



Case Study

In Toto Excision Of Lymphangioma- A Challenge

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ABSTRACT

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Cystic Lymphangiomas are malformations in the lymphatic system that are uncommon and usually presents within the first two years of life. They are extremely rare in adults with a very few reported cases. They are usually located in the head and neck especially in the posterior triangle of the neck, presenting as a painless fluctuant transilluminant mass. In adults trauma and upper respiratory tract infection has been suggested as a trigger for its onset. It can clinically mimic a variety of other conditions, hence imaging and histopathology is crucial to its diagnosis. The treatment of choice remains complete surgical resection though other alternative modalities like sclerotherapy are available. However due to its location and extent a complete surgical removal is often impossible. We report a case series (4 cases) of cystic hygroma, all above the paediatric age group, its varied presentation; the diagnostic as well as intraoperative challenges faced and tackled in its management.

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INTRODUCTION

Cystic swellings in the neck have a wide range of differentials ranging from benign to malignant; common to rare conditions. Cystic hygroma is a rare differential in adults hence posing a diagnostic as well as a surgical challenge for the clinician^(1,2,3). It is believed to arise from early sequestration of the embryonic lymphatic channels. Most of such cases are diagnosed before 2 years of age owing to main period of lymphatic growth⁽³⁾; However its prevalence in adults is such that literature even in the beginning of the twenty first century suggest less than a hundred reported cases⁽⁴⁾. We are reporting a case series of four patients above the paediatric age group, with cystic hygroma of variable extension who had presented to us in a period of 6 months and the challenges in its surgical removal because of its location and extent with due importance in removing the mass in toto with preservation of vital structures close to its vicinity.

CASE 1

32 year old male presented with right sided neck swelling since 1 year, insidious in onset, progressively increasing in size (Fig 1.a). On examination there was right sided supraclavicular single swelling 10 X10 X8 cm, fluctuant and transilluminant (Fig 1. d). CT scan was suggestive of a large well defined non enhancing cystic lesion in right posterior cervical space (Fig 1.b). Fine needle aspiration cytology (FNAC) report was suggestive of Lymphangioma.

Intraoperatively Mass seen extending in the posterior triangle going just below the clavicle, medially to the carotid and internal jugular vein, laterally till the trapezius, posteriorly till the constrictor muscles (Fig 1.c). Mass dissected and removed in-toto (Fig 1. e). Corrugated drain placed till 10 days and removed. Intraoperative and postoperative period was uneventful. No recurrence.

1) Fig1.a- Showing right sided neck swelling.

2) Fig 1.b -CT scan showing a large well defined non enhancing cystic lesion in right Posterior cervical space.

3) Fig1.c- Showing intraoperatively Mass extending in the posterior triangle going just below The clavicle, medially to the carotid and internal jugular vein, laterally till the trapezius, posteriorly till the constrictor muscles.

4) Fig 1.d -Showing right sided supraclavicular single swelling fluctuant and transilluminant

