

## A rare case of microcystic and macrocytic lymphangioma in 12-year-old girl

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### ABSTRACT

**Introduction:** Lymphangioma is an idiopathic digression in the lymphatic vessels morphogenesis. It is a lymphatic malformation that involves the lymphatic vessels of the superficial dermis. The lesions were observed since birth for 50% of the cases. Lymphangioma is typically rare of the lymphatic vessels disorder. This case report aims to explain lymphangioma, which is typically rare of the lymphatic vessels disorder.

**Case report:** Our study reported a chief complaint in a 12-year-old girl was painless multiple small blisters on the left thigh and the left inguinal in the past 11 years. Physical examination revealed the groups of lobulated mass consist of erythematous papules, cysts, skin-colored and purplish papules, purplish macule, tense vesicles & reddish-brown crusts. The blisters were not itchy, followed by the slight swelling under the lesion of the left groin. They were skin-colored papules on the abdomen. It was also found that there was a hypertrophic scar on the upper side of the posteromedial side of the left thigh. Histopathology examination showed hyperkeratosis where the keratinocytes proliferation in the areas lined with endothelium. Eosinophilic amorphous mass found inside the lumen. There were hair follicles, sudoriferous glands and sebaceous glands. The proposed diagnosis was lymphangioma (combined microcystic and macrocystic lymphatic malformation) on the abdomen and soft tissue tumor on the left thigh and the buttock. The treatments were carbon dioxide (CO<sub>2</sub>) laser for lymphangioma and surgical excision for soft tissue tumor.

**Conclusion:** Lymphangioma is a rare lymphatic vessel disorder that manifests on the superficial dermis. It is commonly found in children. This disease can be treated by CO<sub>2</sub> laser.

**Keywords:** CO<sub>2</sub> laser, Frogspawn vesicle, Lymphangioma circumscriptum, Soft tissue tumor

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### INTRODUCTION

Lymphangioma is an idiopathic digression in the lymphatic vessels morphogenesis.<sup>1</sup> It is a rare and hamartomatous congenital malformation of the lymphatic system that involves subcutaneous tissues of the skin. The lesions were observed since birth for 50% of the cases. Lymphangioma is divided into two major groups: the abnormal lymph vessel size and the depth. Lymphangioma circumscriptum is mentioned for the superficial vesicles (microcystic). Besides, the more deep-seated group includes cystic hygroma and cavernous lymphangioma (macrocytic).<sup>2-4</sup>

Lymphangioma is a lymphatic vessel malformation of the superficial dermis.<sup>5,6</sup> Lymphangioma, especially the combined microcystic and macrocystic lymphatic malformation, is a rare type of lymphatic system disorder involving the papillary

and subpapillary layers of the dermis.<sup>3,5</sup> Lymphangioma can be located in any anatomic site, but the most common predilection sites are axillary folds, neck, proximal parts of the extremities, shoulders and tongue. Sometimes the lesions are also found in the conjunctiva, eyelids and genital skin of males and females.<sup>5,7</sup> The lesion consists of numerous small vesicle-like lesions, often with a verrucous surface, grouped in a plaque. It is characterized by frogspawn clusters of thin-walled vesicles on the surface of skin covering either clear fluid or blood-tinged fluid. The lesions are asymptomatic.<sup>7-9</sup>

The superficial vesicles are the result of saccular dilatations of superficial lymphatics, secondary to the increased pressure transmitted from the underlying cisterns. The true extent of the lesions can be identified through magnetic resonance imaging.<sup>5,8,9</sup>

The diseases show dilated vascular spaces involving both deeper subcutaneous and superficial dermis tissues associated with the malformed lymphatic vessels. The superficial lymphatic malformation is accompanied by deep lymphatic dilated cisterns with muscular walls in the subcutaneous fat, causes swelling tissue beneath the superficial vesicles. The superficial side consists of flat endothelial cells in a discontinuous layer, dilated lymph vessels, located in the papillary dermis, and the superficial reticular dermis. Sometimes, the lymphatic vessels are arranged in clusters in the papillary dermis resulting the verrucous or the papillated skin surface. The vessels consist of blood or homogeneous eosinophilic proteinaceous lymph and sometimes foamy macrophages. Scattered lymphocytes may be seen between dilated lymphatic vessels in the connective tissue.

The large irregular lymphatic channels can be observed beneath the superficial vessels in the deep reticular dermis and subcutaneous fat in the extensive lesions.<sup>5,10</sup>

Lymphangioma is a localized lymphatic malformation and relatively superficial. It only causes a cosmetic problem that doesn't need any treatment. But if deep lesions appear and persist, they will need excision.<sup>3</sup> Therefore, the study aimed to discuss lymphangioma, which is a rare case of lymphatic vessel disorder.

