Parapharyngeal lymphangioma - a case report

Dr. Priyadershini A. Rangari¹, Dr. Mahindra Katre², Dr. Farhana Girkar³, Dr. Shaliputra. P. Magar⁴

¹PG student, Department of Oral Medicine and Radiology, Institute of Dental Studies and Technologies, Chaudhary Charansingh University, Meerut (U.P) India.
²Senior Lecturer, Department Of Otolaryngology, Govt. Medical College & Cancer Hospital, Aurangabad, Maharashtra University Of Medical Sciences, Nasik (M.S.) India.
³PG student, Department of Oral & Maxillofacial surgery, Institute of Dental Studies and Technologies, Chaudhary Charansingh University, Meerut (U.P) India.
⁴Reader, Department of Oral Medicine and Radiology, Sri Aurobindo College of Dentistry, Devi Ahilya Vishwavidyalaya, Indore, (M.P) India.

Abstract: Parapharyngeal lymphangioma means cystic hygroma involving parapharyngeal region which can obstructs the airway laterally, this is very rare condition. We present here a case report of a 9-year-old female patient who came with a chief complaint of pain and swelling on left side of face since 8 days and was later diagnosed as having cystic hygroma in parapharyngeal region on the basis of her clinical features and further imaging studies. This report also highlights the surgical and other treatment options.

Keywords: cystic hygroma, lymphangioma, parapharyngeal Lymphangioma, swellings in neck, sclerotherapy.

I. Introduction

Cystic Hygroma was first described in the European Literature by Redenbacher in 1828.³ Cystic hygroma is also known as cystic Lymphangioma, jugular lymphatic obstructive sequence, hygromacollicysticum. The term hygroma means moist tumor. Lymphangiomas are usually classified as shown in fig.1. Cystic hygroma are anomalies of the lymphatic system characterized by single or multiple cysts within the soft tissue, usually involving the cervico-facial regions and axilla. Other less common sites are mediastinum, groin and below tongue. Occasionally, these malformations occur in liver, spleen, kidney and intestine. Ommental cyst in omentum and mesenteric cyst in the mesentery of intestine represents parallel lesions at these locations.⁴ Respiratory distress, recurrent infections or cosmetic reasons are the main indications of the treatment.⁵ Exact etiology is unknown.

There are a number of proposed mechanisms to explain the pathophysiology of cystic hygroma. Embryologically, these lesions are believed to originate from sequestration of lymphatic tissue from lymphatic sacs, during the development of lymphatico-venous sacs. These sequestered tissues fail to communicate with remainder of the lymphatic or venous system. Later on, dilatation of the sequestered lymphatic tissues ensues, resulting in the cystic morphology of these lesions.⁶ They occur most commonly in the neck, which is termed nuchal cystic hygroma (occurs in ~80% of cases) and axilla, with only 10% of cases extending to the mediastinum and only 1% confined to the chest.⁷ The incidence of cystic hygroma is not well defined. Reports range from 1.7:10,000 pregnancies to 0.83% of pregnancies at risk for having a structural anomaly⁸ and there is no hereditary predisposition exists. Over 50% of cystic hygroma present at birth and more than 90% of these congenital malformations are found in children younger than 2 years with males and females equally affected. Many cases occur in the fetal population and the prevalence is 0.2-3%. It is quite rare in adult.⁵,⁹

They occur most commonly in the neck, which is then also termed nuchal cystic hygroma (occurs in ~80% of cases) and axilla, with only 10% of cases extending to the mediastinum and only 1% confined to the chest.¹⁰ The usual presentation of cystic hygroma apparent at birth is a painless mass with worries and queries of the parents about the lesion. The other modes of presentations are related to the complications or effects of cystic hygroma, such as respiratory distress, feeding difficulty, fever, sudden increase in the size and infection in the lesion.¹¹,¹² On clinical examinations, these lesions appear soft, compressible, non-tender, transluminant and without any bruit. Intraoral examination reveals bulging in posterior region of soft palate or pharyngeal wall. In case of lesion anterior lower neck mass causes elevation of tongue, floor of mouth and inability to close the mouth. The other modes of presentations are related to the complications or effects of cystic hygroma, such as respiratory distress, feeding difficulty, fever, sudden increase in the size and infection in the lesion.


