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Intramuscular Hemangioma Complicated by a Volkmann's Like Contracture of the Forearm Muscles

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Intramuscular hemangiomas are rare tumors constituting less than 1% of all hemangiomas. The clinical picture is usually unlike a conventional vascular tumor. Pre-operative diagnosis is very difficult and most often, the condition is recognized only during surgery or after histopathological examination. This is a report of one such rare tumor, which presented as a painful mass in the Flexor Digitorum Superficialis. It was accompanied by the hitherto unreported complication of a Volkmann's like contracture of the deeper forearm muscles. The peculiar feature of this tumor are highlighted and are discussed with a review of relevant literature.

Keywords: Hemangioma, Volkmann's contracture.

Intramuscular hemangiomas are rare tumors constituting a mere 0.8% of all hemangiomas(1,2). They deserve attention

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not only because of their rarity but also because of their invariably confusing clinical presentation as well as intriguing etiopathogenesis.

Case Report

A 12-year-old girl presented with a painful swelling in the left forearm. There was history of significant trauma to the left forearm, at the age of 6 years. Following the injury, she was initially taken to a traditional bonesetter who splinted and immobilized the limb for 4 weeks. The child was asymptomatic and had no movement deficits on splint removal. Over the next few years, she gradually developed a painful swelling in the left forearm and began finding it increasingly difficult to use the affected hand. The swelling was diffuse and slow growing and was not associated with any constitutional symptoms like fever or weight loss.

Local examination suggested a diffuse intramuscular mass in the flexor compartment. It was warm, tender and boggy. The swelling was neither compressible nor pulsatile and no bruit could be heard on auscultation. The wrist and fingers were held in flexion and passive extension was grossly limited and very painful. Active flexion of the fingers and wrist was also restricted and painful. Passive flexion of the wrist permitted some extension of the fingers at the interphalangeal joints (*i.e.*, a positive Volkmann's sign). However, as the patient

had severe pain on both active extension and flexion, the sign was considered false positive and not much significance was attached to it at this stage. A clinical diagnosis of chronic flexor tenosynovitis was made. Radiography revealed a soft tissue mass with multiple calcific spots and the following differential diagnoses were considered: tuber-culous flexor tenosynovitis (with calcified melon seed bodies) or an organized hematoma (with metastatic calcification).

Surgery revealed a multiloculated purple-red mass that had completely involved the flexor digitorum superficialis (FDS). Further, the mass had invaded into the substance of the median nerve at the wrist and had enveloped all other muscles, tendons and neurovascular structures in the distal three-fourths of the forearm. The mass was not compressible even intraoperatively and no feeder vessel could be demonstrated. The median nerve required an internal neurolysis as the mass had insinuated between its fascicles. It was possible to dissect the mass away from all other musculo-tendinous and neurovascular structures in the forearm except the FDS, whose belly was excised along with the tumour. As there was no reason at this stage to suspect any muscle necrosis, no specific attempt was made to explore the flexor profundus or flexor polices longus (FPL) but the muscle looked healthy on gross appearance. Post excision, the remaining long flexor tendons (*i.e.*, FDP and FPL) were found contracted with a persisting Volkmann's sign (*Fig. 1*).

Histopathology of the excised mass revealed muscle tissue with a well-defined lesion comprising closely packed thin walled capillaries, areas of hemorrhage and foci of calcification. The final diagnosis was an intramuscular capillary haemangioma.

Electrophysiological studies of the ulnar

and median nerves confirmed no damage to innervation during dissection of the tumor. Post-operatively, the patient underwent intensive physiotherapy for stretching of the contracted tendons and for restoration of finger flexor power as she was left only with the flexor profundus. Full functional restoration was achieved about 8 months after surgery.

Discussion

The exact cause of the intramuscular hemangioma has been something of an enigma. Some authors feel that it is a congenital neoplasm while others have reported a previous history of trauma(3). Allen and Enzinger(2) have reported trauma to be the cause in 5% patients in their series. A definite history of trauma was available even in the



Fig. 1. Early Postoperative picture showing Volkman's sign.

present case.

Virtually, every report of the intramuscular hemangioma in literature stresses its atypical clinical presentation. It is usually seen as a slow growing mass that may or may not be painful. Features characteristic of vascular tumors like pulsation, thrill or bruit are generally absent and hence, the condition is rarely diagnosed pre-operatively(2). Fergusson(4) reports that the intramuscular haemangioma has been clinically misdiagnosed in more than 90% of cases. Pain is a common symptom, contradictory to that expected in a benign neoplasm. This has been attributed to the peculiar association of abnormal blood vessels with nerve fibers as well as significant levels of Substance P in these tumors(5,6). The current patient presented with all features suggesting an inflammatory process. Most significantly, she had severe pain on direct pressure over the mass as well as on resisted flexion or passive stretching of the fingers. This led to a mistaken clinical diagnosis of flexor tenosynovitis.

Histopathologically, the tumor can be classified into three types, capillary, cavernous and mixed, based on the predominant vascular pattern(2). The present case was of the capillary type. Management of intramuscular hemangioma has evolved over time. Spontaneous resolution has not been known to occur(3). Radiotherapy, cryotherapy and embolization have been found to be largely ineffective(3,4). Rogalski *et al.*(5) in their series of 41 patients could not find a feeder vessel large enough for embolization. There was no feeder vessel evident in the present case as well. Total excision, often requiring *en block* resection of the tumor along with the involved muscle is the universally recommended treatment and was the method employed in the current case.

Various complications have been reported in intramuscular hemangiomas, but so far, contracture of adjacent uninvolved muscles has not been reported. The current case appears to display this unique complication. The features of contracture were very much like that seen in Volkmann's ischaemic contracture (VIC) and involved the FDP and FPL. It is tempting to speculate whether the contracture occurred as a result of chronic muscle ischemia. Factors which contribute to this line of thought are: (i) The Volkmann's sign persisted even after excision of the tumor; (ii) the tumour had invaded the FDS only; (iii) more superficial muscles like the wrist flexors or pronator teres were not involved (iv) The flexor profundus and the flexor pollicis longus were contracted. These are typically the muscles to be affected in the localized type of VIC. The contracted muscles were enveloped but not infiltrated by tumor tissue and there was a clear plane of dissection between the tumor and enveloped muscles. This suggests a cause of contracture other than tumor induced necrosis or fibrosis(7). Robinson *et al.* have reported a Substance-P and CGRP, which are found in intramuscular haemangiomas, are known to divert blood away from muscle fibers into the surrounding connective tissue. It is debatable whether this is confined only to the infiltrated muscle or can exert influence on surrounding muscles, thus contributing to ischaemia and fibrosis. Rogalski *et al.*(5) have reported compartment syndrome following Intramuscular hemangioma in the forearm. Other authors have reported compartment syndrome following other tumors or mass lesions in closed osteofascial compartments(8,9). Anderson and Chandi(10) have reported contracture of the flexor profundus following cysticercosis. Though cysticercus infiltrates of muscles are usually asymptomatic, the excised muscle mass was pale and fibrotic. They speculated that there might have

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been a segmental vascular compromise of the muscle, which led to fibrosis and contracture. The presence of a steadily grow-ing mass in closed osteofacial compartment of the forearm in this case, had in all probability caused a chronic compartment syndrome. Whether this led to ischemia is worthy of consideration. Following surgery, the patient underwent an intensive physiotherapy program comprising stretching and FDP and FPL strengthening exercise. By 32 weeks post-excision, the patient had recovered full range of movement as well as grip strength comparable to the opposite hand.

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