Case Report

Adult Cystic Hygroma– A Rare Entity
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ABSTRACT
Objective: Lymphangiomas (Cystic Hygromas) 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 2 years of age. We report this case to emphasise the need to consider cystic hygroma in differential diagnosis of neck masses in adults. Case Reports: A 50-year-old female presented with a swelling over the antero-lateral aspect of neck of size 8’6 cm since last 4 years, which was not associated with pain, discharge, discoloration or sudden increase in size. Conclusion: The awareness of occurrence of cystic lymphangio main adults is important for its proper management, which includes complete surgical removal, to prevent recurrence.

KEYWORDS: Lymphangioma, Neck masses, Adults, Cystic hygroma, Respiratory tract infection, Sternocleidomastoid muscle, Lymphatic system

INTRODUCTION
Cystic hygroma is an aberrant proliferation of lymphatic vessels resulting from abnormal development of the lymphatic system[1,2]. Within the literature the term cystic hygroma is used interchangeably with lymphangioma and lymphatic malformation[1,3,4]. Cystic hygroma could be classified into septated (multiloculated) or non-septated single cavity (non-loculated). Presentation in adulthood is rare and the cause is uncertain, although trauma and upper respiratory tract infection have both been suggested as possible triggers for onset[5,6]. In this case there was no identifiable cause and onset was sudden and rapid. Most commonly these malformations occur in the head and neck, although they have been described in a variety of other anatomical locations.

To date there have been fewer than 150 reports of adult cervicofacial cystic hygroma in the English language literature and the optimum management of these lesions is still a matter of debate. Diagnosis in adults is considered to present a greater challenge than that in children and initial misdiagnosis, frequently as branchial cleft cysts as in the case reported here, is common[5,7]. Definitive diagnosis is usually based on post-operative histology.

We present a case of cystic hygroma in an adult and discuss the management options for such a presentation.

CASE PRESENTATION
Clinical Presentation
A 50-year-old woman came to General Surgery OPD at MNR Hospital with left antero-lateral neck swelling along the full length of sternocleidomastoid muscle. There is no history of trauma or upper respiratory tract infection. Patient was operated for similar swelling on the right side of the neck 4 years back at Govt. General hospital.

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Clinical Examination
On examination, the patient was found to have a single, oval, painless, freely mobile swelling of size 8x6 cm with well-defined borders on the antero–lateral aspect of neck along the full extent of sternocleidomastoid and the swelling is decreasing on contraction of sternocleidomastoid muscle.

Investigations
USG neck showing multi-septate, irregular cystic lesion noted in the neck region bilaterally (clinically the right-sided mass was not palpable).

On chest X-ray, soft tissue opacities with internal calcifications in middle and upper zones of right lung are seen. Soft tissue swelling is also seen in the left supra-clavicular region.

On CT chest and neck, a large multiloculated cystic lesion noted in both sides of neck, superior and anterior mediastinum measuring 9x6 cm on the left side, 5x4 cm on the right side and 14x7 cm in mediastinum. Mediastinal lesion is encroaching the arch of aorta, trachea and superior vena cava. All the vessels show normal contrast filling. Few loculi are showing wall calcifications. Cardiomegaly with mild pericardial effusion is also seen.

She was unable to perform spirometry and her PEFR was 160ml/min.

2D echo showed conc. LVH with mild pericardial effusion was seen (Figure 1, 2).

On FNAC, straw-coloured fluid was aspirated, which on cytology showed numerous small lymphocytes.

At this point the diagnosis was found to be cystic hygroma.

Surgical Management
As the surgical condition of the patient was poor, excision of the swelling was planned under local anaesthesia. Lateral skin crease incision taken over the neck mass, sub-platysmal flaps raised. Sternocleidomastoid muscle retracted to expose the multilocular cystic, thin-walled lesion throughout its extent. Spinal accessory and internal jugular vein were carefully preserved. Contents of the cyst leaked intra-operatively toward the final phase of the dissection but complete removal of the wall of the mass was achieved. Histological examination revealed cystic spaces lined with flat endothelial-like cells consistent with a diagnosis of cystic hygroma.

DISCUSSION
Cystic lymphangiomas are extremely rare benign tumours occurring in adults. They occur frequently in infants or children >2 years of age. They may be developmental, hamartomatous or neoplastic in origin. The most widely accepted theory about the development of cystic lymphangioma is that they arise from sequestrations of the primitive embryonic lymph sacs. 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. The most common documented site is the neck. 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. The anatomic location of the lymphatic malformation plays an important role in determining the histologic type of lymphangioma. The various sites reported are intraabdominal, mediastinal, axillary, thigh with the neck being the most common. Histologically, lymphangiomas are thin-walled, cystic unilocular or multilocular cystic tumours 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 ~10% cases. A careful evaluation of the extension of the tumour by preoperative imaging using ultrasound, MRI or oropharyngeal endoscopy is strongly recommended, so as to ensure complete removal of the mass and prevent recurrence. Incomplete excision is one of the leading causes of recurrence. Percutaneous aspiration is not preferred because of the risk of bleeding, infection and recurrence. Injection of sclerosing agents such alcohol,
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bleomycin\textsuperscript{17} and OK-432 (a lyophilised mixture of \textit{Streptococcus pyogenes} and Penicillin G potassium)\textsuperscript{18} have been reported with favourable results. Complete surgical excision is the preferred treatment\textsuperscript{14}. Despite all the advanced imaging techniques, the diagnosis of adult lymphangiomas remains a challenge\textsuperscript{16}. A correct diagnosis is ensured only by histopathological examination of the surgical specimen\textsuperscript{15}.

REFERENCES


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Figure 5 and 6: cystic spaces lined with flat endothelial-like cells


[7] Jaroslav Kraus, Jan Plzák, Roberto Bruschini, Giuseppe


