

A Case of Lymphangioma Circumscriptum♦

BİR LENFANGİOMA SİRKUMSKRİPTUM OLGUSU

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SUMMARY

Benign lymphangiomatous disorders of the skin are uncommon. Here in 9-year-old female patient with a large mass in her thigh and buttock is presented and in this subject literature was reviewed.

Key Words: Lymphangioma circumscriptum, Classic type

T Klin J Dermatol 1996, 6:128-130

ÖZET

Cildin iyi huylu lenfangiomatöz hastalıklarına az rastlanır. Burada 9 yaşında bir kız hastada uyluk ve kalçada yer almış büyük kitle şeklindeki lenfangioma olgusu sunuldu ve bu konuyla ilgili literatür gözden geçirildi.

Anahtar Kelimeler: Lenfangioma sirkumskriptum, Klasik tip

T Klin Dermatoloji 1996, 6:128-130

Benign lymphangiomatous disorders of the skin are uncommon (1). Peachey et al. have divided cutaneous lymphangioma into three main group cystic higroma, localized lymphangioma circumscriptum and classic lymphangioma circumscriptum (2). The classic type lymphangioma circumscriptum was first described in 1879 by Fox and Fox under the name lymphangiectoides. Morris first used the term Lymphangioma circumscriptum in 1889 (3). Classic lymphangioma circumscriptum is confined to one region of the body. Common sites of involvement are chest, thigh, buttock or proximal portion of limbs (2,4,5). The perineum, tongue, buccal mucosa, eye lids and conjunctiva are less frequently involved (1,4,6). The lesion of classic lymphangioma circumscriptum may be size of 7 cm or larger (5-8). It is more common in females and usually is present at birth or appear early in life. Lymphangioma circumscriptum characterized of thin walled vesicles usually filled with clear colourless fluid but occasionally discoloured by the presence of altered blood. In these area skin slightly thickened and subcutaneous tissue which may contain palpable cysts. Peachey et al described classic legions involving larger skin areas, which are produced by rhythmically contracting

muscle coated cisterns in the subcutaneous lesions (2,3,5).

CASE REPORT

A 9-year-old female patient presented with a painless soft mass on her thigh and buttock appeared at birth. The mass was 30x40x50 cm in diameter. There were vesicles on the distal part of the lesion. There was not recurrence 6 months after the surgical therapy.

Light microscopic examination showed hyperkeratosis, mild papillomatosis and acanthosis of epidermis. Greatly dilated lymph vessels lined with a single layer of endothelial cells are presented in papillary and deeper zones of the dermis and the subcutaneous fat. Epidermis was thinned over some of the lymph cysts (Figure 1,2). A few vessel were filled with erythrocytes and had valves on their wall (Figure 3).

DISCUSSION

Benign lymphangiomatous disorders divided into primary and secondary lymphangioma that are not differentiate histopathologically from each other (1). Cutaneous lymphangiomas were classified by authors with different terms (2). The classification of these lesions is shown in Table 1.

We used the terms of Peachey classification in our report. Classification of lymphangioma is essential for planning of treatment and prognosis (2). Patients with cutaneous lymphangioma may suffer recurrent episodes of cellulitis, pain, and copious drainage of lymphatic fluid or cosmetic problem and surgical excision may be necessary. The association of lymphangioma circumscriptum

Received: 25.01.1996

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*This case report was presented as a poster in First Clinicopathological Colloquim in Ankara, Turkey (15-17 September 1995)

