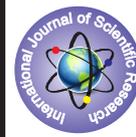


CYSTIC HYGROMA IN AN ADULT



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ABSTRACT

BACKGROUND. Our objective is to present a very rare clinical case of cystic hygroma in an adult that clinically presented as an asymptomatic swelling in the neck and was diagnosed after histopathology as cystic hygroma.

The objectives of this case report are to discuss the clinical presentation, diagnostic differentials, histopathological findings and appropriate management of this malformation.

MATERIALS AND METHODS. A clinical case of a patient with an asymptomatic neck swelling diagnosed as a cystic hygroma is presented. The patient's presentation and the diagnostic procedures, surgery and the follow up procedures are discussed.

CONCLUSION Cystic hygroma is a benign congenital malformation of the lymphatic system that occurs in infant or children younger than 2 years of age. Although cystic hygroma is well recognized in paediatric practice, it seldom presents de novo in adulthood.¹ These commonly present in head and neck but may present anywhere. Cystic hygroma is very rare in adults, but it should be considered in the differential diagnosis of adult neck swellings. Radical excision appears to be the treatment of choice.

INTRODUCTION

A cystic hygroma is a malformation of the lymphatic system most often observed in infants and children.^{1,2} Case reports of cystic hygroma in the adult are exceedingly rare and a recent report has estimated that fewer than 150 cases can be found in English-language scientific literature.¹ Also referred to as lymphangiomas or lymphatic malformations,^{1,2} cystic hygromas are most often found in the cervicofacial region (75%) and are less frequently seen in the axilla (20%) or elsewhere.

The exact nature of cystic hygroma formation is still debated. Once considered to be neoplastic in nature,³ these benign masses are now better understood. Few have suggested the role of trauma or viral infection in their development,⁵ while others discuss a more congenital aetiology involving miscommunication between lymphatic and venous pathways, aberrant lymphatic growths, and tissue sequestration during development. The role of chromosomal abnormalities has been documented, most often involving Turner's syndrome, trisomies 13, 18, and 21, and Noonan syndrome however² review of the literature did not reveal information about the prevalence of chromosomal abnormalities in adult-onset cases. Its development in adulthood has been proposed to be related to several predisposing factors such as trauma, infection, tumor growth or iatrogenic stimuli.⁶

CASE REPORT

A 40 year old female presented with a painless, gradually progressive swelling in right side of the neck since one month. Patient had previous history of similar complaint 6 months back for which incision and drainage was done and healed scar was present as seen in Figure 1. Patient did not have any complaints of pain. On examination, there was a well-defined solitary 6 cm x 4 cm, cystic, nontender, mobile swelling in right side of neck. Medial and lateral border 10 cms and 14 cms from midline of the neck and upper and lower border 3 cms and 9 cms from mastoid process. USG scan showed a well circumscribed cystic lesion. Cytologic analysis of the initial fine needle aspiration revealed lymphocytes and scarce red blood cells consistent with a diagnosis of a cystic hygroma. (Figure 2). Surgical excision of the tumor showed



Figure 1

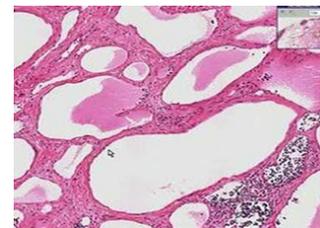


Figure 2

DISCUSSION

In contrast to cystic hygromas in children, lymphangiomas in adults are quite rare and their etiology remains unknown. These lesions may be present congenitally as cystic hygromas and may not become clinically manifest until later in life, when they may start enlarging. Differential diagnosis for a cystic hygroma includes soft tissue sarcoma, abscess, synovial cyst, and hematoma.^{1,2} Diagnosis in adult subjects is difficult to achieve, and also management of these conditions is still challenging because they tend to infiltrate adjacent tissues, causing frequent relapses.

Several staging and classification mechanisms have been proposed and adopted which allow for better diagnosis and management. Smith et al. described cystic hygromas as either microcystic, macrocystic, or mixed, with microcystic containing cysts less than 2 cm in diameter. This allowed for a more accurate prediction of how the mass would respond to sclerotherapy treatment.^{6,7} Mulliken⁸ has described cystic hygromas in terms of histological appearance as either capillary lymphangiomas, cavernous lymphangiomas, or cystic hygromas. De Serres⁹ proposed a more practical classification system based on location, which allowed for a better estimation of prognosis and surgical complication rate, but applies only to masses in the cervicofacial region. Diagnosis is often aided by the use of fine needle aspiration for cytology, tissue histology, and ultrasound, MRI, or CT for definition of the mass. Loculations or cysts can often be appreciated and affected structures can be identified.¹

Continued reporting of adult-onset cystic hygroma is of particular importance, as the nature and management of these rare masses are elucidated. It should be considered in the differential diagnosis of adult neck swellings. Patients presenting with a painless, soft, fluctuant, and enlarging neck mass should have a careful history and physical examination along with radiological imaging to assist with diagnosis.

Surgical excision with encapsulation of the mass is the treatment of choice for this rare condition.

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