

Adult-onset cystic hygroma: a case report and review of management

L. Gow^a, R. Gulati^b, A. Khan^b and F. Mihaimeed^c

^a*Barts and the London Medical School, Whitechapel, London, E1 2AD, UK;*

^b*Department of General Surgery, Newham University Hospital, London, E13 8SL, UK;*

^c*Departments of Surgery and Anaesthesia, Newham University Hospital, London, E13 8SL, UK*

*Corresponding address: Mr F. Mihaimeed, Department of General Surgery,
Newham University Hospital, London, E13 8SL, UK.
Email: faisal.mihaimeed@newhamhealth.nhs.uk*

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Abstract

Cases of cystic hygroma are rare and this report highlights the need for further research into treatment options. A 24-year-old Lithuanian woman was referred for further investigation of a painless but restricting right-sided neck swelling. This case report discusses the differential diagnosis and up-to-date management of cystic hygroma in the adult.

Keywords

Adult; cystic hygroma; lymphangioma; lymphatic malformation; surgical excision; sclerotherapy; OK-432.

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 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.



Fig. 1. A 24-year-old woman presenting with a large right-sided cervical swelling.

Case presentation

Clinical presentation

A 24-year-old Lithuanian woman was referred to the general surgery clinic with an interest in neck surgery by her GP for further investigation of a right-sided neck swelling, along the full length of the sternocleidomastoid muscle (Fig. 1). The swelling was of sudden onset with no preceding upper respiratory tract infection or trauma. The patient reported a previous episode of sudden onset of swelling in the same location at the age of 21 years, for which she had been admitted to hospital in Lithuania and successfully treated with aspiration and antibiotics.

Clinical examination

On examination in our clinic, the patient was found to have a 10×4 cm, freely mobile swelling extending into both the posterior and anterior triangles of the neck. The mass was painless and the patient was asymptomatic apart from some restriction of neck movement. Some degree of social embarrassment caused by the mass was reported.

Investigations

Prior to referral, an ultrasound scan was carried out and revealed a 5×2 cm multiloculated (septated) cystic swelling, with internal septae, related to the right sternocleidomastoid with an initial impression of branchial cleft cyst with internal haemorrhage. A substantial increase in size had occurred in the 4 months since the scan. Magnetic resonance imaging of the neck revealed a 12×2.5 cm mass with smooth margins, extending from the root of the neck to the submandibular gland, deep to the right sternomastoid and lateral to the carotid and jugular vessels (Fig. 2). This was classified as a stage III lesion according to the staging system proposed by de Serres (Table 1)^[8]. Cloudy yellow fluid was aspirated from the cyst and on cytological analysis was found to contain numerous small lymphocytes. At this point diagnosis was thought to be most likely cystic hygroma.

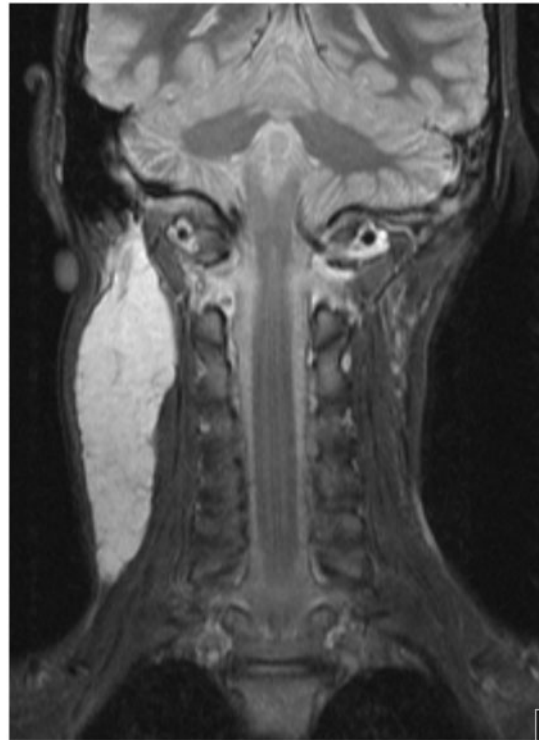


Fig. 2. Coronal T2-weighted magnetic resonance image of the head and neck showing a large hyperintense mass extending from the level of the submandibular gland inferiorly to the root of the neck.

