CASE REPORT

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Arteriovenous synovial hemangioma of the popliteal fossa diagnosed in an adolescent with history of unilateral congenital clubfoot: case report and a single-institution retrospective review

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Abstract

Synovial hemangioma (SH) is a very rare soft tissue tumor; in our department, SH represented 0.07% from all soft tissue tumors (one case from 1311 soft tissue tumors), and 0.78% from all excised hemangiomas (one case from 128 hemangiomas) diagnosed over a five-year period. The aim of this paper was to present the clinicopathologic characteristics of hemangiomas and particularities of one SH of the popliteal fossa diagnosed in an athletic adolescent with previously corrected congenital clubfoot. To our knowledge, this is the 275 case of reported SH. A 13-year-old trick cyclist presented with two-year history of slowly growing mass of the left posterior fossa. The magnetic resonance imaging of the left knee showed a juxta-articular mass with intramuscular component. Open excision of the tumor and partial removal of the synovial membrane was the therapy of choice. Histopathological examination revealed clusters of large arteries and veins embedded in a fibrotic tissue, the tumor mass being lined by synovial membrane. Intramuscular growing was also confirmed. Without any other postoperative therapies, no recurrence or functional disorders were noted after 21 months of follow-up. SH of the knee should be excised as soon as possible to avoid complications such as muscle invasion and risk of recurrence.

Keywords: synovial hemangioma, arteriovenous, malformation, knee, popliteal fossa.

Introduction

Hemangiomas are benign proliferations of blood vessels that can occur anywhere through the human body. It is still unclear if this is a real benign neoplasm or vascular malformation; this is the reason why the therapy of these cases is not yet standardized. As regarding soft tissue hemangiomas, they can display capillary, venous or arteriovenous architecture and can be located in the dermis or in the deeper tissues such as skeletal muscles; well-defined and diffuse growth was described [1, 2].

As regarding the hemangiomas of the joints, they arise in the synovium-lined surfaces, such as intra-articular space or bursa. Synovial hemangioma (SH) is a very rare well- or ill-defined benign proliferation of blood vessels, which underlines the synovium and usually shows a cavernous aspect [1, 3]. To date, about 274 SHs have been reported in English literature; unilateral knee, elbow, wrist, ankle, and temporomandibular joints can be affected [1, 3, 4]. The clinical symptoms vary from localized periarticular swelling or slowly growing painless nodules to recurrent pain, spontaneous atraumatic hemarthrosis, muscle atrophy, and intermittent claudication [1, 3–6]. In less than 5% of patients, degeneration of bone cortex, osteoporosis, periosteal reaction, arthropathy, and chronic hemorrhagic synovitis can occur [1, 4].

The clinical diagnosis of SH is based on arthroscopy but the magnetic resonance imaging (MRI) proved to be the diagnostic procedure of choice that allows not only to identify but also to appreciate the tumor size and the status of the surrounding tissues [1, 3, 5]. However, several cases are misdiagnosed and diagnostic delays of 20–40 years, with an average of 6.8 years, were reported [3, 5].

The aim of this study was to perform a retrospective review of our database for soft tissue tumors, to appreciate the incidence of excised hemangiomas in our institution and to present a historical update of SH based on a representative case and data from literature.

Case report

To identify the incidence of SH in our department, the database for surgically specimens of soft tissue tumors of Department of Pathology, University of Medicine and Pharmacy of Tirgu Mures, Romania, was retrospectively searched for a period of five years (2009–2013). The clinicopathologic and microscopic features of one SH of the left popliteal fossa diagnosed in an adolescent boy were presented in detail. Signed consent of the parents was obtained for detailed publication of this case.

Incidence and clinicopathological aspects of hemangiomas

Out of a total of 1311 soft tissue tumors diagnosed over a period of five years, 128 (9.76%) hemangiomas

were identified. All of the hemangiomas were diagnosed in males with a median age of 29.42±15.34 years, ranging from three months to 84 years. Although similar distribution among the age period was seen, most of the cases (54.69%) occurred in patients younger than 30 years, being localized in head and neck area (Table 1).