CASE REPORT

A 12-year-old girl was noticed with multiple small blisters on her left shoulder since 11 years ago. There were also multiple small bumps on her left thigh and left groin since then and spread widely to her abdomen in the past 8 months. Initially, a few small bumps appeared on her posterior left thigh & slowly increased in number and size subsequently in a period of 4 years since she was still 1 year old. Some of them were localized and grouped on the posterior, lateral and medial left thigh.

There was no abnormality of left thigh skin or in the body parts when she was born. In the last 3 years, the multiple small bumps appeared on her left groin and distributed slowly, increased in number and size. Furthermore, some new multiple, tense, painless and not itchy small bumps similar to the lesions of the left thigh appeared on her abdomen in the past 8 months and increased in number 6 months ago. The lesion has persisted since then. The left thigh was slightly stiffened, painless, and swelling 7 years ago. The patient never complained about referred pain to the bone, especially on the upper left thigh while doing activities or resting. No history of inflammation of the lesion. No history of trauma before the lesion appeared. No family history of a similar disease.

Based on physical examination, a mass was found in the upper lateral portion of the left thigh, abdomen and buttock, resulting an asymmetry lesion. The mass of lesion was tense, 5x4 centimeters, ill-defined borders, not mobile nor painful. The enlargement of lymph node was groundnut-size on the left lateral of

inguinal side. It was also not mobile nor painful and no abnormality was found in other regional lymph nodes (Figure 1).

The dermatologic state was plaque located on 1/3 proximal posterior, medial and lateral left thigh, left groin and abdomen especially umbilical region. There were groups of lobulated mass consist of erythematous, skin-colored and purplish papules, cysts, tense vesicles, purplish macule & brownish red crusts. The hypertrophic scar was on upper posteromedial side of the left thigh. Skin-

colored papules on abdomen with no specific arrangement in size consists of milliary and lenticular lesion with the regional distribution.

The skin biopsy was done to identify the lesions of the abdomen and the left thigh. It resulted that the epidermis was hyperkeratosis and the proliferated dermis was found in spaces lined with endothelium, accompanied by the lumen filled by amorphic eosinophilic mass. There were also sebaceous glands, hair follicle and sudoriferous gland. We



Figure 1. Lesion of lymphangioma on abdomen (A) and soft tissue tumor on left thigh (B), buttock (C)

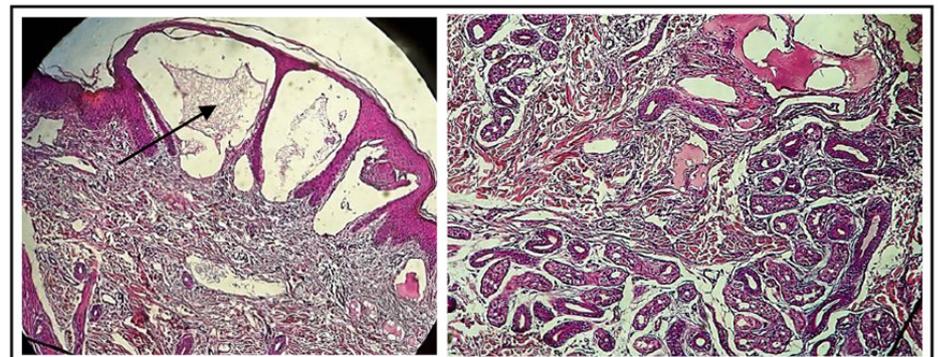


Figure 2. Histopathology of lymphangioma



Figure 3. Abdominal lesion condition one week after laser CO2 treatment

interpreted the working diagnosis as lymphangioma (combined microcystic and macrocystic lymphatic malformation) on the abdomen and soft tissue tumor on the left thigh and buttock. The histopathology of lymphangioma was shown in **Figure 2**. The differential diagnoses were cylindroma and lipoma.

The patient was consulted to several departments. Pediatric Department reported that there were no abnormalities systemically. Pediatric Surgery Department suggested that the patient needed a left pelvis X-ray, CO<sub>2</sub> laser for lymphangiomas and surgery treatment (wide excision) for soft tissue tumor. The left pelvis X-ray showed no abnormalities there and no destructions on the left side of femoral bone. Based on the clinical findings and theoretical background of the disease, the prognosis of lymphangioma and soft tissue tumor on this patient were *bonam for quo ad vitam, dubia ad malam for quo ad sanationam, dubia ad malam for quo ad cosmeticum and dubia ad malam for quo ad functionam*.

