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Case Report

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AN INTRAMUSCULAR HAEMANGIOMA ASSOCIATED WITH MULTIPLE CAFÉ AU LAIT SPOTS: A CASE REPORT

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Abstract

This report details the diagnosis and management of a 13-years-old girl with an intramuscular haemangioma on the left side of the chest wall associated with multiple cafe au lait spots (CALS). All previous cases of intramuscular haemangioma are present in the literature however none of them seems to be associated with multiple CALS. Therefore, it is difficult to ascertain whether such an association, in an otherwise healthy individual, is a new genetic syndrome or a mere coincidental finding.

Case Report

A 13 year old girl was admitted to Mubark Al-Kabeer Hospital in Kuwait in July 2002 with swelling on the left side of the chest. The swelling was of 8 years duration with a progressive enlargement over the last year. There was no history of trauma. On examination the swelling was situated at the level of fifth to seventh ribs on the anterolateral aspect of the left side of the chest. It measured about 6x8 cm but with no skin attachment. There was a thrill over the mass with bruit heard on auscultation. General examination revealed multiple cafe au lait spots (Fig 1) and no other swellings could be detected .The patient otherwise was normal.

A radiograph of the chest and full blood count were found to be normal. A chest magnetic resonance imaging (MRI) axial and coronal, (Fig2) revealed a 9x3 cm fusiform mass at the left side of the chest. The mass had signal void areas in keeping with vascular channels. The vascular lesion

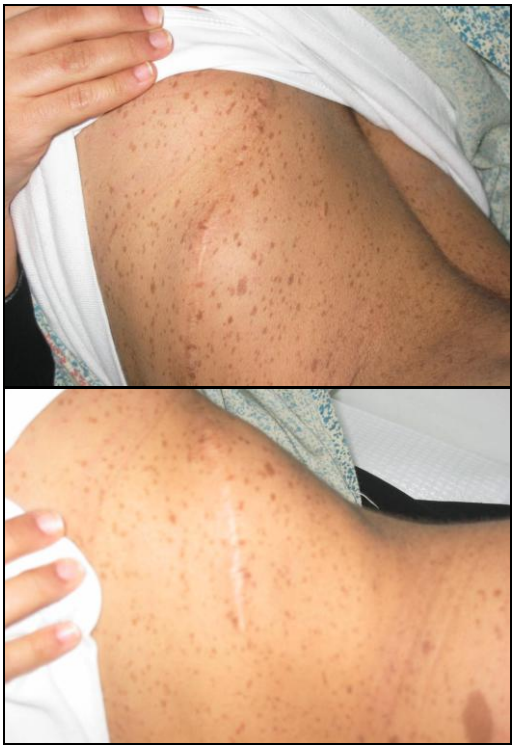

Fig 1 : Multiple café au lait spots

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appeared in the left serratus anterior and latismus dorsi giving it an asymmetric bulge.

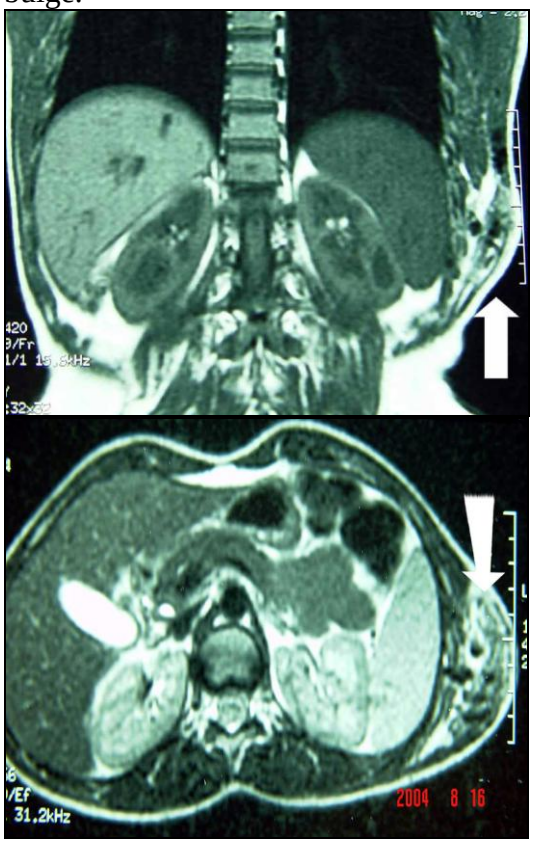

Fig 2: Chest magnetic resonance imaging (MRI) axial and coronal (MRI revealed a 9x3 cm fusiform mass at the left side of the chest. The mass had signal void areas in keeping with vascular channels. The vascular lesion appeared in the left serratus anterior and latismus dorsi giving it an asymmetric bulge).

Arch and selective angiograms (Fig3) revealed enlarged left lateral thoracic artery and enlarged tortuous 7-10 left intercostals arteries with 10x4x6 cm artetiovevous malformation in the lateral aspect of left thoracic wall. Arterial supply for this malformation was coming mostly from 8, 9; and 10 left intercostals arteries, lateral thoracic, intercostobronchial trunk, and internal thoracic arteries. Venous drainage was realized mostly to lateral thoracic and left subclavian veins. Preoperative embolization could not be done because of the multiplicity of the feeding vessels. Skin biopsy showed increased melanin content in both melanocytes and basal keratinocytes without melanocytic proliferation.