Table 1 – ***Clinicopathological characteristics of excised soft tissue hemangiomas***

Characteristic	No. of cases (*n*=128)	Percentage
Gender		
Male	128	100%
Female	–	–
Age		
<1 year	32	25%
1–10 years	16	12.50%
11–20 years	16	12.50%
21–30 years	6	4.69%
31–40 years	18	14.06%
41–50 years	7	5.47%
51–60 years	11	8.59%
61–70 years	12	9.38%
71–90 years	10	7.81%
Localization		
Head and neck	67	52.35%
Trunk/abdominal wall	24	18.75%
Superior limbs	19	14.84%
Inferior limbs	18	14.06%
Histology		
Capillary	90	70.31%
Cavernous	34	26.56%
Arteriovenous	3	2.35%
Synovial	1	0.78%

From the 18 hemangiomas of the inferior limbs, 12 involved the left leg, the other six being seen in the right ones; no bilateral involvement was noted. These tumors involved the thigh (*n*=6), ankle (*n*=8), and knee region (*n*=4). Microscopically, they displayed a cavernous (*n*=10) or capillary (*n*=7) architecture, one of the cases being a SH.

As regarding the four hemangiomas of the knee region, three of them were extra-articular cavernous hemangiomas identified in the medial and lateral area of the right knee (one patient had 18-year-old, the other two having 59 and 43 years, respectively). The fourth case was a SH localized in the left popliteal fossa, which represented 0.07% from all soft tissue tumors, and 0.78% from all hemangiomas, respectively. The details of this case were presented below.

The synovial hemangioma – diagnosis and therapy

A 13-year-old male presented with a history of two-year slowly growing mass in the left popliteal fossa. No significant familial or personal medical history was noted, except for spina bifida occulta S1 and a left congenital clubfoot with moderate Dimeglio score (category II) [7], which was successful corrected in his childhood with an orthosis. He had no complaints in any other joints. The patient's weight was of 48 kg; the tallness was of 1.65 m. All of the serum parameters, including the coagulation profile, were within normal range.

At the present admission, the patient related that he had a bicycle accident two years ago; a hematoma was formed then in the popliteal fossa, without skin lesions. The hematoma was resolved in few days; a persistent swelling of the popliteal fossa was noted. Clinical examination revealed flexion contracture of the left knee (45^0), thigh muscle atrophy, and a prominence of the popliteal fossa and lateral aspect of the thigh. On palpation, we found an approximately 20×5 cm, ill-defined, non-pulsatile soft mass, starting from the left popliteal fossa and continued along the biceps femoris tendon and muscle body. The mass was covered by intact skin, with moderate pain on palpation. Knee range of motion was painful, and reduced by 20^0 in flexion and extension. Tibialis posterior pulse was normal bilaterally. No meniscal or patellofemoral abnormalities were detected on clinical examination.

Plain radiographs of the left knee showed a soft tissue elongated mass without any bone lesion. The MRI revealed an 18×4 cm ill-defined inhomogeneous soft tissue lobulated mass in the posterolateral region of knee joint, without any intra-articular effusion, bony or cartilaginous lesions, or periosteal reaction; the tumor mass had a diffuse growth and infiltrated the biceps femoris muscle (Figure 1).

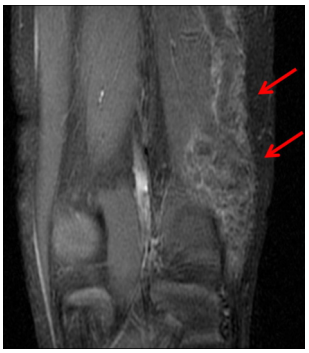

Figure 1 – ***Non-contrast coronal-MRI aspect of the lobulated inhomogeneous synovial hemangioma.***

Open arthrotomy and surgical excision of the tumor was decided to be done. After incision of the skin and popliteal fascia, intraoperative exploration revealed an juxta-articular ill-defined bluish lobulated mass, that also involved the posterior knee joint, in direct relation to the synovial membrane, and extension in the biceps femoris muscle. Tumor excision and partial synovectomy were performed, with free resection margins; the intra- and extra-articular masses were removed, while synovium was electrocoagulated for hemostasis purpose.

The surgical specimen was sent to the pathologist in five fragments with hemorrhagic areas on cut section; the quality of the resection margins was not characterized.