Table 1. Proposal for staging of lymphatic malformations of the head and neck (adapted from de Serres^[8])

Stage	Location of lesion
I	Unilateral infrahyoid
II	Unilateral suprahyoid
III	Unilateral infrahyoid and suprahyoid
IV	Bilateral suprahyoid
V	Bilateral infrahyoid and suprahyoid

Surgical management

The patient underwent surgical exploration of the right neck and excision of the mass via a vertical incision along the anterior border of the right sternocleidomastoid muscle, by the senior author. The mass was encapsulated with a thin wall and although adherent to surrounding musculature, adhesions were flimsy and readily divided with careful blunt dissection using fine artery (Mosquito) forceps. However, it was however necessary to sacrifice the transverse cervical sensory nerve and posterior facial vein. Contents of the cyst leaked intra-operatively towards the final phase of the dissection but complete removal of the wall of the mass was achieved. Macroscopically the specimen measured approximately 13 cm in length (Fig. 3). All the important nerves and arteries encountered during the dissection were seen and preserved. Histological examination revealed cystic spaces lined with flat endothelial-like cells consistent with a diagnosis of cystic hygroma.

Post-operative recovery

Post-operatively the patient recovered well with no signs of any neurological dysfunction or angle of mouth deviation and she was discharged from hospital 1 day post-operatively. For 3 months

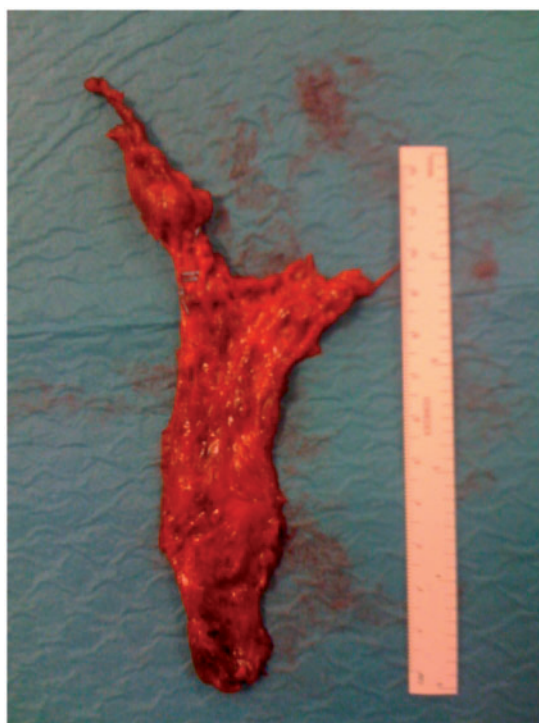


Fig. 3. Post-operative specimen. Mass removed from the neck of a 24-year-old woman presenting with right neck swelling.

post-operatively the patient reported experiencing pain, posterior to the right shoulder. Movement of the shoulder joint and muscle strength was unaffected. At her last follow-up, 4 months post-operatively, this pain had calmed significantly and at this time there was no evidence of recurrence of the mass and no weakness of the trapezius muscle with full range of movement of the shoulder. In a telephone consultation 1 year after surgery, the patient reported no recurrence but she had a slight concern about the appearance of the scar.

Discussion

Cystic hygroma is believed to arise from a congenital malformation of the lymphatic system in which a failure of communication between the lymphatic and venous pathways leads to lymph accumulation. Most cystic hygromas present in-utero or in infancy and therefore most of the literature on management considers paediatric cases. The effect of these lesions depends on their position and relationship to surrounding structures, although the most common adult presentation is of a painless lump in an otherwise asymptomatic patient^[5]. However, rapid enlargement over a short period of time has frequently been reported^[9] and major structures such as the larynx, trachea, oesophagus, brachial plexus and great vessels have known to be compressed or incorporated within the lesion^[9]. In this case the lesion had doubled in size over a period of 4 months and had caused a restriction of neck movement; others have reported presentation with pain, hoarseness, dysphagia and breathlessness^[7,10,11].

Complete surgical excision has traditionally been considered the treatment of choice for cystic hygroma^[12-15]. However, several authors have suggested that sclerotherapy may be a more appropriate first-line therapy^[16,17]. Although sclerotherapy is now well established in the treatment of neonatal and paediatric cystic hygromas, there have been relatively few cases reported of its use in adult patients. Some success has been reported in small numbers of adults with sclerotherapy agents such as OK-432^[18]. Caution has been urged with the use of agents such as OK-432 which induce a local immune response that often results in a rapid temporary increase in the size of the cystic hygroma^[17,18]. Depending on the anatomical relations of the tumour, such increases in size may be intolerable and it has been suggested that such therapy should only be administered in specialised facilities due to the risk of airway obstruction^[19].

