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CASE REPORT

Cavernous Lymphangioma of the Digits: A Rare Cause of Macrodactyly

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ABSTRAK

Limfangioma kavernous adalah kecacatan kongenital sistem limfatik yang menyebabkan sinus limfatik berkembang yang melibatkan kulit dan subkutaneus tisu. Ini adalah satu kes yang menarik tentang macrodactyly dystrophic jari cincin dan jari kelingking dalam seorang bayi perempuan berusia 18 bulan yang menjadi limfangioma kavernous. Tumor ini, walaupun jarang terjadi pada ekstrimiti, mesti dibezakan dari luka vaskular kongenital yang lain, termasuk kecacatan arteriovenous dan hemangiomas. Diagnosis adalah berdasarkan analisis histopatologi mengenai tisu yang dikeluarkan. Pembedahan biasanya diperlukan untuk hasil fungsi dan kosmetik yang memuaskan.

Kata Kunci: kecacatan kongenital, limfangioma kavernous, macrodactyly

ABSTRACT

Cavernous lymphangioma is a congenital malformation of lymphatic system causing dilated lymphatic sinuses that involve the skin and subcutaneous tissues. This was an interesting case of dystrophic macrodactyly of the left ring and little finger in a 18-month-old girl who presented with swollen and sausage like fingers deformity which turned out to be an isolated cavernous lymphangioma. This tumor, although rare to occur in the extremeties, must be differentiated from other congenital vascular lesions of the hand that include arteriovenous malformations and hemangiomas. Diagnosis should be solely based on histopathological analysis of the excised tissue mass. Surgical excision is usually necessary for satisfactory functional and cosmetic outcome.

Keywords: cavernous lymphangioma, congenital malformation, macrodactyly

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INTRODUCTION

Congenital vascular malformation (CVM) is the most common form of lymphatic malformation (LM) which can be classified by new Hamburgh classification into truncular form and extratruncular form of lymphatic malformation (Marler et al. 2005). Both are consequences of a developmental arrest at the different stages of lymphangiogenesis as a result of defective genes. The extratruncular (ET) form of LM is also known as the cystic or cavernous lymphangioma.

Vascular tumours account to 7% of the total tumours occurring in the hand and forearm with cavernous lymphangiomas, accounting for 1% (Takka et al. 2004). With 50 to 65% of these tumors arising at birth, nearly 90% being detected by second year of life (Murase et al. 1992). Cavernous lymphangioma of the hand and fingers as a cause of macrodactyly are extremely rare in occurrence and the treatment of the affected limb is difficult because of the delicate structure of the musculoskeletal system of the hand.

CASE REPORT

An 18-month-old girl presented with gradually enlarging bulbous swelling of her left ring and little finger. These swellings were noted at birth and subsequently increasing in size, dysfiguring the ring and little finger. Examination of the left hand revealed swollen and sausage like deformity of her ring and little fingers (Figure 1). It was soft, non-tender with multilobulated swellings extending over the dorsum of the hand until the distal part of interphalangeal joint. These swellings were not tender, non-transilluminant and non-compresssible with no palpable thrill.

Radiograph of the left hand showed a large soft tissue shadow around the ring and little finger from the metacarpal bones of ring and little finger extending untill the distal phalanx. Magnetic Resonance Imaging of the left hand showed (Figure 2) a large subcutaneous tissue mass of the ring and little fingers with no definite margins. It was described as benign in character, being eccentric with slight deviation of the traversing digital vessels due to the mass effect.

Parents were counselled for debulking surgery which gave histopathological diagnosis and limit the progression of the swelling and resultant deformity. The first surgery involved debulking of the ring finger. Operative findings revealed pale, fibrotic and dense subcutaneous tissue with minimal fat which was sent for histopathological examination.

The histopathological report revealed microscopic findings of

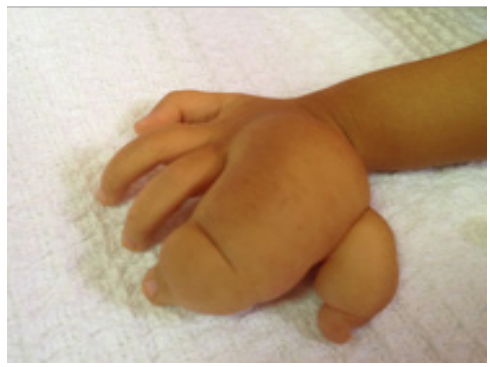

Figure 1: Photograph showing sausage like swelling to patient ring and little finger

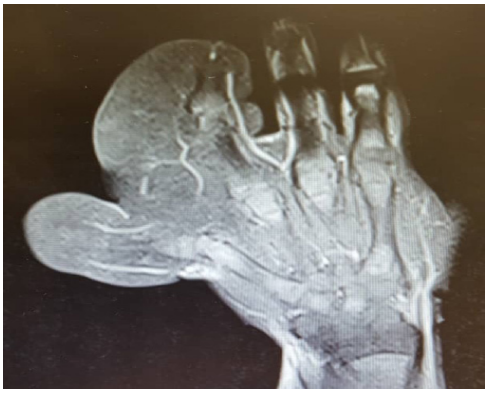

Figure 2: MRI showing a large, homogenous, eccentric subcutaneous tissue mass over the ring and little fingers which was hypointense in T1 and hyperintense in T2 sequence indicating presence of fat.