Under microscope, blood-filled large vascular clusters incorporated in fibrotic tissue were seen among the skeletal muscles and adipose tissue. Most of these fragments were covered by synovial membrane. To determine the type of proliferated vessels, examination in Hematoxylin–Eosin (HE) was completed by Van Gieson (VG) staining. Some of the vessels were veins with thin walls and tortuous lumen, the others were large arteries covered by thick walls lined by smooth-muscle fibers (Figure 2). No arteriovenous shunts even atypical cells were identified. Some of the vessels presented red intraluminal thrombi.

Based on the clinicopathological and microscopic features, the final diagnosis was synovial arteriovenous

hemangioma with intramuscular extension. Postoperative course was uneventful. No recurrences, knee effusions, pain, esthetic or functional disorders were noted after 21-months of follow-up, although the patient is a trick cyclist.

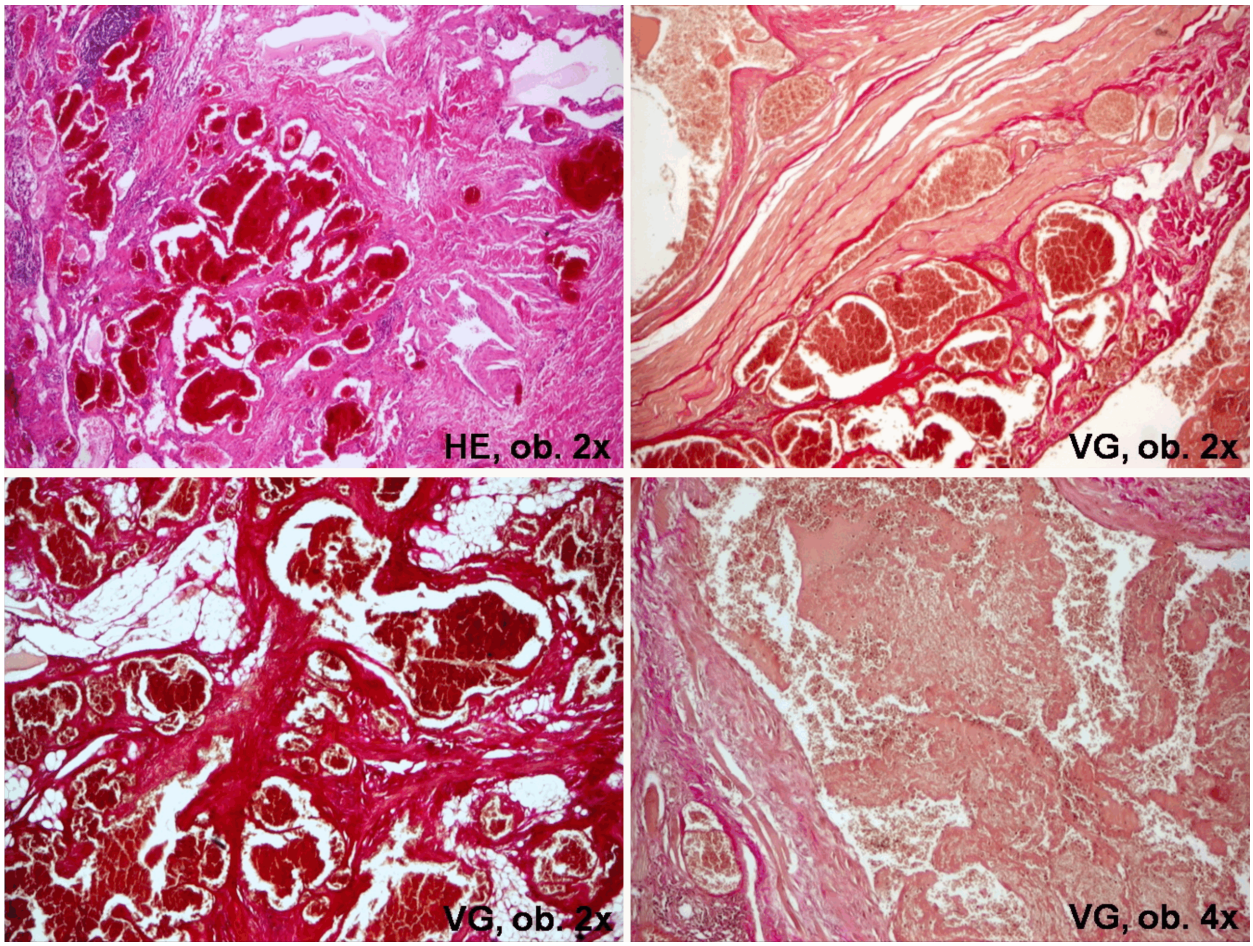

Figure 2 – ***Oversized arteriovenous clusters with intraluminal thrombi, in a synovial arteriovenous hemangioma.***

