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Synovial hemangiohamartomas of the knee joint

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Abstract Hemangioma or vascular malformation of the synovium is rare and presents a difficult problem in diagnosis and treatment. A long history of joint pain and recurrent non-traumatic hemarthrosis usually draws attention to the hemangioma of the knee joint. The lesion can be seen in two different formations; the synovial hemangioma or the arteriovenous malformation named as hemangiohamartomas, both of which involve the synovium and cause non-traumatic episodes of hemarthrosis. MRI scanning together with arthroscopy is a diagnostic tool to demonstrate the extent and the nature of the lesion. We treated the three patients at different ages. All patients underwent standard radiographic examination, CT scans, MRI and diagnostic arthroscopy. After frozen section taken via arthroscopically, the lesions were excised by arthrotomy. The mean follow-up was 38 months (31–45) and all patients are asymptomatic postoperatively. Three additional cases and a review of the literature are presented because of the rarity of the lesion.

Keywords Hemangiohamartoma · Hemarthrosis · Arteriovenous malformation

Introduction

Synovial hemangioma is a scarce benign soft-tissue tumor, which can be observed both localized and diffused [15]. The predominant type of vascular channel observed

at histologic findings classifies the lesions pathologically: cavernous, capillary, arteriovenous or venous [4].

Synovial hemangioma is usually manifested in adolescence or young adulthood. The knee is frequently involved although the ankle, the elbow, the temporomandibular joint and the shoulder can be affected [3].

Pain is a prominent symptom, which exists both with movement and on palpation. Recurrent non-traumatic hemarthrosis, limitation of knee motion occasionally intermittent, tender palpable mass and quadriceps-muscle atrophy are other common findings. No definite prior trauma is noted [3, 14].

The diagnosis is often overlooked because conventional radiographic techniques may omit the presence of the lesion. MRI demonstrates the extension and the nature of the synovial hemangioma. Diagnostic arthroscopy, MRI, red blood cell scans and closed-system venography have been recently used in the diagnosis [19].

Localized or pedunculated lesions are treated easily with arthroscopic excision. Complete open excision decreases potential for local recurrence. However, small doses of irradiation are occasionally applied to the patients with diffuse lesions because of the difficulty in complete surgical removal [1, 2, 6, 19].

Three cases of synovial hemangiohamartomas in suprapatellar pouch and the correlation between MRI scanning and arthroscopic findings are presented in this study.

Case reports

Three patients have been complaining of several episodes of pain, swelling and buckling of their knees for 6 years (4–8 years). We examined the three patients (three females), aged 11, 21 and 22 years at the time of surgery. The right knee was involved in two cases, the left knee in one. There was no history of trauma but tenderness on superolateral aspect of suprapatellar pouch in two cases. We examined palpable mass on the

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medial side of the knee joint within vastus medialis muscle in the other case. All patients had full active and passive range of motion of the knee joint. There was no cellulitis, or fluctance, erythema, discoloration and lacerations. None of the patients had limb length discrepancy.

Case 1

The youngest patient was an 11-year-old girl with a mild effusion, quadriceps-muscle atrophy and palpable mass on superolateral aspect of suprapatellar pouch. She has been complaining of anterior knee pain and spontaneous recurrent hemarthrosis for 4 years. There was no history of trauma. We observed hemarthrosis and limitation of knee motion during physical examination that was considered as positive signs for any internal derangement.

Case 2

She was a 21-year-old woman with quadriceps-muscle atrophy. There was tenderness and palpable mass on superolateral aspect of suprapatellar pouch. The physical examination was negative for any significant internal derangement. She had no limb length discrepancy.

Case 3

A 22-year-old woman has been complaining of buckling of her left knee, spontaneous recurrent hemarthrosis for 8 years. There was palpable mass on the medial side of the knee joint within vastus medialis muscle. She had full painless active and passive range of motion of the knee joint. There was no cellulitis, or fluctance, erythema and discoloration.

In all three cases, routine laboratory investigations were within normal limits and no coagulopathy was detected. Besides, there were no vascular malformations in the other member of their families.

All patients underwent standard radiographic examination, CT scans, MRI and diagnostic arthroscopy. They are all asymptomatic postoperatively.

Plain radiography of the right knee revealed an increase in soft-tissue density in the suprapatellar pouch. There was no sign of phleboliths (Fig. 1) and we used CT in diagnosis.

CT scans demonstrated a mass, which is 4.5×4 cm in diameter, not well circumscribed in suprapatellar pouch. It also revealed minimal bone invasion (Fig. 2).