Smith et al.^[17] compared results from their large-scale trial of OK-432 with pooled results from large surgical case series reported in the literature and reported greater success rates and lower occurrence of major complications with OK-432 sclerotherapy compared with surgical excision.

However, their study focussed mainly on children and only one of the comparative surgical case series included adult patients. Several authors have expressed the opinion that surgical excision of cystic hygroma is an easier procedure in adult patients, because these lesions are better circumscribed, and as such the success rate is greater^[2,5]. In addition, clinical success of OK-432 treatment was defined as a greater than 60% reduction in size^[17]. This may leave a substantial proportion of the lesion in situ and the acceptability of this may depend on the exact location and relations of the tumour. It is therefore difficult to apply these results to our case of an adult patient with a lesion in close relation to several important structures in the neck.

In addition to lack of evidence surrounding the best management of adult cystic hygroma, there are barriers to obtaining OK-432 for use in the United Kingdom. OK-432 is not licensed for use in the European Union but was previously obtainable within the United Kingdom on a named patient basis via IDIS World Medicines^[18]. On investigation of this, however, we were informed that IDIS have now stopped supplying the medicine due to a lack of documentation on transmissible spongiform encephalopathies. Moreover, treatment with sclerotherapy does not allow the diagnosis to be confirmed because definitive diagnosis requires excision for full tissue analysis.

In this case, it was thought that the ideal treatment would be complete surgical excision as multiloculated cystic hygroma may not respond to sclerotherapy. Success of surgery has been found to correlate with histology, encapsulation, complete excision, anatomical location and stage of the lesion^[3,8,12]. Imaging appeared to show smooth margins, indicating a lack of infiltration, which is a good prognostic feature, facilitating complete removal and low recurrence. However, this was an extensive stage III lesion with close relations to major structures and therefore a difficult procedure was anticipated. It proved to be impossible to remove the cystic hygroma completely without rupture, a recognised problem as these tumours usually have a fragile thin wall. Intra-operative rupture of the lesion complicates complete removal as it obscures the limits of the structure^[4]. However, Riechelmann^[12] reported very low levels of recurrence (1/9 patients) following subtotal excision when small plaques of tumour wall were known to be left in situ. To the best of our knowledge we were able to remove the cystic hygroma completely.

Complications of surgery should be discussed with the patient before consent is obtained for surgery, including

- Scar: there may be a long scar depending on the size of the cystic mass but this should usually improve over time.
- Injury to important structures and nerves: all blood vessels and nerves located between the mandible and the sternoleidomastoid muscle are vulnerable to injury, specifically the mandibular branch of the facial nerve, the spinal branch of the accessory nerve and the greater auricular nerves.
- Venous bleeding is a possibility but injury to the carotid sheath and its content and some external carotid branches should be rare.
- Wound Infection is usually rare in neck surgery.
- Recurrence after surgery is a possibility.

Further research into the effectiveness of sclerotherapy, particularly OK-432, in the management of both multiloculated and single cavity cystic hygroma in adult patients is necessary to inform treatment decisions; however, this may not be feasible because of the rarity of this pathology in adults. Few studies present long follow-up although recurrence has been reported as much as 6 years after surgery^[5], indicating the need for long-term close monitoring. Management of cystic hygroma, particularly for more advanced stage lesions, remains a complicated issue and decisions should be made on a case-by-case basis with full patient involvement. We present a case of a stage III cervicofacial lesion, successfully excised with no recurrence at the present time. Case reports of the management of cystic hygroma in adults could be the only source to inform the surgical community about management outcomes and should be encouraged, as it would be impossible for one centre to accumulate enough cases to draw conclusions on a definitive management plan.

Teaching points

- Congenital neck masses usually present at birth but may present at any age
- Cystic hygroma is a rare presentation in adults
- Malignancy should be excluded in all adult patients presenting with a cystic neck swelling

- Differential diagnoses for a congenital neck mass in adults
 - Branchial cleft cyst (lateral to midline)
 - Dermoid cyst (midline)
 - Thyroglossal cyst (midline)
 - Haemangioma
 - Thymic cyst (midline/lateral)
 - Neck malignancy (midline/lateral)
- Investigations should include:
 - Ultrasound scan
 - Fine-needle aspiration (important to exclude malignancy)
 - Magnetic resonance imaging or computed tomography
- Final diagnosis depends on tissue analysis
- Treatment options in adults include:
 - Surgery
 - Sclerotherapy
- In multiloculated cystic hygromas, surgery may be the preferred option but it is important that surgical complications are kept to a minimum.

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