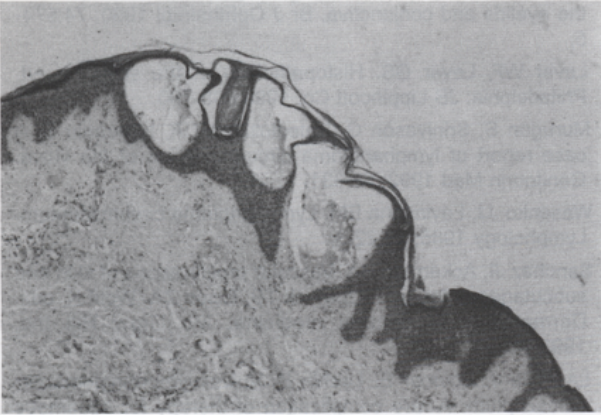


Figure 1. Epidermis was thinned over some of the lymph cyst. HEx80

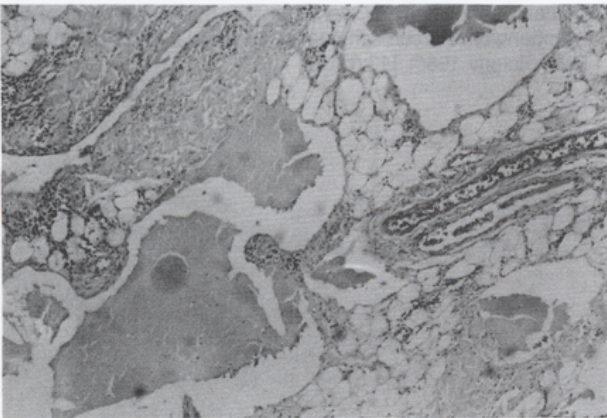


Figure 2. Dilated lymph vessels in the deeper zone of the dermis. HEx200

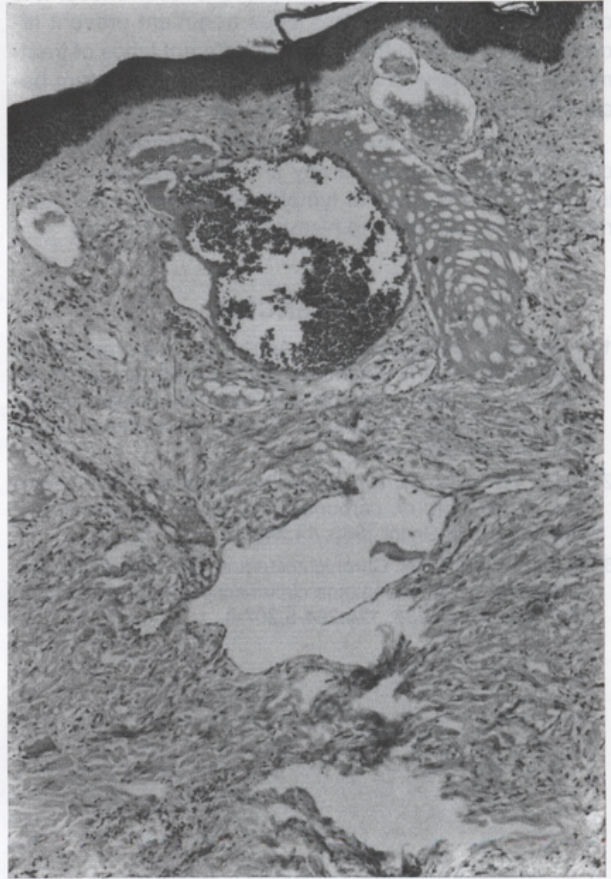


Figure 3. A few vessels were filled with erythrocytes in the superficial zone of the dermis. HEx200

with diffuse edema of a limb is seen rarely (5). However lymphangiosarcoma arising from a previous area of lymphangioma circumscriptum has not been recorded (9). In two cases the site of lymphangioma circumscriptum had

been exposed X irradiation lymphangiosarcoma had been developed.

Surgical excision of skin and underlying subcutaneous tissue is 75-100% curative (4,10). Cautery, cryotherapy, X-ray, CO₂ laser vaporisation have been used in treating surgically in accessible sites to improve cosmesis, and to prevent complications. However none

Table 1. Classification of cutaneous lymphangioma

Classifications	Clinical Features		
	I Deep Cystic Mass (no surface changes)	II Superficial vesicles (no deep mass)	III Combined (deep mass and superficial vesicles)
PEACHEY	Cystic Hygroma	Localized Lymphangioma Circumscriptum (Lymphangioma Simplex) (Capillary Lymphangioma)	Classical Lymphangioma Circumscriptum
FLANAGAN and HELWIG	Lymphangioma Cavernosum	Lymphangioma Circumscriptum	
WEGENER	Cystic Lymphangioma	Simple Lymphangioma	Cavernous Lymphangioma

of these methods of treatment prevent recurrences. It is difficult to evaluate different types treatment prevent recurrences. It is difficult to evaluate different types of treatment in patients with lymphangioma circumscriptum because there have been no comperative trials (2,10,11).

Because of big size and localization of our case had difficulties for surgical therapy and may change to recur in future. This is case of lymphangioma circumscriptum regard to her clinical and histopathological findings.

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Figure 2. A few vessels were filled with erythrocytes in the superficial zone of the dermis. HEx200

Figure 3. Dilated lymph vessels in the deeper zone of the dermis. HEx200

Surgical excision of skin and underlying subcutaneous tissue is 75-100% curative (4,10). Carbon dioxide laser vaporization have been used in treating surgically in accessible sites to improve cosmetic, and to prevent complications. However none

Figure 4. Diffuse edema of a limb is seen rarely (2). However lymphangiomas arising from a previous case of lymphangioma circumscriptum has not been recorded (9). In two cases the site of lymphangioma circumscriptum had

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Table 1. Classification of cutaneous lymphangiomas

Clinical Features		Classifications
I	Deep Cystic Mass (no surface changes)	BRACEY
II	Superficial vesicles (no deep mass)	FLANAGAN and HELWIG
III	Combined (deep mass and superficial vesicles)	WEGENER
	Localized Lymphangioma Circumscriptum (Lymphangioma Simplex) (Capillary Lymphangioma)	
	Lymphangioma Circumscriptum	
	Simple Lymphangioma	
	Cystic Lymphangioma	
	Lymphangioma Cavernosum	
	Classical Lymphangioma Circumscriptum	
	Combined (deep mass and superficial vesicles)	