

THE DIAGNOSTIC CHALLENGE OF SYNOVIAL HEMANGIOMA

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ABSTRACT

Synovial hemangioma is an uncommon benign vascular tumor, which is most commonly localized in the knee and is often difficult to diagnose in its early stage. We report 3 children who presented with knee monoarthritis, which was later found to be due to a synovial hemangioma. In all patients, the diagnosis was confirmed by magnetic resonance imaging (MRI), which dictated additional appropriate invasive investigations. Our report emphasizes the inclusion of synovial hemangioma in the differential diagnosis of atypical monoarthritis of the knee and the value of MRI in identifying the lesion.

INTRODUCTION

The differential diagnosis of monoarthritis in the pediatric and adolescent age is broad and not only includes the acute or chronic inflammatory arthritides, but also a number of extremely rare entities, such as pigmented villonodular synovitis, loose bodies, lipoma arborescens, juxta-articular myxoma, xanthoma, bursitis, meniscal tears, synovial sarcoma, posttraumatic organizing hemorrhage, tuberculosis, sarcoidosis, bleeding disorders, and synovial hemangioma [1-3]. Synovial hemangioma is a rare benign vascular tumor that usually affects young patients and is most common in the knee [4-9]. The diagnosis of this lesion, which frequently causes recurrent joint effusions, is often delayed. When it presents with the sudden onset of painful knee swelling after a local trauma, it is commonly interpreted as traumatic arthropathy. In case of insidious development, usually as painless swelling with no history of trauma, it can be misdiagnosed as juvenile idiopathic arthritis (JIA) [1,4]. As a result, several years frequently lapse between onset of symptoms and treatment [6,7,9].

This report highlights the diagnostic challenges posed by this condition and the usefulness of magnetic resonance imaging (MRI) in delineating the lesion.

CASE REPORTS

Case 1.

A 16-month-old boy developed a painless swelling of his right knee after a fall to the ground. He was brought to his local hospital, where a knee radiograph was performed, which was negative, and a diagnosis of traumatic arthropathy was made. A few days of weight-bearing avoidance and ice-pack application led to prompt reduction of joint swelling, which however did not heal completely. Over the following months the right knee always appeared to the parents slightly more swollen than the contralateral, and a recurrent exacerbation of swelling was often noted following minor trauma. One year after

the onset of symptoms, due to the persistence of knee swelling, he was hospitalized elsewhere. All laboratory tests, including inflammation parameters, antinuclear antibodies, rheumatoid factor, and a repeat plain radiograph were negative. Based on the persistence of knee swelling, a diagnosis of JIA was made and nonsteroidal antiinflammatory (NSAID) therapy was prescribed, which had no effect. The boy was seen at our outpatient clinic at the age of 3 years, shortly after a further episode of knee swelling, preceded by a minor injury. On clinical examination, the right knee appeared swollen, but was modestly tender and painful on passive motion. A significant joint effusion and a slight restriction of flexion were appreciable. The remaining joints and the general physical findings were within normal limits. Knee ultrasonography showed marked effusion and also revealed an area of hyperechogenic material in the superpatella region, suggesting prominent synovial hypertrophy. Since there was a discrepancy between a clinical and ultrasonographic picture potentially consistent with JIA and a history of a close relationship between the exacerbation of joint swelling and trauma, an arthrocentesis was performed. Aspiration revealed 20 ml of frank blood. To rule out a bleeding disorder, the clotting tests (including platelet count, prothrombin time, activated partial thromboplastin time, and fibrinogen level) were requested, which yielded normal results. An MRI of the knee was then performed, which demonstrated an extensive lesion, with low signal in T1-weighted images and high signal in T2-weighted and stir images, nearly occupying the entire synovial cavity and a large fluid collection ([figure 1](#)). A diagnostic arthroscopy led to the histopathologic diagnosis of synovial hemangioma ([figure 2](#)). One week later, the lesion was resected surgically. At 3 months after surgery, the boy was asymptomatic.

Case 2.