5) Fig 1.e- Showing dissected and removed in-toto Mass



Fig 1.a

Fig 1.b

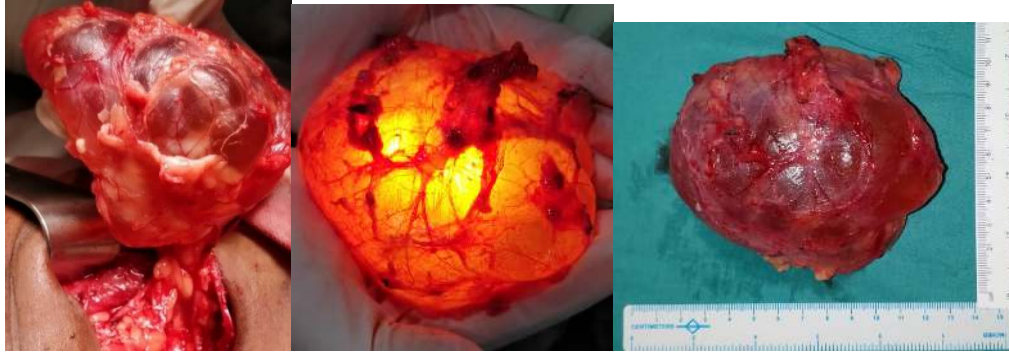


Fig 1.c

Fig 1. d

Fig 1. e

Case 2

14 years old male came with complaints of Right sided anterior neck swelling, insidious in onset, gradually progressing in size, painless, Size 8 x 12 cm soft in consistency extending from mentum till 1 finger breadth above the clavicle, laterally till the posterior border of sternocleidomastoid muscle medially till midline (Fig 2 a), Transilluminant, transmitted pulsations present. CT scan was suggestive of cystic lesion arising from lateral wall of nasopharynx extending to right parapharyngeal area, abutting right Common carotid artery, medially to thyroid suggestive of cystic hygroma (Fig 2 b). Fine needle aspiration cytology (FNAC) report was suggestive of Right side cystic lesion.

Surgical excision was done. Mass dissected from all around, separated from the anterior border of the sternocleidomastoid, inferiorly extending just above the clavicle (Fig 2.d), superiorly extending uptill pharynx (Fig 2. C). Mass excised after separation from the submandibular gland (Fig 2.f). The extension into pharynx was marsupialised and walls sutured to the adjacent structures (Fig 2 .e).Corrugated drain inserted and was removed on post operative day 14.Glove drain reinserted on POD16, in view of collection, Neomycin instilled. Symptomatically improved , hence discharged after a week following drain removal. Patient followed up every month postoperatively for five months . He recovered without any morbidity.

Fig 2 a- Showing Right sided anterior cystic neck swelling

Fig 2 b- CT scan showing of cystic lesion arising from lateral wall of nasopharynx

Extending to right parapharyngeal area, abutting right Common carotid artery, Medially to thyroid

Fig 2 c- showing Surgical excision of mass extending uptill superiorly pharynx.

Fig 2 d - Mass dissected from all around, separated from the anterior border of the Sternocleidomastoid, inferiorly extending just above the clavicle

Fig 2 e- Showing extension into pharynx was marsupialised and walls sutured to the Adjacent structures

Fig 2 f- showing Mass excised after separation from the submandibular gland

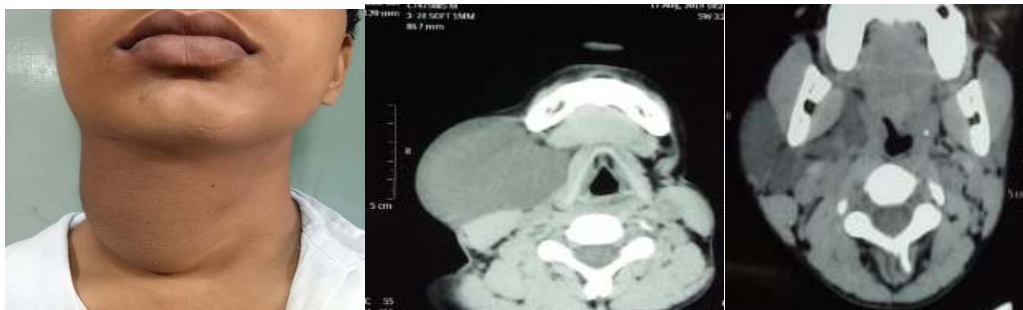


Fig 2 a

Fig 2 b



Fig 2. C



Fig 2.d



Fig 2 .e



Fig .2.f

Case 3

24 year old male presented with right sided neck swelling since 10 to 12 months , insidious in onset, progressing to chest and axilla (Fig 3 a). On examination two separate swelling , one in right supraclavicular region soft , 8x6 cm, non tender, fluctuant, crossing midline and extending below the clavicle. Another swelling 10 x 6 cm soft to firm in consistency, in right axilla, transilluminant with engorged veins. CT Scan and MRI done, suggestive of well defined thin walled non enhancing cystic lesion 12.4x6.6x2.9 cm in right lower posterior cervical space, extending into infraclavicular space and axilla (Fig 3 b).