Magnetic resonance imaging scans showed a non-contrast T1-weighted image that demonstrated intermediate signal intensity and isointense with adjacent muscle (Fig. 3a). T2-weighted and fat suppressed



Fig. 1 Case no.1: patient aged 11 years with a mild knee effusion. X-ray shows an increased soft-tissue density in the suprapatellar pouch

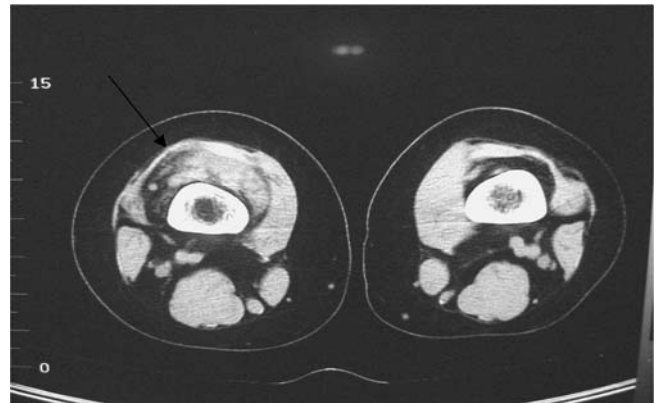


Fig. 2 Case no.2: 21-year-old-woman with a 2.5 cm quadriceps-muscle atrophy. CT scans demonstrate minimal bone invasion

images revealed high signal intensity in the suprapatellar pouch (Fig. 3b).

Arthroscopic findings revealed a synovial lesion in the suprapatellar pouch embedded in synovium. During the puncture of the synovium acute bleeding occurred, and soon after the coagulation diffuse bleeding focal points on the synovium in the suprapatellar pouch appeared (Fig. 4a–d). We observed minimal erosion on the metaphyseal bone. The appearance of the lesion warned us against synovial hemangioma.

A 0.50 cm³ sample was arthroscopically taken for frozen section. Frozen section diagnosis was a benign lesion originated from vascular structures. After frozen section, the lesions were excised by standard medial parapatellar approach. Remnants of the lesion were removed using curettage and the joint capsule was closed in a routine manner.

Histologically on gross examination, a purplish, soft, lobulated mass was connected to the synovia. The blood existed inside the lesion. Microscopically, a melange of large venous, cavernous and capillary sized vessels

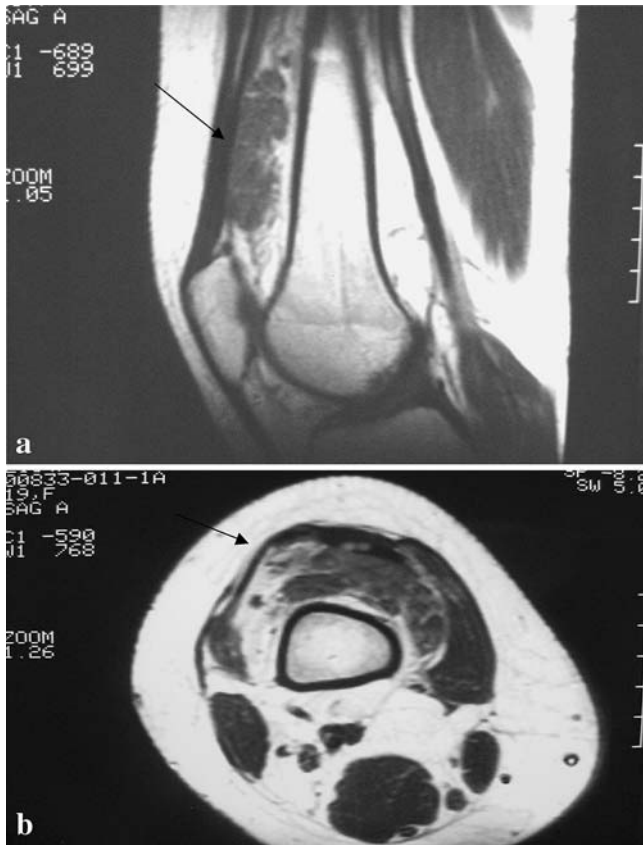


Fig. 3 Case no.2: **a** non-contrast T1-weighted image demonstrated intermediate signal intensity and isointense with adjacent muscle. **b** T2-weighted and fat suppressed images revealed high signal intensity