A 3-year-old girl presented to her family physician with a painful swelling of the medial aspect of her left knee, which was ascribed to a recent bump into home furniture. The joint symptoms resolved within few days with NSAID therapy. During the following years, she experienced intermittent episodes of swelling of her left knee, often preceded by trauma. Six years after the onset of symptoms, an arthrocentesis revealed hemarthrosis. A bleeding disorder was sought and the coagulation tests showed decreased fibrinogen levels. A defect in fibrinogen synthesis was hypothesized. For this reason, the subsequent episodes of knee swelling were treated with an anti-fibrinolytic agent (tranexamic acid), which, however, did not affect symptoms. At admission to our hospital, one year later, the left knee appeared slightly swollen, but was not tender, and no effusion was detectable clinically. A knee ultrasonography revealed only a slight effusion in

the suprapatellar pouch. MRI showed a mass in the anteromedial aspect part of the synovial cavity, which appeared slightly intense in proton density images and hyperintense in T2-weighted images. Diagnostic arthroscopy and biopsy revealed synovial hemangioma. The mass was excised surgically. At one year, the girl is free of symptoms.

Case 3.

A 7-year-old boy was admitted to his local hospital because of the occurrence, in the absence of a previous trauma, of swelling, aching and functional limitation of his right knee. A knee radiograph was negative. A traumatic arthropathy was diagnosed. The knee was casted and NSAID therapy was given for one month with good results. Two months later, the swelling of the right knee recurred after a minor injury. Arthrocentesis of the right knee was performed and bloody fluid was obtained. The screening for coagulation defects was negative and the swelling regressed spontaneously within a few days. After two years without complaints, the boy had a new episode of painful knee swelling following a trauma. Repeat coagulation screening and a plain radiograph did not reveal abnormalities. At admission to our hospital, the right knee appeared slightly swollen, but neither painful nor functionally limited. Based on the patient history, a working diagnosis of synovial hemangioma was entertained. An MRI was therefore performed, which revealed a mass in retropatellar region, that was best demonstrated on T2-weighted images ([figure 3](#)), without synovial effusion. Arthroscopy and biopsy confirmed the diagnosis of synovial hemangioma and the mass was removed surgically. At 6 years, there has been no recurrence.

DISCUSSION

The clinical diagnosis of synovial hemangioma is often elusive. Diagnosis was made preoperatively in only one third of the cases reported in the literature [6]. In the patients described herein, the lesion was identified only 1.5, 7, and 2 years after the onset of symptoms, respectively. All lesions were localized. Early diagnosis of synovial hemangioma is important because recurrent hemarthrosis may lead to chronic inflammatory synovitis and joint damage [1,7]. In the reported patients, clinical examination and plain radiographs did not prove helpful and ultrasonography was misleading. The diagnostic suspicion was based on the history of recurrent joint swelling after a minor trauma and was confirmed by MRI, which dictated additional appropriate invasive investigations. The main clinical features and results of typical JIA tests in the study patients are reported in table 1.

Synovial hemangioma has been defined as a benign vascular lesion arising from any structure lined by synovium, including the intra-articular region, bursal spaces, and tendon sheaths [4]. Some authors do not include vascular lesions arising from tendon sheaths in the classification of synovial hemangioma, based on the fact that such lesions are not confined by a synovial structure [4]. Synovial hemangioma can occur as a single pathology in one or several joints or as a systemic disease (Maffucci syndrome, von Hippel-Lindau syndrome, or Kasabach-Merritt syndrome). The most typical form of synovial hemangioma is the intra-articular type in which the tumor forms a mass lined by synovial membrane [7]. These tumors almost invariably involve the knee joint, although they have also been found in the elbow, wrist, finger, ankle, and temporomandibular joints, as well as in the tendon sheaths [4,7,9]. The nature of the lesion is also controversial: it is unclear whether it is a true neoplasm or simply an hamartomatous (nonneoplastic) vascular proliferation [4,7]. The presentation of most lesions at a young age suggests that synovial hemangioma is a form of vascular malformation [10]. Trauma is unlikely to be of relevance in the pathogenesis. Anatomically, it has been described as synovial, juxta-articular, and intermediate and can be pedunculated or diffuse. The hemangiomas can also be further classified by the nature and size of the blood vessels that predominate within the hemangioma. These classifications include capillary, cavernous, arteriovenous, and venous types. [11-14].

