



Unusual Presentation of Cystic Lymphangioma Neck: A Case Report

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Abstract

Cystic lymphangioma, also known as water tumor, is a hamartomatous lesion usually noted in infants and younger children. This is basically due to the malformation of lymphatic channels. It is usually not seen in adults; however, few case reports describe the occurrence of this benign entity in elderly people as well. We describe a case of large cystic lymphangioma in the posterior triangle of the left side of neck in a 75-years-old asymptomatic female. In the present case, the role of fine needle aspiration cytology is also highlighted as an important simple investigative tool in early diagnosis of this benign lesion.

Keywords: Cystic lesion; neck, FNAC; Histopathology

Introduction

Cystic lymphangioma is a benign congenital malformation of the lymphatic system. It is usually characterized by dilatation of capillary and sinusoidal lymphatic vessels. This entity is usually noted in infant or children younger than 2 years of age [1]. It is also found as de novo in adulthood but is very rare [1-3]. The common site involved are head and neck but can be seen anywhere in the body. The patients are usually asymptomatic, but may present with cough, dysphagia, vascular compression or any other respiratory complications due to the pressure effect of enlarging mass. This condition should be kept as an important differential along with other lesions like thymic cyst, pericardial cyst, bronchogenic cyst, cystic teratoma, etc., while evaluating a neck swelling in adults [3,4]. A careful clinical, pathological and radiological assessment should be done in elderly patients who present with a painless, soft, slowly enlarging neck mass, so that early intervention can be planned, and patients can be managed accordingly.

Herein, we report a case of large cystic lymphangioma in a 75-years-old female patient in the neck region. We emphasize an importance of proper clinical examination along with role of a simple investigative modality, i.e., Fine Needle Aspiration Cytology (FNAC) in the initial diagnosis of such a benign condition which can be managed accordingly.

Case Presentation

A 76-years-old female presented with a large swelling on the left side of neck for the last 2 years (Figure 1). The swelling had progressively increased in size. On physical examination, the swelling was non-tender, soft and cystic with a smooth margin. Ultrasound of the neck showed evidence of a large cystic septate lesion, measuring 118 mm × 96 mm × 79 mm (volume- 475 cc) on the left side of neck in the posterior triangle (Figure 1). An impression of lymphangioma was given on radiological examination and pathological correlation was advised.

Her complete blood count and other biochemical parameters were within normal limits. Fine needle aspiration from the large cystic swelling on the left side of neck was done and yielded 80 mL of pale straw colored fluid. Cytological smears showed numerous mature lymphocytes admixed with many macrophages in a proteinaceous fluid background (Figure 2). Cytological diagnosis was consistent with the radiological findings of lymphangioma. The swelling was subsequently excised and sent for histopathological examination. Grossly, a skin covered tissue, measuring 10 cm × 4 cm × 1 cm was received (Figure 2). Sections examined confirmed the diagnosis of cystic lymphangioma in an elderly patient (Figure 2).

Discussion

Cystic lymphangioma is a benign malformation of lymphatic channels and is also known as water-tumor [2-4]. This soft tissue tumor was reported by Reden Backer in 1828 for the first time

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Figure 1: Clinical image showing a large cystic swelling in the left side of the neck. Ultrasound of the neck with evidence of a large cystic septate lesion, measuring 118 mm x 96 mm x 79 mm in the posterior triangle of the left side of neck.

[3-5]. It was also termed as “cystic hygroma” by Wernker in 1834 [4]. The usual sites where this can occur are, in the head, neck, axilla, cervico-facial regions, groin, and below the tongue, mediastinum, and many other anatomical sites. This is usually found in children when there is an active lymphatic growth. It is found in literature that approximately 70% to 90% of cases are found from birth till 2 years of age [2-5]. However, rarely it can also be noted in adulthood [1-6].

The cause of its occurrence in adulthood is uncertain, however, can be due to delayed proliferation of congenital or acquired lymphoid cells following trauma or upper respiratory tract infection [4-6]. Its diagnosis in adults is difficult to establish owing to its rarity; however final diagnosis is based on post-operative histological examination. Histologically, it is characterized by the proliferation of small lymphatic vessels with intervening fibrous tissue.

They have no gender predilection and present as a painless mass that progressively enlarges. Typically, the mass is soft, non-tender and ill-defined. *Majority of the patients are asymptomatic; however, can present with symptoms related to pressure effects from the slowly enlarging huge masses or due to sudden hemorrhage or infections in the swelling.*

Surgical excision is the treatment of choice for cystic lymphangioma and helps in alleviating the pressure symptoms. These are locally aggressive, benign lesions that are difficult to manage due to high recurrence rate post-surgery. *Radiological evaluation helps primarily in establishing the benign nature of the lesion along with its cystic component.*

In the present case, the lesion was correctly characterized by ultrasound with demonstration of cystic component with fluid content. In our case, the initial diagnosis was established first by a simple cytological evaluation followed by radiology and histopathology. However, there is no consensus in the literature regarding the use of fine-needle aspiration cytology in the diagnosis of these lesions. Complete surgical excision of this benign entity has shown a cure rate of 80% to 84% [5-8].

Our case is rare in the fact that this benign entity occurred in a 75-years-old female, who presented with large, slowly and progressively increasing de novo swelling with no symptoms and no history of trauma or significant respiratory infection in the past. Our

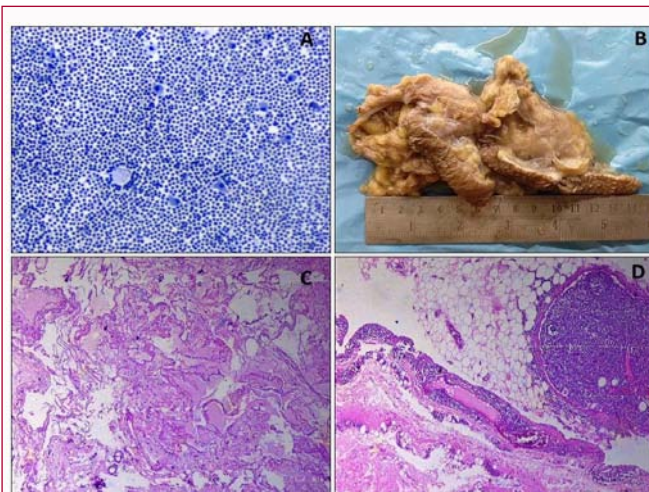


Figure 2: Fine needle aspiration cytological smear showing numerous mature lymphocytes admixed with many macrophages in a proteinaceous fluid background [Pap smear-200X]. Gross image of the excised skin covered surgical specimen of the neck swelling. H&E section showed evidence of numerous dilated lymphatic channels [H&E-00X] along with presence of lymphoid follicles [H&E-200X] confirming the diagnosis of cystic lymphangioma.

patient underwent complete surgical removal of the lesion and was doing well till last follow up of 6 months.

Conclusion

Cystic lymphangioma is a benign entity, usually found in infants and younger children. It is very rarely noted in elderly people. It is an important differential which should be kept while evaluation of any large neck swelling in adulthood. A comprehensive clinico-radiological and pathological approach helps in early diagnosis of such lesions. The present case report highlights the importance of simple investigative modality, i.e., FNAC in early diagnosis of this entity in an elderly female.

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