

Adult Cystic Hygroma: An Enigma for the Clinician

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ABSTRACT: Cystic Hygroma is a benign lymphatic malformation/aberration presenting usually in perinatal period, almost always before age of 2. Presentation in adults is rare. We hereby present a rare occurrence of Cystic hygroma with rapid onset in a 60 year old female. This case report discusses the presentation, diagnosis and options of management.

KEYWORDS: cystic hygroma, lymphangioma, lymphatic malformation, surgical

I. INTRODUCTION

Cystic hygroma is a benign aberration in development of lymphatics either due to sequestration of or by obstruction of outflow of developing lymphatics. It usually presents in perinatal period upto age of 2 years.[1],[2] Adult occurrence is rare with fewer than 150 cases reported in literature.[3],[5] Diagnosis in adults is considered to present a greater challenge than in children and initial misdiagnosis, frequently as branchial cleft cysts is common.5 Definitive diagnosis is usually based on post-operative histology.

Adult presentation is rare, usually with no identifiable cause. Upper respiratory tract infection and trauma are some of the identifiable triggers.[6],[7] This case came with no identifiable cause with sudden and rapid onset. These usually present in head and neck region but other anatomic locations have been described.

We present an adult onset cystic hygroma case with discussion on the presentation and management. The lymphatic system is the network of vessels responsible for returning to the venous system excess fluid from tissues as well as the lymph nodes that filter this fluid for signs of pathogens. These malformations can occur at any age and may involve any part of the body, but 90% occur in children less than 2 years of age and involve the head and neck. These malformations are either congenital or acquired. Congenital lymphangiomas are often associated with chromosomal abnormalities such as Turner syndrome, although they can also exist in isolation. Lymphangiomas are commonly diagnosed before birth using fetal ultrasonography. Acquired lymphangiomas may result from trauma, inflammation, or lymphatic obstruction.[21]. These malformations can occur at any age and may involve any part of the body, but 90% occur in children less than 2 years of age and involve the head and neck. These malformations are either congenital or acquired. [22]

II. RELATED WORK

Case report:

A 60 year old female came to our clinic with history of a rapidly growing (onset to presentation in 3 months) disfiguring mass on left side of neck with breathlessness more in supine position especially in night, with some dysphagia and hoarseness of voice from 1 month. There was no preceding history of trauma or upper respiratory tract infections. Physical examination showed a 10 x 7cm lobulated, soft, fluctuant, transilluminant mass occupying the left

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side of neck extending to posterior triangle starting from root of neck, fixed to deeper structures and free under the skin. FNA showed straw coloured fluid with mature lymphocytes and histiocytes. MRI of neck revealed cystic malformation mostly lymphatic with no vascular element within the cyst. The patient underwent complete excision of the multilocular lesion by a Macfee incision under general anaesthesia. There was Intraoperative leak of the fluid but lesion was excised completely. Histopathology confirmed the lesion to be a lymphangioma. No recurrence was seen at 3 months follow up.

III. METHODOLOGY

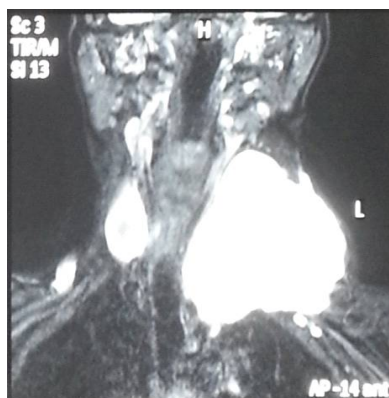
- STUDY DESIGN: Case study
- SAMPLING TECHNIQUE: Convenient Sampling
- STUDY SETTING: D.Y. Patil Hospital, Nerul, Navi Mumbai.
- SAMPLE SIZE: Single Case Study
- POPULATION FOR STUDY: Patients suffering from Cystic hygroma.
- DURATION OF STUDY: 3 months
- MATERIAL USED: Surgical Equipment in Operation Theatre

PROCEDURE: The patient underwent complete excision of the multilocular lesion by a Macfee incision under general anaesthesia. There was Intraoperative leak of the fluid but lesion was excised completely. Histopathology confirmed the lesion to be a lymphangioma.

IV. RESULT AND INTERPRETATION

A 60 year old female came to our clinic with history of a rapidly growing (onset to presentation in 3 months) disfiguring mass on left side of neck with breathlessness more in supine position especially in night, with some dysphagia and hoarseness of voice from 1 month. There was no preceding history of trauma or upper respiratory tract infections. Physical examination showed a 10 x 7cm lobulated, soft, fluctuant, transilluminant mass occupying the left side of neck extending to posterior triangle starting from root of neck, fixed to deeper structures and free under the skin. FNA showed straw coloured fluid with mature lymphocytes and histiocytes. MRI of neck revealed cystic malformation mostly lymphatic with no vascular element within the cyst. The patient underwent complete excision of the multilocular lesion by a Macfee incision under general anaesthesia. There was Intraoperative leak of the fluid but lesion was excised completely. Histopathology confirmed the lesion to be a lymphangioma. No recurrence was seen at 3 months follow up.

