Cavernous hemangioma of the left forearm

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Abstract

Cavernous hemangiomas are benign hamartomatous proliferation of endothelial tissues. They can arise virtually anywhere in the body and do not contain tissues native to the organ or structure in which they are located. Contrary to what is seen in capillary hemangiomas and the cavernous cutaneous variety, intramuscular hemangiomas almost always never regress. We report a case of intramuscular cavernous hemangioma in an 18 year-old male Nigerian with swelling in the left forearm of 9 years duration. The swelling progressively increased in size and nearly involved the entire forearm with occasional pain and no preceding history of trauma. Pre-operative plain radiographs showed an ill-defined soft tissue swelling with multiple calcifications or phleboliths. Surgical excision with ligation of feeding vessels was done and histology confirmed the diagnosis with free margin of excision. Post-operative clinical improvement was marked.

Introduction

Hemangiomas are the most common benign soft tissue tumors of infancy and childhood and occur in 5-10% of infants. They can be of cavernous or capillary variety and the cavernous type can either be cutaneous or deep. Deep cavernous hemangioma is a rare entity and can be intramuscular, bony or visceral; in an increasing order of frequency of occurrence. However, despite the frequency of these tumors, their pathogenesis is not completely understood and the best approach to their management remains controversial.1,2

Less than 1% of all hemangiomas of the body occur in the muscles. Intramuscular hemangiomas are non metastasizing benign hamartomatous congenital neoplasms, that after remaining unrecognized for long periods, may suddenly start to grow in the second and third decades of life.3 Approximately 55% of these tumors are present at birth and the remainder develops in the first weeks of life.4

Non-operative treatment options for intra-muscular hemangioma include cryotherapy, radiotherapy, laser therapy, subcutaneous injection of recombinant interferon alpha (IFN-α), injection of sclerosing agents and corticosteroids. Other therapies discussed in the literature include duplex-guided sclerotherapy with foam injection and angiographic embolization.

Surgery is often indicated in the treatment of residual cosmetic deformities and it is a reasonable option in some cases, such as pedunculated hemangiomas that are almost certain to result in residual abnormalities and hemangiomas that are life threatening or impair function and for which non-operative therapy is not effective or well tolerated. Recurrence is normally caused by incomplete excision.

Case Report

An 18-year-old boy presented with a 9 year history of left forearm swelling. The swelling was initially small but progressively increased in size to almost involve the entire left forearm. No similar swelling was noticed in other parts of the body. He developed clawing of the fingers of the left hand 2 years prior to presentation. There was associated sharp pain that was intermittent and of sudden onset with no known aggravating or relieving factors. No preceding history of trauma was obtained, neither was there weight loss, malaise, bone pains, jaundice, abdominal or chest pain or cough nor family history of similar pathology.

Examination findings at presentation were a young boy with a diffuse swelling on the volar aspect of the left forearm, extending from the mid forearm to the wrist, measuring 7×13 cm, smooth surface and soft with poorly defined edges but tender on deep palpation and a positive Tinel’s sign over the median nerve (Figure 1A).

Plain radiographs of the left forearm showed soft tissue swelling with multiple calcifications and no involvement of the forearm bones (Figure 1B). Excision biopsy with ligation of the feeding vessels was done and mass was sent for histology. Intra-operative findings were that of a nodular mass within the common belly of the long flexor group of forearm muscles with the short flexor tendon embedded within it. Some calcific deposits were also seen in the mass (Figure 2).

Histology came out as cavernous hemangioma (Figure 3) and post-operative period was essentially uneventful. He has been keeping follow-up clinic appointments and regular physiotherapy sessions to improve the function of the digits.