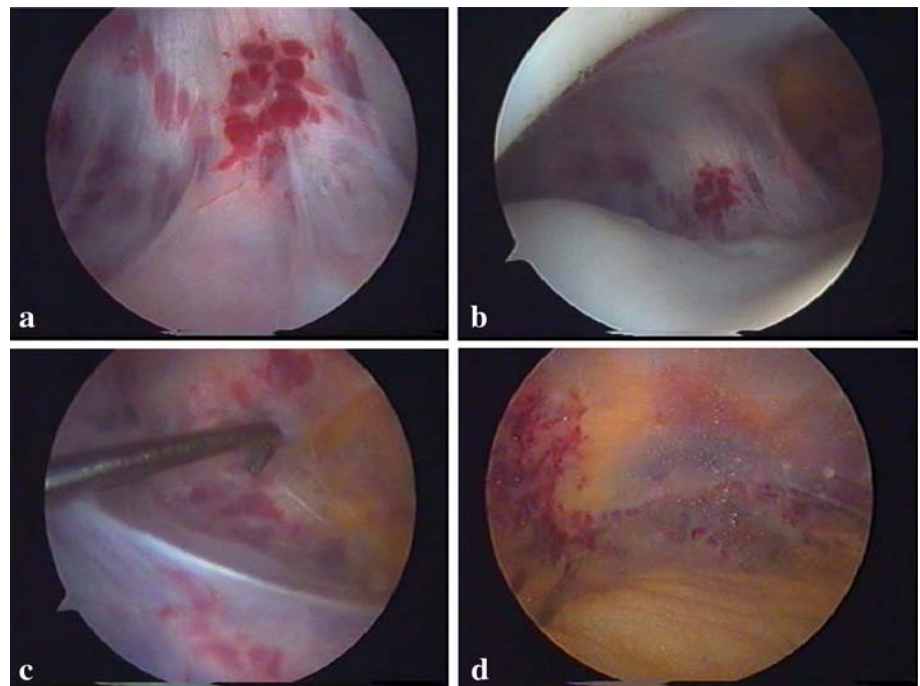
scattered incidentally throughout soft-tissue. The venous vessels were remarkable for their thick walls, which consisted of occasional attenuation and herniations of the wall (Fig. 5). Diagnosis is angiomatosis. The mean follow-up was 38 months (31–45) and all patients are asymptomatic postoperatively.

Discussion

The hemangiomas originate from subsynovial tissues and they are appealingly hamartomas or congenital vascular malformations rather than true neoplasm. The lesion has been described anatomically as synovial, juxta-articular and intermediate. It may invade throughout the entire synovium, fat tissue and muscles unless they are diagnosed in an early stage [1]. Histologically, they may be grouped as a capillary, cavernous, arteriovenous or purely venous. Hence, most of those in the synovial membrane of knee joint have been of cavernous type [14, 24]. There is only one reported case in which synovial hemangioma invades the femur [21]. We did not observe apparent bone invasion; however, in one case there was adjacent muscle invasion in vastus medialis. Pathologic diagnosis of our cases is angiomatosis, with the lesions inside the knee joint and there is minimal erosion on the metaphyseal bone.

Synovial hemangioma usually affects children, adolescent and young adults. Approximately 75% of patients are symptomatic prior to age 16. Girls have a high propensity in the first few decades of their lives [3]. We examined three patients aged 11, 21 and 22 years and their initial complaints began 6 years ago (4–8 years). In no case, bilateral involvement has been reported [14]. In

Fig. 4 Case no.3: 22-year-old-woman with buckling of her knee for 8 years. Arthroscopic findings show a synovial lesion in the suprapatellar pouch embedded in synovium



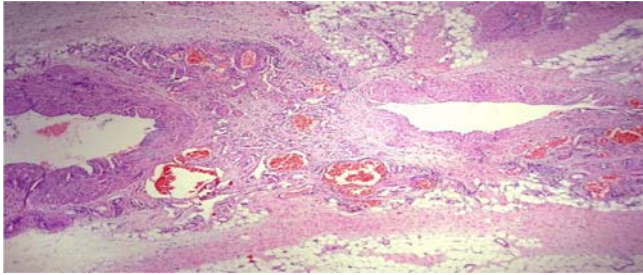


Fig. 5 Case no.3: histological aspect of biopsied synovial tissue. Melange of large venous, cavernous and capillary sized vessels scattered incidentally throughout soft tissue. (Hematoxylin and eosin, $\times 40$)

general perspective, no definite history of trauma was noted [3, 14], as it was the case in our patients.

A history of intermittent pain and swelling is the most frequent symptom. Recurrent spontaneous hemarthrosis of the knee should warn the orthopedic surgeon against synovial hemangioma. Limitations of knee motion caused by mechanical interference, tender palpable mass and quadriceps-muscle atrophy increased limb length on the involved side. Other findings are vascular involvement of the superficial skin and seemingly disappearance of the palpable mass upon limb elevation or bed rest [8, 9, 10, 13, 14, 18]. Our patients had tender palpable mass, quadriceps-muscle atrophy, several episodes of pain and swelling as well as spontaneous recurrent hemarthrosis.