DOI: 10.9790/0853-14842125 www.iosrjournals.org 21 | Page
II. Case Report

A 9 year old female Hindu patient presented with a swelling on left side of the face which was gradually increasing in size since 8 days, she also developed difficulty in breathing and pain on palpation. History of present illness reveals patient had same swelling two years back for that she had taken antibiotics and anti-inflammatory medication. Then the swelling get subsided and recurred 3-4 times up till now. Physical examination revealed a 8 X 6cm bulging soft mass over the left side of the neck, extending 3cm anterior to the ala tragus line superio-anteriorly to the 2cm posterior to ear superio-posteriorly. Inferiorly it involves midline of mandible at anterior end to angle of mandible at posterior side giving an oblique elongated appearance (Fig. 3 a and b). This swelling was a soft, compressible, elongated and tender on palpation. Local temperature was normal. Thetransillumination test was positive. Small other firm tender masses were palpable surrounding the biglobulated mass. The patient's voice was normal. There was no cervical or supraclavicular adenopathy. Initially 7 days antibiotic course was given. Intraoral examination showed bulging of left faucial pillar, soft palate and pharyngeal wall.

Prior sonography report suggested the presence of salivary gland inflammation. Recent Pulsed Doppler sonography image of the neck posterior to the left mandibular angle shows large well defined multisepted cystic mass lesion involving parapharyngeal, lingual and parotid space. Measuring 8x6cm in size consist of thick fluid inside the cystic cavity. Absence of any calcification noted. The lesion is seen adjacent to tonsillar fossa and tongue; this also displaces blood vessels in neck posterolaterally. No movement observed with tongue movement and deglutition. These findings denote lymphocele or cystic hygroma.

The echogenic portions of the lesion correlate with clusters of small, abnormal lymphatic channels. Fluid filled level can be observed with a characteristic echogenic component layering in the dependent portion of the lesion.

CECT scan of the neck and thorax showed a large loculated cystic mass lesion of near water density with few enhancing septa within. It occupied the posterior cervical space on the left side of the neck lateral to the carotid artery. Axial postcontrast CT image of the neck at the level of the mandibular angle obtained during the current admission displaced the left internal jugular vein anterior-medially and the sternocleidomastoid muscle interio-laterally. In its crano-caudal extent, it extended from C1 to the level of vertebra C4.

Axial postcontrast CT image of the neck obtained reveals a largewell defined unilateral multiloculated hypodense lesion more prominent over the left submandibular region spreading through the soft tissue measuring about 8x6x6cm in size. The major component of the lesion involved the left side of the neck from the level of the parotid gland to the thoracic inlet. There was also moderate enlargement of the adenoids and tonsils, which, together with the parapharyngeal space was partially obstructing the airway. Report of routine blood, urine and x-ray chest examination was normal.

After FNAC, microscopic examination comprised of endothelium lined cystic spaces with scanty stroma. A final diagnosis of cystic hygroma was made. Because of the increasing size and pain during the current admission, it was elected to remove the lesion surgically. Transcervical transmandibular approach through the digastric triangle was used to access the submandibular and parapharyngeal space. Submandibular gland excision done for good exposure. Multiple lymph filled sacs were encountered. A cystic area with old hemorrhagic elements due to prior bleeding was observed in the left side of the neck posterolateral to the mandibular angle. Complete excision of mass removed in toto. Neck drain kept rest postoperative period was uneventful.

Histopathology revealed thin connective-tissue stromaseparating the cystically dilated spaces lined by asingle layer of benign endothelial cells which was consistent with cystic hygroma. The patient tolerated the procedure well and suffered only minor weakness of the marginal mandibular branch of the facial nerve.
Cystic hygromas are congenital vasculolymphatic malformations that are frequently present at birth. They have no predilection for sex or race, and they have no malignant potential. Typical cystic hygromas cause no symptoms unless they enlarge in size or surround or invade adjacent normal anatomic structures. In this situation, cystic hygromas may cause symptoms such as feeding problems or breathing difficulties\(^2\). Our patient showed a tender swelling with a little difficulty in swallowing.

