Imaging strategies in intramuscular haemangiomas: an analysis of 20 cases

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Abstract The purpose of this study was to review the correlation between the imaging studies and the histological findings in the diagnosis of this disease. We retrospectively reviewed 21 lesions in 20 patients (median age, 23.7 years old) who had been diagnosed with cavernous haemangiomas ($n=11$), capillary ($n=6$), and mixed ($n=3$) types. The imaging studies were obtained with plain film radiography ($n=20$), Tc-99 m bone scans ($n=5$), angiography ($n=7$) and magnetic resonance imaging (MRI; $n=20$). All the patients underwent marginal to wide excision. Based on the imaging studies, the rate of accurate prediction of intramuscular haemangioma using MRI in our study was 90%. Using the preoperative imaging studies and surgical excisions, only one (5%) local recurrence happened 2 years after marginal excision. The remaining patients were free of disease. For the avoidance of recurrence, wide excision is necessary with the help of the imaging studies, which can provide more specific information, making possible the preoperative identification of characteristic features of the tumor.

Résumé Le but de cette étude est de montrer la corrélation entre les images radiologiques et les constatations histo-

logiques dans le diagnostic des hémangiomes. Pour cette étude, 21 lésions chez 20 patients (âge moyen 23.7 ans) pour lesquels ont été diagnostiqués un hémangiome caverneux (11), capillaire (6) et mixte (3). Le diagnostic de ces hémangiomes ayant été réalisé par radiographie simple (20), scanner avec technecium (5), angiographie (7) et IRM (20). Tous ces cas ont bénéficié d'une excision large. A partir des images IRM, la prédiction d'hémangiomes intra-musculaires a été de 90%. Seuls 5% des patients présentaient une récurrence locale 2 ans après l'excision, les patients restant n'ayant pas présenté de problèmes particuliers. Afin d'éviter la récurrence, il est nécessaire de réaliser une excision large à partir du bilan radiographique, scanner, IRM.

Introduction

Intramuscular tumors represent the most common type of deep soft-tissue haemangioma, although they are themselves still quite rare [14]. In Watson and McCarthy's study, only 0.8% of haemangiomas were intramuscular [13]; this incidence was also confirmed by Derchi et al. [3]. The rate of correct diagnosis before surgery was only 8% to 19% in the reported literature [9]. Of the reported cases, approximately 45% have been located in the lower extremities [10]. The disease occurs predominantly (90%) within the first 3 decades [9]. The aetiology of intramuscular haemangiomas is still controversial, but has been predominantly ascribed to trauma or congenital theories [10, 12, 13]. Allen [1] (1972) classified intramuscular haemangioma into the small-vessel, large-vessel and mixed types. On the basis of the vascular lumen and endothelial walls, these tumors were classified into the capillary, cavernous and mixed types.

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Pain is a cardinal symptom occurring in 60% of the cases. A mass is always present, and it is found in 98% of the cases [1]. Deeply situated haemangiomas that arise in the deep subcutaneous soft tissue or muscle pose diagnostic difficulties.

MRI is superior in many aspects and may be the modality of choice for the preoperative evaluation of local extent [6, 11]. Most intramuscular haemangiomas show tissue characteristics of intermediate signal intensity on T1-weighted and high-signal intensity on T2-weighted images. Magnetic resonance imaging (MRI) has also been proposed as the diagnostic procedure when an intramuscular haemangioma is strongly suspected. In our observations, the internal structure of these neoplasms on T2-weighted images showed that the cavernous type is predominately lobulated, but the capillary type is serpiginous. The mixed type is mixed lobulated and serpiginous.

Soft-tissue haemangiomas may recur. The mixed type has the greatest tendency for local recurrence (as many as 28% of cases), followed by the capillary (20%) and the cavernous (9%) types [1, 4].

We postulated that preoperative imaging findings for the patients presenting with suspected soft-tissue haemangiomas would have the predictive correlation of magnetic resonance imaging with the final pathological diagnosis. This information could provide an accurate preoperative assessment for planning for the surgeon.