Nodular pigmented villonodular synovitis, synovial chondromatosis, xanthomas, hemangiomas and lipomas are among the most frequent benign tumors of the knee joint. Other entities that should be taken into consideration are lymphangioma, myxoma, meniscal cyst, inflamed bursa, neuroma, myoma and less frequently synovial sarcoma [3, 6, 22]. Most of the cases have normal plain radiographs. Abnormal radiographic views are as follows: soft-tissue mass, phleboliths, arthritic changes, periosteal thickening of the femur or tibia, bone atrophy, osteolysis of femoral or tibial condyles [14]. In our case, plain radiographs reveal an increased soft-tissue density in the suprapatellar pouch but they show no phleboliths. The diagnosis is often overlooked because conventional radiographic techniques may omit the presence of the lesion. That is why the symptom of our patients lasts for 6 years (4–8 years).

Angiography is beneficial in demonstrating the vascular nature of the lesion, however it is not diagnostic for cavernous hemangiomas because they do not reveal a balanced relationship between the lesions and the surrounding structures [1, 5, 11, 19]. Thermographic assessment is used to define the extent of synovial hemangioma [12]. For this reason we did not perform angiography, since it is an invasive method.

Demonstration of an intra-articular lesion on CT scan can be useful in the diagnosis, however it is not specific. On the other hand, the relationship of the lesion to the adjacent muscle is not obvious [1, 5]. We obtained CT scans and they reveal a mass, which is 4.5 \times 4 cm in

diameter, not well circumscribed in the suprapatellar pouch.

MRI scanning is a diagnostic, non-invasive and non-irradiating method. It reveals superior tissue contrast and is more reliable than CT in demonstrating the extent and the nature of the soft-tissue tumors [1, 5, 19]. Furthermore, the extent and the location of the lesion are crucial in the classification and definitive treatment of vascular malformation [15, 16, 19]. MRI findings of our cases are non-contrast T1-weighted image that demonstrated intermediate signal intensity and isointense with adjacent muscle. T2-weighted and fat suppressed images revealed high signal intensity.

Both MRI and arthroscopy offers valuable and reliable information for diagnosis and treatment [1, 2]. In our case, arthroscopic findings reveal a synovial tumor in the suprapatellar pouch embedded in the synovium. During the puncture of the synovium, acute bleeding occurred and after coagulation a purplish, soft, diffuse lesion appeared.

A thorough review of the literature demonstrates different treatment options namely arthroscopic excision, open surgical resection with partial or total synovectomy, embolization and arthroscopic ablation using a holmium:YAG laser [7, 13, 17, 18, 20]. The treatment modality in localized or pedunculated form is arthroscopic surgical resection with or without laser [7, 19, 23]. Diagnostic arthroscopy is a beneficial method, however, in most of the reported cases arthrotomy is inevitable because of the diffusion of the lesion and the bleeding [7, 17, 19]. Arthroscopic ablation using a holmium:YAG laser is useful in decreasing joint trauma and intra-operative bleeding, but it is not usually indicated in diffuse lesions [21]. If there is chondral degeneration, the incidence of local recurrence is relatively high [1]. Open complete excision should be considered in diffuse form together with pre-operative embolization in the existence of large feeder vessels [1, 19, 23]. In addition to this, open complete excision decreases the risk for local recurrence. During arthroscopy, we observed that all of the lesions were diffuse and there was no chondral degeneration. We preferred total excision of the lesions via arthrotomy. After open complete excision, limitation of the knee joint motion is a possible complication but we did not observe such morbidity. However, low doses of irradiation should be used in diffuse lesions because of the difficulty in complete surgical removal [14, 16]. Radiation should not be routinely used because of late adverse effects.

Conclusion

If a clinical portrait of a young girl complaining of unilateral intermittent knee pain, swelling for several years together with non-traumatic episodes of recurrent hemarthrosis, limitation of the knee motion and quadriceps-muscle atrophy is drawn, then the orthopedic surgeon should be alert against synovial hemangioma.

Only the plain radiograph may omit the presence of the lesion. MRI scanning in collaboration with arthroscopy offers valuable information for pre-operative planning. If the tumor is localized, well circumscribed and appealingly encapsulated, arthroscopic excision should be the choice of the treatment. Diffuse lesions are difficult to excise arthroscopically, so that open complete excision should be exercised. In addition to this, open complete excision decreases the risk for local recurrence. After open complete excision, limitation of the knee joint motion is a possible complication.

Therefore, we recommend routine MRI scanning together with diagnostic arthroscopy in case of synovial hemangioma.

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