**III. Discussion**

Cystic hygromas are congenital vasculolymphatic malformations that are frequently present at birth. They have no predilection for sex or race, and they have no malignant potential. Typical cystic hygromas cause no symptoms unless they enlarge in size or surround or invade adjacent normal anatomic structures. In this situation, cystic hygromas may cause symptoms such as feeding problems or breathing difficulties\(^2\). Our patient showed a tender swelling with a little difficulty in swallowing.
They usually occur in the fetal/infantile and paediatric populations with most lesions presenting by the age of two. The estimated prevalence in the fetal population is 0.2-3%. Patients in the infantile or paediatric population can present with pain, dyspnoea, infection, haemorrhage or respiratory compromise. In contrast our patient was 9 yr old and she never felt any swelling or respiratory discomfort before.

Pathologically, Cystic hygromas are multilobulated, thin-wall, lymph containing sacs. The fluid within the sacs is usually produce milky, serous, sero-sanguinous or straw coloured fluid, when aspirated with a widebore needle. Cystic hygroma are thought to arise from delayed development/maldevelopment/failure of the lymphatic system to communicate with the venous system of the neck. Like other lymphangiomas, they are endotheilial lined cavernous lymphatic spaces.

They can vary significantly in size. Lymphatic vascular malformations may be mixed with other forms of vascular malformation, including capillary or venous.

They occur most commonly in the neck, which is then also termed nuchal cystic hygroma (occurs in ~80% of cases) and axilla, with only 10% of cases extending to the mediastinum and only 1% confined to the chest. In our case the lesion was above neck region and deep involving parapharyngeal region up to airway and less likely to bulging peripherally.

Associated with aneuploidic anomalies: ~65% (range 50-80%) like Turner syndrome; Down syndrome, trisomy 13, trisomy 18, triploidy and non aneuploidic like congenital cardiac anomalies oractication hypoplastic left heart syndrome, pentalogy of Cantrell, Apert syndrome, Cornelia de Lange syndrome, fetal alcohol syndrome, Fryns syndrome, lethal multiple pterygium syndrome, limb hypertrophy, Noonan syndrome, Pena Shokeir syndrome.

The mainstay of lymphatic malformation treatment has been surgical resection, which has been refined through lesion and radiographic characterization. Sometimes, this may be impossible due to the infiltrating nature of the hygroma within and around neurovascular structures, muscles and blood vessels.

In this condition, unroofing, partial cystectomy and drainage of the cystic content should be performed and all adjacent crucial structures should be preserved. Recurrence rate of 10-15% is reported.

The other techniques recently introduced in the management of lymphangiomas are radiofrequency ablation and laser excision (using CO₂, Nd-YAG, or potassium-titanylphosphate lasers) is minimally invasive and is much less painful than surgery. Complications following resection are postoperative muscle weakness, nerve injuries and neural weakness, bleeding and wound infection. In our patient the spinal accessory nerve was splayed anteriorly all across over the cystic mass. On the medial aspect, the mass could be carefully separated from the facial nerve, parotid gland, internal jugular vein, carotid vessels, vagus nerve and hypoglossal nerve.

An alternative to surgery, intralesional sclerotherapy in macrocystic lymphatic malformation, is effective and reduces the need for other forms of therapy for some cases. Intralesionalsclerosing agent like bleomycin, OK-432, boiled water, quinine, sodium morrhuate, urethane, iodine, dicycline and nitromin are used after aspirating the cystic fluid.

**IV. Conclusion**

The cystic hygromas situated in parapharyngeal region calls for extra attention as it is deep neck spaces and containing important neurovascular structure and per se lymphangioma of parapharyngeal space is rare entity. It is difficult to diagnose it purely on clinical examination. Preoperative imaging plays crucial role in identification of exact origin such lymphatic anomalies. This information can help to delineate the vascular surgical map and avoid complications during surgical exploration. The use of reconstructed CT images and color Doppler sonography are useful to evaluate the neck lesions and analyze their relationship to adjacent anatomic structures. Transcervicaltransmandibular approach provides good exposure of parapharyngeal space and for excision such lesion. Considering rarity of entity cystic hygroma of deep neck space will be interesting reading for all.

**References**


DOI: 10.9790/0853-14842125 www.iosrjournals.org 24 | Page
Parapharyngeal lymphangioma - a case report


