

Adult Lymphangioma - A Rare Entity: A Report of Two Cases

Erişkin Lenfanjiyomu - Nadir Bir Antite: İki Olgunun Sunumu

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ABSTRACT

Objective: Lymphangiomas are rare congenital malformations commonly seen in children. Its occurrence in adults is uncommon with very scarce reports in the literature. It mainly occurs in infants or children younger than two years of age. We report these cases to emphasize the need to consider cystic lymphangiomas in differential diagnosis of neck masses in adults.

Case Reports: We describe two cases in adults, one a 27-year-old man and the other a 59-year-old male. The former presented with recurrent neck mass for 5 years, and the latter presented with asymptomatic neck mass of a short duration. Both were not associated with any pain, discoloration, discharge or sudden increase in size.

Conclusion: The awareness of occurrence of cystic lymphangioma in adults is important for its proper management which includes complete surgical removal, to prevent recurrence.

Key Words: Lymphangioma, Neck masses, Adults

ÖZ

Amaç: Lenfanjiyomlar çocuklarda görülen nadir konjenital malformasyonlardır. Erişkinlerde nadirdir ve literatürde bildirilmiş az sayıda olgu vardır. Genellikle 2 yaş altındaki çocuk ve infantlarda gözlenir. Erişkinlerde boyun kitlelerinde ayırıcı tanıda kistik lenfanjiyomlara dikkat çekmek için 2 olgu sunulmaktadır.

Olgu Sunumları: Erkek olan olgulardan birisi 27, diğeri 59 yaşındaydı. İlk olgu 5 yıl süren tekrarlayan kitle ile, diğeri kısa süre önce fark edilen kitle ile başvurdu. Her iki olguda da ağrı, renk değişikliği, akıntı ve ani boyut büyümesi tanımlanmıyordu.

Sonuç: Erişkinlerde kistik lenfanjiyomların ayırıcı tanıda akılda tutulması ve nüksün önlenmesi için tam olarak rezeke edilmesi önemlidir.

Anahtar Sözcükler: Lenfanjiyoma, Boyunda kitle, Erişkin

INTRODUCTION

Lymphangioma is a benign congenital malformation seen almost exclusively in children less than two years of age. It is extremely rare in adults, with only about 100 cases reported in literature (1). These are considered to be the result of sequestration of lymphatic tissue that has retained its potential for growth. The three variants which have been described are: 1) Capillary - characterized by small thin walled vascular channels; (2) Cavernous - large channels with a fibrous coat; and (3) Cystic- large cystic endothelial lined spaces (2). Although they can occur anywhere in the body, the most common sites are in the posterior triangle of the neck (75%), axilla (20%), mediastinum (5%), groin, retroperitoneal space and pelvis (3).

We report two cases of cystic lymphangioma in adults, and contrast it with those presenting in the pediatric age group.

CASE REPORTS

Case 1

A 27-year-old man presented with recurrent swelling in the nape of the neck since 5 years. The swelling was gradually progressive, and not associated with pain, discharge or sudden increase in size. He was otherwise asymptomatic. On examination, the mass was 8.0 x 8.0 cm non-tender, soft in consistency, nodular, non-fluctuant and transilluminant with well defined rounded borders. Imaging studies showed a heterogenous lesion superior to the muscle with few small cystic areas, and no calcification. A clinical diagnosis of neurilemmoma was rendered. An excision biopsy was done and the mass was sent for histopathology. The specimen consisted of a skin covered nodular mass measuring 8.0x7.0x4.0 cm with multiple cystic spaces containing clear fluid (Figure 1). Microscopic examination showed epidermis overlying dermal and subcutaneous tissue with large caliber ectatic lymphatic channels lined by

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endothelial cells containing intraluminal lymphocytes and surrounded by a fibrotic stroma consistent with cavernous lymphangioma (Figure 2).

Case 2

A 59-year-old male presented with a cystic swelling on the left side of the neck since fifteen days. The swelling was cystic, not associated with any pain, discoloration or sudden increase in size. The excised mass measured 2.5x2.0x1.0 cm and cut surface showed grey white lobulated areas with multiple cystic spaces (Figure 3). Microscopy showed dilated vascular channels lined by attenuated cells and surrounded by bundles of smooth muscles. The stroma showed focal lymphoid aggregates (Figure 4).

DISCUSSION

Cystic lymphangiomas are extremely rare benign tumors occurring in adults. They occur frequently in infants or children younger than two years of age (3). They may be developmental, hamartomatous or neoplastic in origin (4). The most widely accepted theory about the development of cystic lymphangioma is that they arise from sequestrations of the primitive embryonic lymph sacs (5). However, the etiology in the adult population is controversial. Some authors attribute adult lymphangioma to delayed proliferation of the congenital or acquired lymphoid rests following trauma or preceding respiratory infection (6). The most common documented site is the neck (5). In children cervical lesions can cause dysphagia and airway obstruction, however, this is rare in adults. Adults usually present with an asymptomatic, soft fluctuant, well defined mass with a capsule, but it is less defined in children (7).

