as it is more common in people older than 20 years. It is also associated with an erosive arthropathy in more than 90% of cases, particularly in joints where the synovial space is restricted, such as the hip. Bone destruction is

Table 1. The Clinical, Diagnostic, and Treatment Considerations of PVNS and Synovial Hemangioma

	PVNS	Synovial Hemangioma
Clinical course	Age—typically 20-40 y old Principally hip, knee, and ankle Insidious presentation months or years after onset of symptoms of joint pain Effusion with bone and joint erosion common Recurrence rate significant Malignancy reported	May be seen in children Relatively uncommon Principally affects the knee Palpable swelling, joint pain, restricted movement Effusion and bone erosion uncommon Recurrence rate low Malignancy not reported
Diagnostic and imaging criteria	Plain radiographs and computed tomography may demonstrate high-density effusion and erosions MRI—hyperplastic synovium and heterogeneous lobulated synovial swelling, areas of high SI on T2, with areas of low SI on all sequences. High hemosiderin deposition causing “blooming” on gradient echo sequence Bone and synovial destruction common	Diagnosis often delayed due to failure of plain radiographs to identify abnormality MRI very useful—T2-weighted shows high SI lobulated lesion with low SI linear septae Arthroscopy useful for determining extent of lesion and surgical planning
Histopathologic examination	Proliferation of mononuclear cells admixed with multinucleated cells, foamy histiocytes and hemosiderin deposits Nodular architecture	Randomly distributed blood vessels with capillary, cavernous, or arteriovenous architecture Diffuse hemosiderin deposits
Treatment	Open resection usually preferred as has lower recurrence rate than arthroscopic resection Arthroplasty may be required	Arthroscopic removal may be performed for well-localized or pedunculated lesions but can result in bleeding necessitating subsequent open resection Diffuse lesions require open resection to prevent recurrence

much less common with synovial hemangioma [13,19] and when present, usually results from localized erosion by the adjacent tumor. Widespread synovial erosions and arthropathy with cystic change, particularly with such heavy hemosiderin deposition, would invariably lead to the preoperative diagnosis of PVNS rather than synovial hemangioma. We can find no published case reporting arthropathic destruction due to synovial hemangioma.

The diagnosis of the intraarticular lesion was complicated by the intramedullary femoral findings thought to be cystic fibrous dysplasia after MRI. Cystic foci, which are occasionally quite large, may be seen in juxtaarticular locations in joints affected by PVNS, and this finding made the preoperative diagnosis of synovial hemangioma even less likely.

The only other case of synovial hemangioma of the hip reported in the literature is a 20-year-old woman who presented with a 10-month history of left hip pain and limitation of activity [6]. A small heterogeneous lesion with minimal effusion was identified on T2-weighted MRI, although a radiologic diagnosis was not reached. When the pain did not resolve after 3 months of conservative treatment, arthroscopy of the hip joint was performed and a fragmented lesion was removed. The joint was normal. Histologic investigation confirmed a cavernous synovial hemangioma.

Delayed diagnosis of hemangioma can be problematic. As the lesion grows, it infiltrates the surrounding soft tissue [17] and bone [18] making resection more difficult. Long-standing lesions may result in an inflammatory synovitis and in the knee, and joint damage resembling hemophilia due to recurrent hemarthroses has been described [20]. This appearance has not been described in the hip, and this case demonstrates the destructive capacity of synovial hemangioma that was severe enough to warrant total hip arthroplasty. We can find no other report of synovial hemangioma associated with articular damage sufficiently severe to require prosthetic joint arthroplasty.

Pigmented villonodular synovitis was the major differential diagnosis in this case, and like synovial hemangioma, it may be diffuse or focal [21]. Plain radiographs may appear normal. Focal PVNS appears as a heterogeneous lobulated synovial swelling, often with areas of high T2 SI but characteristically shows some low-signal areas on all sequences, with "blooming" on gradient echo sequences, due to hemosiderin deposition [3,19].

Conclusion

Although MRI increases the accuracy of diagnosing synovial hemangioma, it remains difficult to diagnose and it can be mistaken for PVNS. In this case, the typical MRI features of synovial hemangioma were not seen, and imaging was strongly suggestive of PVNS, particularly the presence of low-signal tissue within the joint. Together with the age of the patient and the joint destruction seen,

these made PVNS the working diagnosis. Hence, histologic evaluation is required for definitive tissue diagnosis. Synovial hemangioma of the hip is extremely rare; this is only the second reported case and the first to describe widespread articular damage necessitating joint arthroplasty. Patients presenting with long-standing nonspecific hip pain should be investigated with MRI to rule out a potentially damaging synovial hemangioma.

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