Materials and methods

Patients and assessment

From 1 December 1992 to 30 June 2004, 20 patients underwent marginal to wide surgical excision of an intramuscular haemangioma at our institute. Twelve men and eight women aged 11 to 41 years enrolled in this study. The mean age was 23.7 at the index surgery, and 18 patients (90%) were younger than 30 years old. The duration of symptoms was from 1 month to 10 years. Intramuscular haemangiomas were found in seven in the forearms, five in the thighs, five in the calves, one in the back, one in the buttocks, and one in the foot. All of the patients received the reperfusion test: performing elevation of the upper extremities at more than 90 degrees and the lower extremities at more than 60 degrees, respectively, for 5 min and then putting them down to check how the mass dissipated and emerged again. The patients were studied using plain film radiography ($n=20$), Tc-99 m bone scans ($n=5$), angiography ($n=7$), and MRI ($n=20$).

MRI was performed on a 1.5-T Signa unit (Siemens Vision, Germany) to obtain proton density, T1- and T2-weighted spin echo images as well as multiplanar gradient recalled (MPGR) images in the coronal, sagittal, and axial

planes. Because of time constraints and lack of tolerance by patients, not all of the above pulse sequences were used in every patient; however, at least two orthogonal planes and two sequences of echo were available for review in each patient. Angiography and bone scans were performed using standard peripheral angiographic techniques. After finishing these examinations, intramuscular haemangiomas could be just strongly suspected, but the tests were not pathognomonic. The accurate diagnosis depends on the histological study. The Spearman test was used to detect the correlation between the diagnosis and the characteristics of imaging studies.

Results

The findings of the various studies in the 20 patients are summarised in Tables 1 and 2. According to the Allen (1972) classification of the intramuscular haemangioma, 6 cases were capillary haemangiomas, 11 cases were cavernous, and 3 cases were mixed type. Eleven (55%) out of 20 intramuscular hemangiomas were located in the lower extremities, and 5 lesions were found in the quadriceps muscle group. Seven lesions were in the upper extremities. Eighteen patients (90%) complained of painful masses. The rate of accuracy of the reperfusion test was 60% (12/20). All the positive results were located in the lower extremities. In 7 out of 20 (35%) intramuscular lesions, the plain film demonstrated the phleboliths, which suggested the diagnosis. An unusual calcification appearance with phleboliths and hyperostosis within the same area was shown (Fig. 1a). The plain film could show the abnormality of soft tissue enhancement, calcification, and associated bony erosion when lesions increased in size. We found intramuscular fatty streaks over the painful area in 5 out of 20 patients (Fig. 2). In one intramuscular lesion, a periosteal reaction was seen, yet plain radiographs were normal in seven cases. All of the arteriograms ($n=7$) of the tumour showed several small arteries supplying the lesions. The angiogram could not easily determine the entire extent of the tumour. Five patients underwent technetium 99 m methylene diphosphonate bone scintigraphy, which demonstrated the increased radiotracer uptake over the lesions.

Eighteen of the 20 investigations of MRI showed intermediate signal intensity on T1-weighted spin-echo images and a hyperintense signal in the T2-weighted spin-echo images (Fig. 1b,c). The heavily T2-weighted images showed the best contrast differentiation between the haemangioma and the surrounding normal tissues. The other two patients showed slightly higher signal intensity than the around-muscle signal in the T1- and hyperintense T2-weighted spin-echo images, for which the preoperative impressions were haematoma and fluid accumulation, such as ganglion cyst, respectively. For the correlation of the