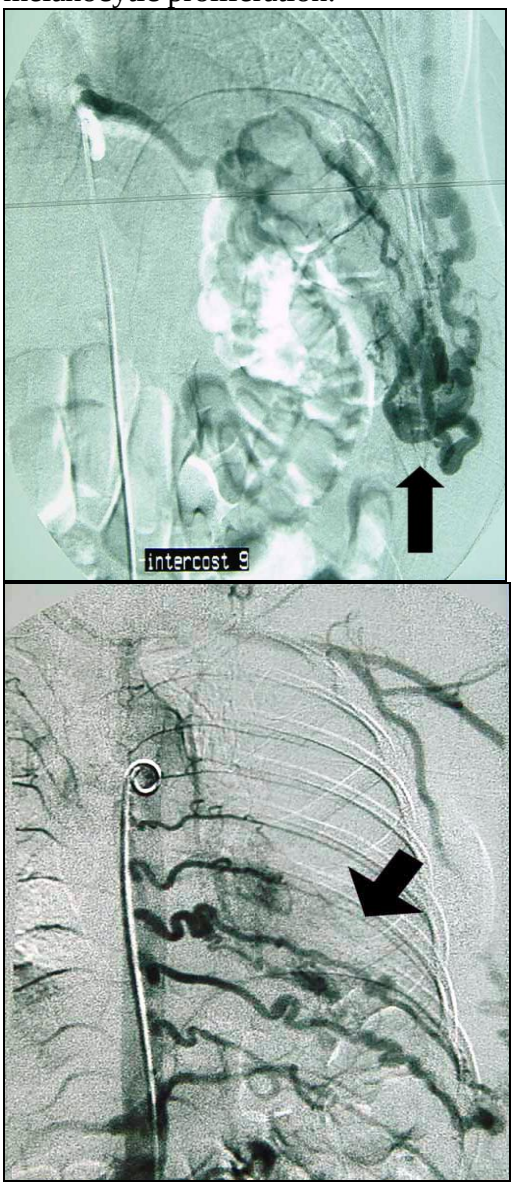

Fig 3: Selective angiograms (Enlarged left lateral thoracic artery and enlarged tortuous 7-10 left intercostals arteries with 10x4x6 cm artetiovevous malformation in the lateral aspect of left thoracic wall. Arterial supply for this malformation was coming mostly from 8, 9; and 10 left intercostals arteries, lateral thoracic, intercostobronchial trunk, and internal thoracic arteries.).

Surgical resection of the swelling with the involved part of the muscles down to and including the periosteum of the

underlying ribs was performed. The intraoperative bleeding was minimal. The microscopic appearance confirmed the diagnosis of intramuscular cavernous haemangioma.

Discussion

Intramuscular hemangiomas are basically rare benign tumours, making up 0.7% of all reported hemangiomas[1]. Despite their wide anatomic distribution, the quadriceps muscle seems to be the most distinguished location compromising about 19% of all cases[2].

There are two main theories attributed to the aetiology of intramuscular haemangioma. One being the theory of congenital origin which is mostly accepted because of the larger percentage of hemangiomas recorded in young patients. This is also supported by a review of 1363 haemangiomas of which 73% were evident at birth[1,3]. The second theory being that of trauma related history. In 1957 a study done by Scotts showed that 17% of the cases were associated with a history of trauma at the site of the tumour[2]. Winchester in 1992, explained further that the role of the trauma may be to augment a pre-existing condition or to produce small areas of granulation tissues and enlargement with continued disturbance[4].

Patients with intramuscular hemangiomas may have soft-tissue complaints, such as pain and swelling which is present for years[3]. These hemangiomas are difficult to pre-diagnose due to their infrequent presence and variable depths in the muscle. Despite the less frequent presence of thrills or bruits, they do provide us an important insight into the diagnosis, as was useful in our case. La Sorte in 1960, reports a bruit in 1.8% and pulsation in 3% of cases[5].

The appearance of these intramuscular hemangiomas can lead to suspicion of malignancy, which can only be definitely confirmed by blood aspiration or histological study of the biopsy (needle or excisional)[6]. Angiography and magnetic resonance imaging (MRI) may also be an indispensable tool for their diagnosis. The latter has become the method of choice because of its non invasive nature which usually shows high signal intensity on both T1 and T2 weighted images, serpiginous pattern, and associated focal muscle atrophy[7].

Multiple CALS have been reported in several syndromes including neurofibromatosis[8], Albright syndrome[9], Banayan-Riley-Ruvalacaba syndrome[10], Watson syndrome[11], Silver Russel syndrome[12], and Westerhof syndrome[13]. Of these syndromes, multiple CALS is mostly associated with neurofibromatosis. The findings in our case cannot be associated with any of these syndromes or any other published syndromes.

According to Rao in 1992, an angioma is a true neoplastic process involving vascular or lymphatic tissues[14]. Complete wide surgical excision is attempted, unless cosmetic or functional considerations arise, to prevent recurrence[4,15]. In a review of 89 cases of skeletal haemangioma done by Allen in 1972, 18% recurred locally. However, each patient is treated individually depending on the exact location and accessibility of the tumour found[16]. Cohen in 1993 has advocated preoperative embolization of intramuscular haemangiomas to minimize blood loss and facilitate a more meticulous dissection[17]. The multiplicity of feeding arteries in our case was not in favour of this technique. On a two year follow up, there has been no recurrence in our patient and she is enjoying a pain free life. It is concluded that although CALS usually indicate neurofibromatosis, yet one must not exclude the possibility of an intramuscular haemangioma to avoid unexpected intraoperative bleeding.

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