Discussion

SH was first described by Bouchut in 1856 as "erectile tumor of the knee joint" [8]. From 1856 to 1971, another 104 cases have been added; Forrest and Staple established first positive arthrographic diagnosis of SH of the knee [9]. The number of reported cases increased to 197 in the year 2000 [3] and to about 250 cases in 2011 [10]. Up to now (November 2014), another 24 cases were published on PubMed, the present case being the 275 ones.

SHs typically affect children and young males, with an average age at diagnosis of 13.5 years [3, 11]. Although most of the SHs arise in the knee joint, the anteromedial part is especially involved, without involvement of the bone, cartilage, or muscles [4, 9]. An accelerated epiphysis maturation and disparity in length of the limbs was described in some cases [4, 9].

Similar to the other joints, the knee SH can be diagnosed based on MRI that can be completed by X-ray, angiography, and ultrasonography [5, 6, 11]. Angiograph can identify feeder vessels and allow tumor embolisation [5]. Contrast MRI is necessary to differentiate knee effusion from hemangioma [4, 5]. Echo-Doppler ultrasonography can relieve the segmental popliteal artery stenosis, exacerbated during knee hyperflexion, that is rather characteristic for a popliteal artery adventitial cyst, a degenerative lesion that can communicate with the synovial membrane and is associated with loss of foot pulse with knee flexion (Ishizawa sign) and a popliteal fossa murmur [6]. In our case, angiography was not performed but, however, the thrombosed vessels would not allow a proper examination [5, 9].

As regarding the therapy, SH of the knee can be removed through selective embolisation, arthrotomy arthroscopic excision, arthroscopic ablation with laser or holmium, cautery, freezing, or using of sclerosing agents, in small-sized or pedunculated tumors [4, 5, 11]; open excision is mandatory for large lesions [4].

The SH arising from the popliteal fossa present a challenge with regard to differential diagnosis and therapy. First, a range of lesions should be excluded, including other vascular malformations such arteriovenous malformations that are diagnosed based on the direct connection between oversized arteries and veins lacking the capillary beds, other hemangiomas (cavernous, venous, capillary), lymphangiomas, artropathies (pigmented villonodular synovitis, rheumatoid/juvenile chronic arthritis, Lyme disease, hemophilic arthropathy, synovial osteochondromatosis, osteochondritis dissecans, lipoma arborescens, meniscal tears, discoid menisci, loose bodies, etc.), Baker cyst, and soft tissue sarcomas, especially synovial sarcoma [3, 5, 12]. The last ones should be taken into account especially in diffuse-growing hemangiomas with intramuscular extension, such in the present case. As most of

the patients report a history of knee trauma [3], the aneurysms secondary to traumatic popliteal arteriovenous fistula [13] and postarthroscopic popliteal artery pseudoaneurysm [14], which can be seen on computed tomography [13], should also be considered. Other rare specific lesions that should be taken into account are the Klippel–Trénaunay and Parkes Weber syndromes, which consist of regional prominent capillary-lymphatic-venous malformative dilatation of superficial, muscular, or deep leg veins, most common in the popliteal fossa, soft tissue hypertrophy and overgrowth of bone, with leg length discrepancies [15].

However, to avoid the risk of neurovascular/cartilaginous compromise and functional morbidity that, even in benign tumors, depends on the tumor size [12], soft tissue tumors of the popliteal fossa should be excised as soon as possible [5]. Moreover, recurrent hemarthrosis can cause hemophilia-like joint destruction [3]. Due to proximity of neurovascular structures, wide resection margins are very difficult to be obtained [12], especially in diffuse SH; postoperative radiation therapy may be recommended [5, 9]. In open excision, postoperative complications such as delayed wound healing, wound infection, persistent postoperative pain, postoperative seroma, arteriovenous fistula, and long-term orthopedic complications or ankle weakness, were described [12, 13].