Table 1 Patient data

Patient no.	Sex	Age (years)	Duration	Initial complaint	Localization	Reperfusion test	Size (cm)
1	M	12	1 months	Pain and growth of mass	Soleus	–	2.5×2.0×1.0
2	F	41	10 years	Extension block of thumb	Surrounding FCU and FPL	±	FCU:5.0×3.0×2.0 FPL:10×5.5×4.0
3	F	26	2 years	Back pain and palpable mass	Back, Latissimus dorsi	–	6.0×5.0×3.5
4	M	20	4 months	Painful and enlarging mass	Gluteus maximum	+	5.5×2.5×2.0
5	M	32	3 months	Pain and growth of mass	Vastu lateralis	+	8.0×5×2.5
6	M	23	2 years	Pain and Achilles tendon contracture	Soleus	+	5.0×4.0×3.0
7	F	15	12 years	Pain and rapid growth of mass	Proximal forearm	–	4.0×3.0×2.0
8	M	11	5 months	Mass and swelling	Vastu lateralis	+	8.0×5.0×3.0
9	M	20	2 years	Growth of mass and mild pain	Vastu lateralis	+	8.0×4.0×3.0
10	M	20	5 years	Pain and growth of mass over medial foot	Foot, abductor hallucis and quadratus plantae muscles	+	6.0×3.0×2.0
11	F	16	3 months	Rapid growing mass and pain	Brachioradialis	+	9.0×5.0×4.0
12	M	25	1 year	Pain and growth of mass	Rectus femoris	+	9.0×5.0×3.0
13	F	28	6 months	Growth of mass and mild pain	Brachioradialis	±	5.4×3.2×2.5
14	F	29	8 months	Painful and enlarging mass	Quadriceps	+	8.0×4.0×3.0
15	M	26	10 months	Pain and growth of mass	Supinator	–	6.3×5.1×3.5
16	M	24	3 months	Growth of mass and mild pain	Gastrocnemius	+	7.5×5.5×4.0
17	F	27	2 months	Mass and swelling	Triceps brachioradialis	–	3.2×2.5×1.8
18	M	23	2 years	Painful and enlarging mass	Peroneous	+	7.0×6.5×4.2
19	F	29	6 months	Rapid growing mass and pain	Biceps	–	5.5×4.5×3.0
20	M	27	4 months	Mass and swelling	Gastrocnemius and soleus	+	8.0×7.5×5.0

FCU, flexor carpi ulnaris; FPL, flexor pollicis longus; M, male; F, female; + positive; – negative; ± not sure

histological results and preoperative imaging study, the rate of accurate prediction of intramuscular haemangioma in our study was 90% (18/20). The specificity and sensitivity of the cavernous haemangioma were 66.7% and 93.3%, respectively, using the presentation of the lobulated pattern in MRI. The specificity and sensitivity of the serpiginous pattern in MRI to predict the capillary-type haemangioma were 100% and 55.6%, respectively. The values of the Spearman correlation for the diagnosis and the imaging studies about the cavernous and capillary groups were 0.636 and 0.645, respectively ($P<0.005$). This means they are correlated.

Complications

There were two patients (10%) with tendons involved leading to a functional block of the extremities. One patient received Achilles tendon lengthening because of contracture and shortening of the tendon. The other underwent flexor pollicis longus (FPL) reconstruction and lengthening because of tendon contracture and functional impairment of

the forearm. One female patient (5%) developed recurrence of the tumor in the back after 2 years of follow-up.

All patients were followed up for an average of 45 months (range, 15 to 96 months) after surgical intervention. The rate of correct diagnosis of intramuscular haemangioma in our study was 90% (18/20) by the imaging studies. All other patients are free of symptoms and recurrence. No distal metastasis was noted in this series.

Discussion

Liston reported the first skeletal haemangioma in 1843. They constitute about 10% of all benign vascular tumors. A wide anatomical distribution of skeletal haemangiomas has been noted. However, the most commonly involved muscle is the quadriceps muscle, accounting for 19% of all cases [10]. In our series, 55% of the lesions were found in the lower extremities, and 25% were located in the quadriceps muscle group. These findings were compatible with the other reports.