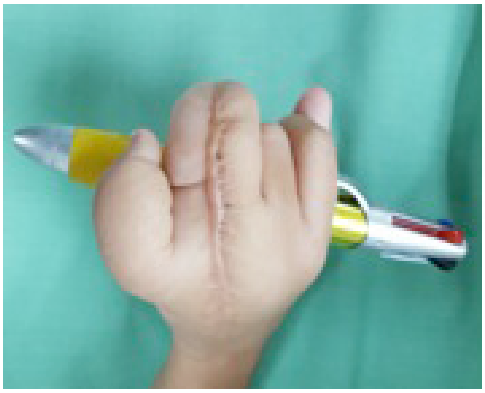

Figure 3: Improved grip and size of digits after first debulking surgery

dense fibrocollagenous tissue with few small lobules of mature adipose tissue. Variable sizes of thin walled lymphatic vessels, lined by endothelium with uniform nuclei was seen. These findings guided us to the final diagnosis of lymphangioma. On follow-up, her surgical wound healed well and has improvement in hand function with reduction in size of her ring finger (Figure 3).

Second stage of debulking her little finger was then performed. Upon incision over the dorsal aspect of the little finger, similar findings as the first debulking were noted. Exploration over the volar aspect of the digit revealed clear fluid filled cystic tissue. (Figure 4). Despite the variation in tissue consistency, the second histological report was similarly reported as lymphangioma. Post-operative follow-up showed both the digital swellings had markedly reduced and and she had regained more hand function. Her parents were happy with the results and was informed about the risk of recurrence.

DISCUSSION

Lymphangioma can appear at birth or shortly thereafter, and present as a slow growing mass anywhere in the body but commonly in the head and neck region (Bhayya et al. 2015). Lymphangioma of the upper extremity is rare and can occur at the dorsum of the hand, due to the rich lymphatic channels (Murase et al. 1992). In our case, the involvement of the fingers were extremely rare. Diagnosis is

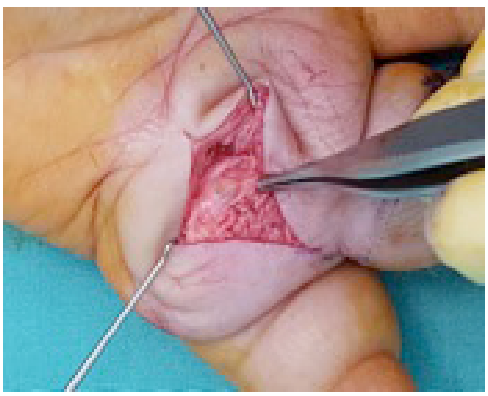

Figure 4 : Volar view of the little finger showing less dense tissue with fluid filled cysts

usually clinical, with pain being a classical exhibit in bigger patients along with evidence of functional limitation in the deformed looking bulky digits. Pain can occur in some tumors due to nerve compression by the mass, which grows in a confined space. Other differential diagnosis can be arteriovenous malformation, cutaneous lipoma or hemangiomas.

Treatment of lymphangioma can be challenging, which includes surgical excision, radiation therapy, aspiration, and sclerotherapy. Radiation treatment is no longer been used due to unable completely destroy the lymphatic malformation and possibility damaging the growth of local structures, and the potential for inducing a malignant lymphangiosarcoma (Upton et al. 1999). Cauterization and laser therapy are not helpful. Aspiration therapy is only recommended to relieve local compressive symptoms as an emergency measure (Jacobs et al. 2010). Sclerotherapy can shrink the tumour but not as curative treatment. Surgical treatment may still be required to remove the lesion.

Complete surgical resection is still the recommended treatment for lymphangiomas (Wall et al. 2018). But local recurrences are common as adequate excision of lymphangiomas can be difficult and, at times unfeasible (Blair et al. 1983). This tumor has an 'invasive' nature with lack of demarkation of a clear border between superficial cutaneous tissue and the contiguous muscle and fascial layers causing insufficient removal of the involved tissue and resultant post-operative fluid leakage and recurrences. Overlying skin, surrounding adipose tissue and paratenon should be removed en bloc in order to prevent recurrence (Takka et al. 2004).

In this case, the primary aim of the debulking surgery was to improve functional status and dexterity of the affected digits were achieved to the patient's and parents' satisfaction. However, parents were carefully prompted on the invasiveness and poor demarkation of lyphangiomas which resulted in fairly high rates of recurrence. Individual compression stockings of digits and sclerosant injections were the alternative resort in case of future recurrence and deterioration in hand function. Further or repeated debulking procedures may have guarded outcome due to scar fibrosis, adhesion and joint stiffness.

CONCLUSION

Functional limitation of the enlarged and cosmetically disfigured fingers associated with pain makes surgical resection the preferred option. Parents should be thoroughly informed regarding the post-operative risk which includes recurrence, scar adhesion and multiple debulking with large tumour or multiple finger involvement.

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