Conclusions

Differential diagnosis of SH of the popliteal fossa should take into account other rare lesions such as soft tissue sarcomas and adventitial cyst. Even benign tumors, SHs of the popliteal fossa represent a therapeutic challenge, especially in large tumors. Early excision is necessary to avoid complications such as infiltration of the muscles and risk for recurrences, especially in healthy athletic adolescents and young adults.

Conflict of interests

The authors declare that they have no conflict of interests.

Acknowledgments

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References

[1] Calonje JE. Haemangiomas. In: Fletcher CDM, Bridge JA, Hogendoorn PCW, Mertens F (eds). WHO classification of tumours of soft tissue and bone. IARC Press, Lyon, 2013, 138–140.
[2] Ciurea M, Ciurea R, Popa D, Pârvănescu H, Marinescu D, Vrabete M. Sinusoidal hemangioma of the arm: case report and review of literature. Rom J Morphol Embryol, 2011, 52(3): 915–918.
[3] De Schrijver F, Geens S. Recurring hemarthrosis of the knee due to synovial hemangioma: report of two cases of which one was treated successfully by arthroscopic excision. Acta Orthop Belg, 2000, 66(1):86–92.
[4] Choudhari P, Ajmera A. Haemangioma of the knee joint: a case report. Malays Orthop J, 2014, 8(2):43–45.
[5] Vakil-Adli A, Zandieh S, Hochreiter J, Huber M, Ritschl P. Synovial hemangioma of the knee joint in a 12-year-old boy: a case report. J Med Case Rep, 2010, 4:105.
[6] Taurino M, Rizzo L, Stella N, Mastroddi M, Conteduca F, Maggiore C, Faraglia V. Doppler ultrasonography and exercise testing in diagnosing a popliteal artery adventitial cyst. Cardiovasc Ultrasound, 2009, 7:23.
[7] Derzsi Z, Gozar H, Gurzu S, Prisca R, Nagy O. Congenital clubfoot in children after walking age: management and evaluation of 41 feet with the Dimeglio score. J Clin Diagn Res, 2013, 7(12):2841–2843.
[8] Bouchut GE. Tumeur erectile de l'articulation du genou. Gaz Hop, 1856, 29:379–381.
[9] Forrest J, Staple TW. Synovial hemangioma of the knee. Demonstration by arthrography and arteriography. Am J Roentgenol Radium Ther Nucl Med, 1971, 112(3):512–516.
[10] Sasho T, Nakagawa K, Matsuki K, Hoshi H, Saito M, Ikegawa N, Akagi R, Yamaguchi S, Takahashi K. Two cases of synovial haemangioma of the knee joint: Gd-enhanced image features on MRI and arthroscopic excision. Knee, 2011, 18(6):509–511.
[11] Dunet B, Tournier C, Pallaro J, Boullet F, Fabre T. Arthroscopic treatment of an intra-articular hemangioma in the posterior compartment of the knee. Orthop Traumatol Surg Res, 2014, 100(3):337–339.
[12] Huh WW, Guadagnolo BA, Munsell MF, Patel S, Lewis VO. Soft tissue sarcomas of the popliteal fossa: a single-institution retrospective review. Cancer, 2011, 117(12):2728–2734.
[13] Masood M, Abdullah SS, Yousuf KM. Giant iliac vein aneurysm secondary to long standing traumatic popliteal arteriovenous fistula. J Coll Physicians Surg Pak, 2012, 22(4): 264–266.
[14] Alserr AH, Antonopoulos CN, Papapetrou A, Kakisis JD, Brountzos E, Liapis CD. Endovascular repair of popliteal artery pseudoaneurysm with arteriovenous fistula after knee arthroscopy: case report and literature review. Vasc Endovascular Surg, 2014, 48(2):166–170.
[15] Uihlein LC, Liang MG, Fishman SJ, Alomari AI, Mulliken JB. Capillary-venous malformation in the lower limb. Pediatr Dermatol, 2013, 30(5):541–548.

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