Discussion

Hemangiomas are hamartomatous proliferation of endothelial tissues.2 They are the most common of the angiomatous lesions and account for 7% of all benign soft tissue tumors in the general population. Hemangiomas are the most frequently diagnosed soft tissue tumors in children and can be classified clinically as capillary or cavernous. Cavernous hemangiomas are less common than the capillary variety and are composed of large dilated vascular channels, with poorly defined edges and involve deep structures like viscera, bone and muscles. Intramuscular cavernous hemangioma is a rare entity, literatures are few and mostly case reports.

Unlike other benign tumors, the life cycle of a hemangioma is different in that there is a phase of rapid proliferation that is followed by spontaneous involution. This is especially true for the capillary and cutaneous cavernous varieties while the deep cavernous type almost always never regress and malignant transformation is extremely rare.

Hitherto, there has been a poor understanding of intramuscular cavernous hemangiomas and vascular malformations as separate entities as many cases of reported intramuscular hemangiomas were in actual fact skeletal mus-
cle vascular malformations. Mulliken and Glowacki in their study, elucidated vascular malformations and hemangiomas by emphasizing that hemangiomas are characterized by hyperplastic endothelium or increased mitotic activity, make their appearance during the late fetal or early neonatal life, grow rapidly and usually undergo regression. Vascular malformations on the contrary, never regress, are present at birth and lined by quiescent endothelium with walls deficient in smooth muscle.

Also, immunohistochemistry shows that hemangiomas express angiogenic proteins during the phase of proliferation, specifically, vascular endothelial growth factor, basic fibroblast growth factor, type IV collagenase and urokinase; both enzymes involved in remodeling extracellular matrix. Conversely, this up-regulation of angiogenic factors or matrix enzymes is absent in vascular malformations. GLUT-1 (an erythrocyte-type glucose transporter protein) specific for hemangiomas of infancy, is not expressed in vascular malformations. Hein and co-workers, in a review of 176 cases of vascular malformations, including three lesions with capillary structures, discovered negative staining of all specimen to GLUT-1. They concluded that there is a benign vascular tumor of muscle with prominent capillary structure that is histologically similar to infantile hemangioma but does not express GLUT-1 surface protein or involute.

Hemangiomas are in most cases, asymptomatic, however, presentation is usually due to pain, the presence of a mass or soft tissue swelling, subcutaneous discolorations or cosmetic concerns (by parents or patient) and less frequently, neurologic symptoms secondary to impingement of a nerve bundle.

Clinical examination, usually reveals a soft mass with varied shape and may be fluctuant, with or without differential warmth, may be tender with ill-defined edges, may empty and a thrill may be felt or a bruit heard over the mass.

Radiological evaluation include plain radiographs which would show a soft tissue swelling without bony involvement and calcifications may be evident. Magnetic resonance imaging shows densely lobulated mass with mixed intensities. Computed tomography scan shows a homogenous mass with large feeding vessels with intense and persistent enhancement after administration of contrast. Ultrasonography with Doppler studies is a cost effective and non-invasive technique and demonstrates the high flow pattern that is characteristic of hemangiomas thus differentiating them from the low flow pattern of vascular malformations.

A clinical photograph of the lesion is also taken and other ancillary investigations done to optimize the patients for surgery.

Treatment of hemangiomas is initially conservative which entails observation and periodic evaluation. Modalities like cryotherapy, radiotherapy, laser therapy, subcutaneous injection of recombinant interferon alpha (IFN-α), injection of sclerosants and intravenous corticosteroids, duplex-guided sclerotherapy with foam injection and angiographic embolization have all been tried with variable results.

Surgical excision with ligation of feeding vessels is the optimal management for intramuscular hemangiomas in order to prevent recurrence. The indication for surgery include; progressively increasing mass or swelling, pain and neurologic compromise from pressure effect and significant cosmetic concerns. A structured post-operative rehabilitation protocol is beneficial to improve limb function and prevent joint stiffness.
Conclusions

Prognosis after complete surgical excision and ligation of feeding vessels as a treatment modality for intramuscular cavernous hemangioma is usually very good and malignant transformation is rare, however, incomplete excision is attended by recurrence.

References