Table 2 Imaging data

Patient no.	Plain film	Bone scan	Angio.	Preoperative MRI (T2)	Histology diagnosis	Treatment	Recurrence
1	Intramuscular fatty streaks	+	++	Serpiginous and lobulated	Cavernous	Wide excision	No
2	Phlebolith and hyperostosis	+	–	FCU: lobulated FPL: lobulated	FCU: cavernous FPL: cavernous	Wide excision and FPL reconstruction	No
3	Negative	–	–	Lobulated	Cavernous	Marginal excision	Yes
4	Negative	–	–	Lobulated	Cavernous	Wide excision	No
5	Phlebolith	–	++	Lobulated	Cavernous	Wide excision	No
6	Periosteal reaction	+	++	Serpiginous and lobulated	Cavernous	Wide excision and Achilles tendon lengthening	No
7	Negative	–	–	Serpiginous	Capillary	Wide excision	No
8	Phlebolith	–	++	Serpiginous	Mixed	Wide excision	No
9	Phlebolith	–	++	Lobulated	Cavernous	Wide excision	No
10	Negative	–	–	Lobulated	Cavernous	Wide excision	No
11	Intramuscular fatty streaks	+	–	Lobulated	Cavernous	Wide excision	No
12	Intramuscular fatty streaks	–	++	Lobulated	Cavernous	Wide excision	No
13	Intramuscular fatty streaks	–	–	Serpiginous and lobulated	Capillary	Wide excision	No
14	Phlebolith	–	–	Serpiginous and lobulated	Mixed	Wide excision	No
15	Negative	–	–	Serpiginous	Capillary	Wide excision	No
16	Intramuscular fatty streaks	–	–	Lobulated	Cavernous	Wide excision	No
17	Phlebolith	+	–	Serpiginous	Capillary	Wide excision	No
18	Negative	–	–	Serpiginous	Capillary	Wide excision	No
19	Phlebolith	–	–	Serpiginous and lobulated	Mixed	Wide excision	No
20	Negative	–	++	Lobulated	Capillary	Wide excision	No

Angio, angiography; + increased radiotracer uptake of bone scan; ++ done; – not done

An enlarging mass, occasionally pain and neurological symptoms secondary to the mass impinging on a nerve bundle are the common presenting symptoms. In our cases, the growth of the mass was found in 14 (70%) and pain in 16 (80%) patients. Functional blocks with tendon contractures were also recognised in two patients clinically. As a result, the intramuscular haemangioma should be strongly suspected when a deeply situated painful growing mass over the extremities or trunk is recognised.

The duration of symptoms caused by intramuscular haemangiomas can range from 2 months to 18 years [1, 14]. The median duration of clinical symptoms in the large-vessel group was longer, and most of the patients complained of pain (90%) [1]. But there was a case with symptoms for only 1 month in our series. So we should not exclude the rapidly growing mass within from suspicion of intramuscular haemangioma.

Skeletal muscle haemangiomas that grow rapidly may easily be misinterpreted as malignancies [1, 5]. Microscopically, the capillary haemangiomas may show marked

mitotic activity, but it does not mean malignancy. The differential diagnosis from large-vessel-type intramuscular haemangioma to angiosarcoma is the most important consideration due to their different treatment and prognosis. The presence of adipose tissue within large-vessel-type tumours is common, and at times may be as conspicuous as to suggest a lipoma. Large-vessel-type tumours were slightly larger than in the small-vessel group, and the postoperative morbidity was more pronounced [1].

Plain radiography, ultrasound, scintigraphy, angiography, CT scan, and MRI may be helpful in the evaluation of intramuscular haemangiomas. The plain film may show the abnormal soft tissue enhancement, calcification and associated bony erosion when lesions increased in size. The adjacent bony change can be categorised as periosteal, cortical or medullary, and the finding of osseous change does not correlate with patient's symptoms [7]. The presence of smooth, round, laminated phleboliths in a lesion where a large venous plexus does not normally exist is the specific finding of cavernous haemangiomas.

Fig. 1 Case no. 4: a 41-year-old woman with cavernous haemangioma in her left forearm. Lateral radiograph (a) shows pronounced soft tissue swelling of the forearm with multifoci of phleboliths and calcifications. Axial T1-weighted (b) spin-echo MR image shows an infiltrative mass (arrows) of intermediate signal intensity within the muscles. Note atrophy and fatty infiltration of the involved muscles. Axial fat-suppressed T2-weighted (c) fast-spin-echo MR image shows a lobulated-like mass (arrows) of high signal intensity infiltrating the muscles. Axial contrast-enhanced T1-weighted (d) spin-echo MR image shows enhancement of the lesion (arrows)

Ultrasound may be useful in confirming the presence of a mass in doubtful cases or if MRI is unavailable. The first use of Doppler sonography or colour-flow imaging in the evaluation of soft-tissue haemangiomas was reported by Greenspan et al. [6] in 1992. They concluded that the flow within most haemangiomas, except for the arteriovenous type, is so slow that it is below the detection sensitivity of Doppler instruments. It is also useful as an intraoperative detector for smaller and non-palpable lesions.