In terms of treatment, we decided to do a CO<sub>2</sub> laser for the lesion on the abdomen and perform surgical excision for the lesion on the left thigh and the buttock. We educated the family that the disease represents benign lymphatic malformations and not premalignant lesions. The risk of recurrence remained high; hence, the treatments were explained carefully.

A week after treatment with laser CO<sub>2</sub> from the abdomen, the efflorescence showed multiple papules with the same skin color & erythematous papules, which has a black crust on top of it on the abdomen area (**Figure 3**). There was redness, tingling, stinging, and no pruritic. The lesions on the upper left thigh, left groin still appeared & no changes compared before. We were planning for surgical excision from the pediatric surgery department.

## DISCUSSION

A lymphangioma represents a congenital proliferation of lymphatic vessels. The cases of lymphangioma of the skin and subcutaneous tissue are rare. There were approximately 26 % of benign vascular tumors and 4 % of vascular tumors in

children. The patient with lymphangioma (combined microcystic and macrocystic) seems to be a rare case.<sup>11,12</sup> In our department, there were no reports about lymphangioma in recent years.

The diagnosis of lymphangioma in this patient was based on clinical findings and histopathology examination, resulting in the left thigh, left groin and umbilical region with groups of lobulated mass consisting of cysts erythematous papules, skin-colored papules, purplish papules, tense vesicles, purplish macule & red-brownish crusts. The hypertrophic scar on the upper posteromedial portion of left thigh. The skin-colored papules were found on the abdomen and hypertrophic scars were found on the posterior upper left thigh. The mass on the upper lateral portion of the left thigh was tense, 5x4 cm, ill-defined borders, not mobile nor painful, and the enlargement on the left lateral inguinal lymph node was neither mobile nor painful. The clinical finding of this lesion was mimicking cylindroma. The lesions of cylindroma consist of multiple papules, nodules, or variously-sized tumors. It has tendency to confluence. Commonly found in women and often familial. The most common location is on the scalp & sometimes on the face or trunk. The histopathology examination showed a jigsaw puzzle-like pattern which is circumscribed dermal and/or subcutaneous lesion composed of irregularly-shaped tumor islands and cords of basaloid cells in lipoma, overlying skin with no lesions or changes.<sup>1</sup> The result of histopathology examination showed hyperkeratosis with the proliferated dermis found in spaces that are lined with endothelium with the filled lumen of amorphous and eosinophilic mass. There were sebaceous glands, hair follicles and sudoriferous glands got affected. The disease can be further complicated due to excessive drainage and recurrent cellulitis. There was no history of infection including cellulitis on the lesion in this patient.

The first-line therapy is surgical excision, although other modalities, such as sclerotherapy and laser therapy, have also been reported successful in treating the patients.<sup>12-14</sup> We planned this patient to undergo CO<sub>2</sub> laser and surgical excision. Indications for treating this disease

are cosmetic issues and prevention of complications, such as cellulitis. However, lymphangioma has a tendency to recur.

The previous research showed that three women with two vulvar lymphangioma and vulvar lymphangiectasia were treated with CO<sub>2</sub> laser. All of them improved in symptoms by the mean follow-up time 22 months. Localized persistence and focal recurrence were noted in two lymphangioma patients.<sup>13</sup> A study reported two children with congenital giant cystic lymphangioma were diagnosed through histological examination of the surgical specimen. Early treatment is needed to cease the disease's progress by considering surgery or sclerotherapy based on indications accordingly.<sup>14</sup> Another study reported a newborn with a huge congenital lymphangioma, presented as a cervical swelling on the neck, was diagnosed by ultrasound during the intrauterine period. The patient was treated by surgical excision. The disease recurrence happened in the upcoming 9 months.<sup>15</sup>

## CONCLUSION

Lymphangioma is a rare case, especially the combined microcystic and macrocystic lymphatic malformation. This is an idiopathic digression in the lymphatic vessels morphogenesis that usually occurs during infancy or at birth. The first-line treatment is surgical excision, followed by a carbon dioxide (CO<sub>2</sub>) laser.

## CONFLICT OF INTEREST

The authors declare that there is no conflict of interest regarding the manuscript.

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## AUTHOR CONTRIBUTION

All authors are contributed equally to the content of the study

## ETHICAL STATEMENT

The informed consent was declared from the patient's parent regarding the publication in this journal

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