

A Case Of Lymphangioma Circumscriptum: Magnetic Resonance Imaging (MRI) in Diagnostic Evaluation

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A 21-year-old male visited our clinic with an erythematous plaque studded with papulovesicles on the right thigh and calf which had been noticed at 3 months after birth. Moving the limb caused intermittent pain. He was diagnosed with lymphangioma circumscriptum which involved muscle layers after checking the skin biopsy and magnetic resonance imaging (MRI). Partial removal of mass resulted in relieving pain on motion.
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Key Words : Lymphangioma circumscriptum, Magnetic resonance imaging (MRI).

Stereotypical lymphangioma circumscriptum may exhibit diffuse swelling of the subcutaneous tissue beneath the pseudovesicular lesions¹. Involvement of the muscle layer is not uncommon, and the depth and extent of involvement cannot be adequately estimated from the cutaneous examination. So magnetic resonance imaging (MRI) may be a useful tool in evaluating patients with lymphangioma circumscriptum.

REPORT OF CASE

A 21-year-old male patient visited our clinic because of painful subcutaneous masses on the right thigh and calf. They developed at 3 months after birth and had not caused any specific problem until the development of intermittent pain during exercise one year ago. A physical examination disclosed well defined 2 × 3 cm erythematous to brownish plaque studded with grouped vesicles and crusts on the right thigh (Fig. 1) and calf. A skin biopsy re-

vealed multiple cystic lesions lined by a single layer of endothelium in the upper dermis (Fig. 2) consistent with lymphangioma circumscriptum. In MRI, huge cystic structures invading the muscle layers were found in the right thigh and calf (Fig. 3). He was transferred to the orthopedic surgery and had a partial removal of the tumor mass. Post operative histopathologic examination showed multiple cystic vascular channels in the muscle layer (Fig. 4). The pain was relieved after the operation.

DISCUSSION

Lymphangioma circumscriptum is an uncommon hamartomatous malformation. Clusters of translucent vesicles (frog spawn), occasionally blood tinged, and with superimposed hyperkeratosis are characteristically present on a circumscribed area of the skin¹.

Lymphangioma circumscriptum lesions are often apparent at birth or develop by the age 5 years. Although lymphangioma circumscriptum has been described as first appearing in adulthood, the lesion occurs less commonly in older age groups^{1,2}. The sites mainly involved are the neck, axillae, glands, abdomen and thigh, but there were some reports that it developed in the glans³, scrotum⁴, and oral cavity⁵. In our case, the developing time and predilection site are consistent with typical lymphangioma circumscriptum.

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Fig. 1. 2 × 3cm erythematous to brownish plaque studied with grouped vesicles and crusts in the right thigh and calf

Fig. 3. huge vascular structures invading the muscle layers (MRI findings)

It was postulated that the lesion of lymphangioma circumscriptum consists essentially of a collection of large muscular-coated lymphatic cisterns, lying deep in the subcutaneous plane and communicating via dilated dermal lymphatics with the superficial vesicles. In 1976, however, Whimster⁶ found no histological evidence of communications between the cysts and the adjacent normal lymphatics and suggested for the etiology of the lymphangioma circumscriptum that the

Fig. 2. multiple cystic lesions lined by a single layer of endothelium in the upper dermis (H&E, × 40).

Fig. 4. multiple cystic lesions in the muscular layer ; post-operative histopathologic findings (H&E, × 40).

subcutaneous cysts were sequestered segments of the primitive lymph system. The absence of a connection between the subcutaneous lymphatics and the cysts has been confirmed by lymphography⁷.

The histopathological appearance of lymphangioma circumscriptum is characterized by solitary and grouped dilated cystic spaces in the papillary dermis. The cystic spaces are lined by endothelial cells, and they often contain red blood cells as well as lymphatic fluid. In the deep dermis and subcutaneous fat, dilated lymphatics are also seen, some of which contain thickened muscular walls^{1,8}. In 1970, Peachey et al¹. reviewed 65 cases collected from several centers and divided lymphangioma circumscriptum into two major groups (classic and localized form) and carvenous form in the aspect of histopathologic involvement. In our case, the lesions involve the muscular layers and have large

cystic lesions in the histopathological finding, which is consistent with carvenous type of lymphangioma.

In evaluating lymphangioma circumscriptum, MRI definition of the entire anatomy of the lesion and the extent of involvement may be an invaluable tool. In 1989, Siegels *et al*¹². reported 17 cases of lymphangioma circumscriptum which were imaged with MRI to define the nature, extent, and anatomic relationships of these lesions and suggested that the information obtained with MR imaging can help in providing a preoperative diagnosis, in planning surgical resection, and in defining recurrence.

The search for an effective therapy of lymphangioma circumscriptum has been an elusive one. Superficial destruction of lesions using cryotherapy and cautery has often been followed by a recurrence of lesions⁹. Recent studies using the carbon dioxide laser showed good results, but the deep component of the lymphangiomas was not treated¹⁰. Complete surgical excision is the treatment of choice in the case of deeply involved lymphangioma circumscriptum¹¹.

In our case, we could find the invasion of lymphangioma in muscular layer via MRI findings and choose the surgical removal in treatment which resulted in the relief of pain. Deeply invaded lymphangioma can recur after superficial treatment, so it is necessary to evaluate the extent of invasion via MRI for complete treatment. Future developments in MRI, including dedicated pediatric or skin surface coils, chemical shift imaging, and shorter imaging times, may further improve image quality and enhance its utility in both preoperative assessment of lesion extent and postoperative assessment of residual or recurrent disease¹³.

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