No information exists on the incidence or prevalence of synovial hemangiomas. It has been estimated that fewer than 200 cases have been reported to date [7]. It has been reported that the average age of onset is 10.9 years in girls and 12.5 years in boys, and that 75% of patients are symptomatic prior to age 16 [15]. The typical patient with synovial hemangioma is a child or young adult (with a slight male predominance) who presents with a swollen, painful knee or elbow. The complaint is often chronic, with reports of previous relapses and remissions. Relapses are due to bleeding from the hemangioma and are frequently, although not invariably, preceded by a minor trauma. With time, quadriceps muscle atrophy and some restriction of joint motion can become evident. Occasionally, on physical examination, a tender spongy or firm mass may be palpable, which may decrease in size with elevation of the extremity. Aspiration of the lesion or synovial fluid often yields bloody fluid. Radiographs are generally unrevealing, although a vague soft tissue mass or localized phleboliths may be seen in some cases on plain films of the affected joints. Phleboliths, which are most common in cavernous hemangiomas, may offer a clue to the diagnosis. Ultrasonography may allow the definition of the size and location of the lesion, but images may be confusing, like in our first case, with prominent synovial hypertrophy.

As demonstrated in our patients, the extent of the lesion is more accurately judged by MRI, which allows an earlier diagnosis and provides valuable information for planning the treatment. Angiography and CT scan are generally not necessary when MRI is used [6]. The MRI appearance of hemangioma is frequently characteristic [16]. On T1-weighted images, it is typically poorly marginated and isointense to skeletal muscle. Within the lesion are areas of increased signal, approximating that of subcutaneous fat. On T2-weighted images, which are highly suggestive for the diagnosis, the lesion is typically well marginated and markedly hyperintense as compared with subcutaneous fat. Yet portions of the hemangioma also may be isointense to fat and/or muscle. Diagnostic arthroscopy and biopsy are useful adjuncts to confirm the nature of the lesion. Accurate and adequate preoperative assessment assists in the classification of the lesion and guides definitive management with the aim of complete resection to minimize the risk of recurrence [9]. The diagnosis is made histologically on the operative specimen [4]. Various treatment strategies have been suggested in the past, including embolization, open surgical resection with partial or total synovectomy, arthroscopic excision, radiotherapy, cautery, freezing, use of sclerosing agents, and laser arthroscopic ablation [7,8]. In the localized type of tumor, the treatment of choice is open or, if possible, arthroscopic surgical excision [7]. Most authors prefer the open technique because arthroscopic surgery may produce bleeding problems and arthroscopic radical resection is not always possible [9].

Because synovial hemangiomas are frequently misdiagnosed, leading to a diagnostic delay of many years, a high index of suspicion is necessary to improve early diagnosis. This condition should be suspected in any child who presents with a history of intermittent pain and swelling in a single joint, particularly a knee, which is often exacerbated by minor traumas and in whom an inflammatory or infectious process or a bleeding disorder is ruled out on clinical grounds and/or through appropriate laboratory tests. In such patients, MRI represents the diagnostic procedure of choice because it may allow an earlier diagnosis and sometimes obviates the need for more invasive investigations such as arthroscopy. More in general, children with monoarthritis should be selected to receive an MRI if they have symptoms, signs or radiographs atypical for an inflammatory arthritis, they fail to respond to conventional therapy for arthritis, or they have features more typical of internal derangements [2].

In summary, we emphasize two things: [1] the inclusion of synovial hemangioma in the differential diagnosis of atypical monoarthritis of the knee; [2] the value of MRI in identifying the lesion.

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Table 1. Main clinical features and results of typical JIA tests in the study patients.

	Patient #1	Patient #2	Patient #3
Gender	Male	Female	Male
Age at onset of symptoms	16 months	3 years	7 years
Initial diagnosis	Traumatic arthropathy, then JIA	Traumatic arthropathy, then bleeding disorder	Traumatic arthropathy
Time lag onset-diagnosis of hemangioma	20 months	6 years	2 years
Location	Right knee	Left knee	Right knee
ESR (mm/h)	13	9	9
C-reactive protein (mg/dl)	Neg	ND	Neg
Antinuclear antibodies	Neg	Neg	Pos (1:80)
Rheumatoid factor	Neg	Neg	ND

JIA: juvenile idiopathic arthritis; ESR: erythrocyte sedimentation rate; ND: not done

FIGURE LEGENDS

[Figure 1.](#) Case #1. Synovial hemangioma occupying nearly the entire synovial cavity of the right knee seen on magnetic resonance T1-weighted image.

[Figure 2.](#) Case #1. Cavernous blood spaces beneath the proliferated synovial cells.

[Figure 3.](#) Case #3. Synovial hemangioma in the retropatellar region on magnetic resonance T2-weighted image.