Picture 1: Sagittal T2 image



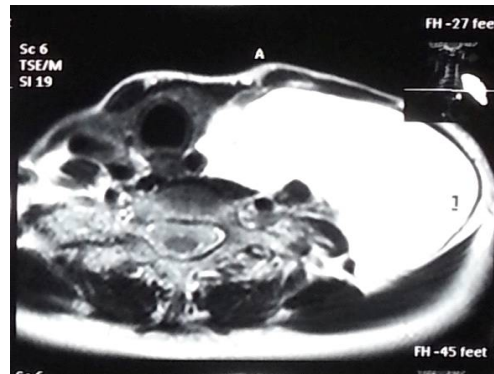
Inference: Sagittal Image shows Cystic hygroma

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Picture 2: Transverse T2 image



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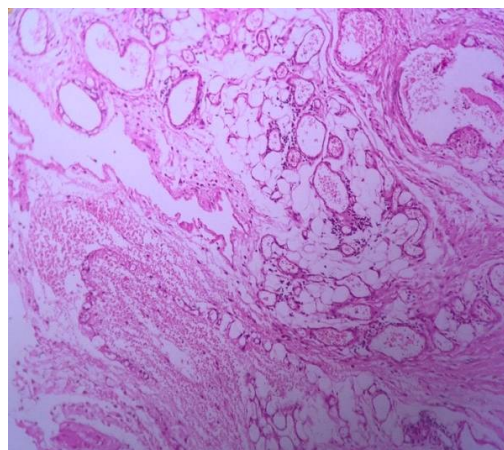
Inference: Transverse T2 image shows Cystic hygroma

Picture 3: Intra-operative Photo



Inference: Intra-operative multiloculated lesion with colour change due to cystic multiple FNAC's

Picture 4: Light Microscope Image



Inference: Histopathologic slide of lesion under light microscope

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V. DISCUSSION

Cystic hygroma is believed to arise from lack of communication between the lymphatic and the venous system. This leads to accumulation of the lymph. It is usually seen in the perinatal period and so most of the literature is regarding management in paediatric cases.[1-5] Presentation depends on the site and size of the lesion. Our patient presented with a rapid onset and progression and with some compression of trachea and esophagus. The usual presentation in adults is a painless growth in an asymptomatic patient. There are reports of rapid onset and compression or incorporation of vital structures in the malformation.

In 1956, Landing and Farber[10] classified lymphatic malformations according to the size of their vascular spaces into lymphangioma simplex composed of capillary-like vasculature, cavernous lymphangiomas composed of dilated sinuses that do not communicate, and cystic hygromas, which are larger and multiloculated. Kennedy[9] has recently classified lymphangiomas into four types: superficial cutaneous, cavernous, cystic hygromas, and diffuse systemic. A more recent classification based on CT, anatomical location, and histology was proposed by McGill and Mulliken[9],[11] in 1993 and is more clinically practical. In this classification, type I malformations are often macrocystic and develop below the mylohyoid muscle. They involve the anterior and posterior triangles and are most consistent with the classic cystic hygroma. In contrast, type II malformations are often microcystic and invasive and are found in the neck above the level of the mylohyoid. They usually involve the lip, tongue, and oral cavity and are difficult to resect.

MRI is the radiological investigation of choice. The hygroma appears hyperintense in T2 weighted sequences. Ultrasonography and CT scan are other options. MR Lymphangiogram can detect patency of communications.[12], [19] Surgical excision of the lesion has traditionally been the gold standard of care.[1],[2],[3],[15],[16],[17] Complications may include postoperative muscle weakness, nerve damage, neural weakness, bleeding, sepsis, infection, scar as well as recurrence. Intraoperative rupture complicates excision as it obscures margins. Riechelmann 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.[13]

Some authors have suggested use of sclerotherapy in line with paediatric cases as the first line of therapy[16],[17]. But efficacy of the same remains unproved. Drugs shown to be useful in sclerotherapy are Picibanil (OK-432), Doxycycline, Bleomycin, hypertonic saline, alcohol, acetic acid. Picibanil (OK-432) is mixture of lyophilised group A streptococcus pyogenus.[18] Efficacy of the same remains unproved.[20] Other options include repeated aspirations, intralesional steroids, radiotherapy. The literature is unclear as to the efficacy of these alternative treatments.[1][7]

VI. CONCLUSION

Being rare, continuing reporting of the cases of Cystic hygroma in adults will help elucidate various presentations, diagnostic dilemmas, management options and complications. This case contributes to the body of literature in the diagnosis of neck masses in adults.

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