Scintigraphy is rarely used in the evaluation of soft tissue haemangiomas; it is usually reserved for cases in which the plain film revealed bony destruction. The use of three-phase scintigraphy for diagnosing haemangiomas of the extremities could be the other choice. Murata et al. [8] reported that mixed-type haemangioma showed a diffuse increase in radioactivity on both perfusion and blood-pool imaging.

Computed tomography (CT) may delineate a mass lesion with bony involvement that is seen on plain film well. Its greater sensitivity in detecting phleboliths was also superior to the plain film. CT scans may help to differentiate the

Fig. 2 Case no. 1: a 12-year-old boy with mixed-type intramuscular haemangioma in his right calf. The boy had a history of contusion about 1 month before. The plain film shows soft tissue swelling and intramuscular fatty streaks (arrows). Haematoma could not be ruled out. The arteriogram shows enlarged and tortuous anterior and posterior tibial arteries

soft-tissue masses like lipomas, which are well defined and display lower density.

Yuh et al. [15] stressed that haemangiomas are more likely to exhibit an overall nonhomogeneous pattern with serpentine type and signs of muscular atrophy and fat overgrowth in MR imaging. Based on our experience, the internal structures of these neoplasms on T2-weighting have characteristic appearances that can be differentiated: the cavernous type is lobulated, but the capillary type is serpiginous. The contrast-enhanced T1-weighted spin-echo MR images also could show enhancement of the lesions (Fig. 1d). According to our study, the sensibility of using the lobulated type in T2-weighted MR imaging to detect the cavernous type is 93.3% and the serpiginous to the capillary type is 55.6%. The specificity is 66.7% and 100%, respectively. The application of the lobulated type in T2-weighted MR imaging to detect the cavernous type is superior to the capillary type in our study. This is maybe due to inadequate case numbers for capillary haemangioma. These are the preliminary results, and more studies are needed in the future to support them. These findings could provide useful preoperative information to differentiate whether haemangiomas belong to the large-vessel or small-vessel group because the recurrence rate of the capillary type is much higher than the cavernous. These findings could provide surgeons with much more preoperative information for surgical planning for cases of intramuscular haemangiomas. Angiography is also useful in planning a surgical approach for larger lesions, and preoperative embolisation could reduce the amount of blood loss and provide a better view of the surgical field to facilitate surgical dissection [2].

All intramuscular haemangiomas are benign, and no distal metastases were reported [2]. In Allen and Enzinger's report, 18% of the patients had local recurrences, 7% more than once. In the mixed type, the clinical presentation was similar to the large-vessel haemangiomas [1]. While the recurrence rate was the highest, the reason is unknown. In our series, local recurrence happened in only one case of cavernous haemangioma located on the back about 2 years after marginal excision. It may have been due to inadequate determination of the extent of the tumor in deeper layers of the back muscles. In order to reduce recurrence rates, we use the available imaging tools to help us to define intramuscular haemangiomas accurately and to approach the tumor with the plan of wide rather than marginal excision.

An adequate wide excision should be done for all the intramuscular haemangiomas. If wide excision is impractical due to extensive infiltration or the likelihood of risking permanent deformity and/or functional impairment, the preoperative imaging diagnosis could help surgeons to adapt the surgical plan to lower the recurrence rate. Knowing that the mixed- and capillary-type haemangiomas

have the higher recurrence rates, the ultimate goal is to develop imaging strategies preoperatively that could provide accurate imaging diagnoses and optimise the functional outcome.

Conclusion

The preoperative diagnosis of deeply situated intramuscular haemangiomas remains fairly complicated. The choice of various imaging modalities challenges the clinician's decision making and the availability of equipment and financial resources. The demonstration of phleboliths and fatty streaks in plain film is a more specific finding in the initial evaluation of intramuscular haemangiomas. As lesions increase in size, they may show an associated periosteal reaction or even bone erosion. The findings of MRI for these lesions consist of intermediate signal intensity on T1-weighted spin echo images and a strikingly intense signal on the T2-weighted spin echo images; these are characteristic, but not pathognomonic. Careful preoperative judgment and wide excision could prevent local recurrence, and this is usually curable. The associated reconstruction procedure may be considerable if intramuscular haemangiomas interfere with the function of the extremities. No distal metastasis was seen.

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