The anatomic location of the lymphatic malformation plays an important role in determining the histologic

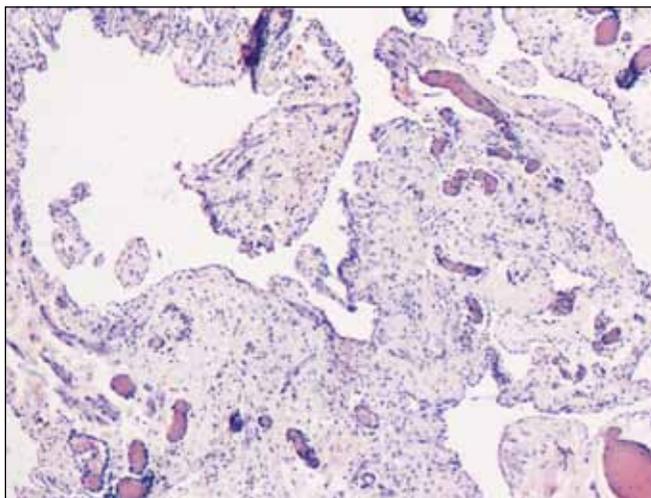


Figure 2: Ectatic lymphatic channels containing lymphocytes, (H&E, x100).

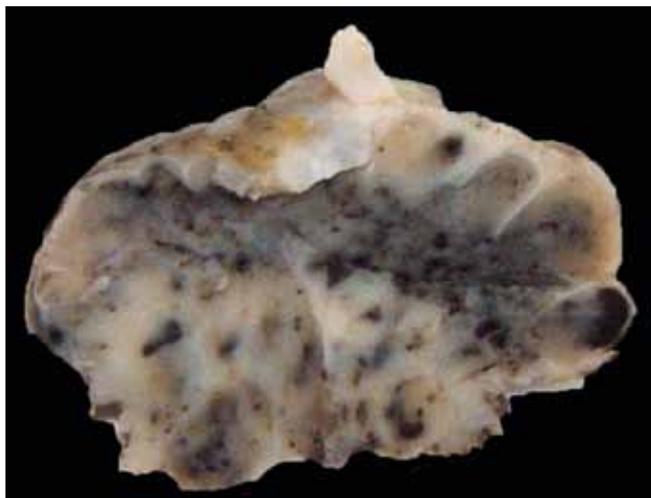


Figure 3: Lobulated grey white mass with cystic spaces.



Figure 1: Spongy mass with multiple cystic spaces.

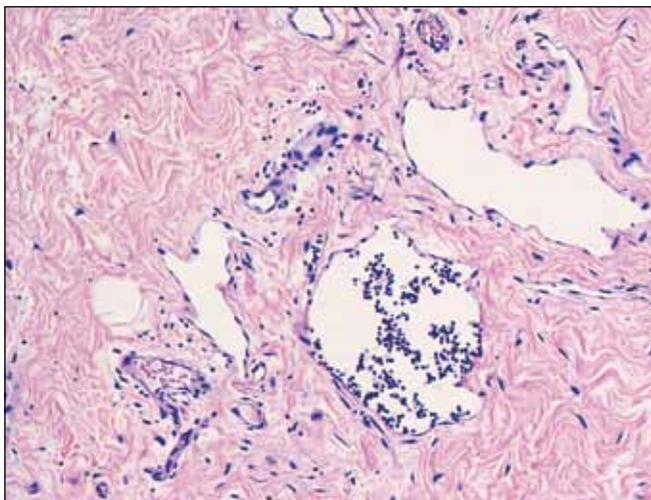


Figure 4: Dilated vascular channels surrounded by bundles of smooth muscles, (H&E, x100).

type of lymphangioma. The various sites reported are intraabdominal, mediastinal, axillary, thigh with the neck being the most common (1-8). Histologically, lymphangiomas are thin walled, cystic unilocular or multilocular cystic tumors lined by endothelial cells containing clear yellow fluid. A radiological diagnosis can be difficult. Extension into the oropharynx is present in 20% cases, and extension to the mediastinum is found in about 10% cases. A careful evaluation of the extension of the tumor by preoperative imaging using ultrasound, MRI or oropharyngeal endoscopy is strongly recommended, so as to ensure complete removal of the mass and prevent recurrence (7). Incomplete excision is one of the leading causes of recurrence. Repeated aspiration and depomedrone injections often fail to prevent recurrence. Complete surgical excision is the preferred treatment (7).

Despite all the advanced imaging techniques, the diagnosis of adult lymphangiomas remains a challenge (9). A correct diagnosis is ensured only by histopathological examination of the surgical specimen (8).

We report these cases to further emphasize the need to consider cystic lymphangioma in the differential diagnosis of neck masses in adults.

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