The surgical resection was done with the assistance of cardiothoracic team. Intraoperatively Neck crease incision taken over supraclavicular region, separate vertical incision taken in axillary region. Large cystic swelling seen extending infraclavicularly towards right axilla (Fig 3 c). Mass separated from surrounding structures including right Internal Jugular vein, carotid artery and subclavian vessels, brachial plexus, axillary vessels. Complete excision of mass done and delivered through supraclavicular incision in toto (Fig 3 d). Brachial plexus injury identified during supraclavicular dissection through neck incision. Exploration done by plastic surgeon and incision extended to join

axillary and neck incision. Cut ends of branches from upper trunk identified (Branch to suprascapular) and Primary epineural repair was done .Post operatively advised shoulder immobilization in adducted position with elbow

physiotherapy for two weeks. Postoperative Histopathology was consistent with cystic hygroma. Patient followed for 6 month postoperative. There was no recurrence of swelling and no gross restriction of shoulder movements.

Fig 3.a – Showing right sided neck swelling since progressing to chest and axilla.

Fig 3.b- CT scan and MRI showing non enhancing cystic lesion in right lower posterior Cervical space, extending into infraclavicular space and axilla

Fig 3.c- Showing intraoperatively Neck crease incision taken over supraclavicular region, Separate vertical incision taken in axillary region & large cystic swelling seen Extending infraclavicular region.

Fig 3.d- Showing Complete excision of mass done and delivered through supraclavicular Incision in toto



Fig 3.a

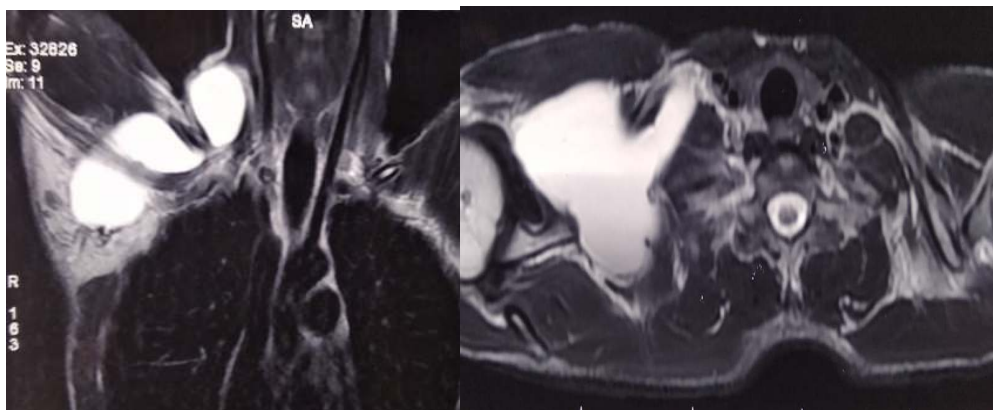


Fig 3 b



Fig 3 c



Fig 3 d

Case 4

30 year old male presented with right sided neck swelling of 7- 8 months duration, insidious in onset, gradually increasing. On examination there was swelling 7x5x3 cm in the right submandibular region, non tender, and fluctuant (Fig 4a).

Fine needle aspiration cytology was suggestive of benign cystic lesion consistent with lymphangioma, CT scan showed well defined homogenous isodense mildly enhancing soft tissue lesion 6x2.4x2.4cm within right submandibular region, lateral to submandibular gland abutting and displacing gland posteriorly (Fig 4 b), Suggestive of lymphatic collection/cyst. USG was suggestive of

Fig 4a – Showing cystic swelling in the right submandibular region

Fig 4b- CT scan showed well defined homogenous isodense mildly enhancing soft tissue Lesion within right submandibular region, lateral to submandibular gland abutting And displacing gland posteriorly

Fig 4c- Showing Intraoperatively neck incision given 2 finger breadths below angle of Mandible over maximum bulge of swelling

Fig 4d - Showing large cystic swelling extending toward oral cavity, suspecting Plunging ranula

Fig 4e- Showing Complete surgical excision of neck mass done with marsupilisation in the floor of mouth

Collection/cyst measuring 38x37 mm in right submandibular region extending to the floor of mouth.

Intraoperatively neck incision given 2 finger breadth below angle of mandible over maximum bulge of swelling (Fig 4 c), large cystic swelling extending toward oral cavity (suspecting- plunging ranula, Fig 4 d).

Complete surgical excision of neck mass done (Fig 4e) with marsupilisation in the floor of mouth . Glove drain placed. Oral Glycopyrrolate given postoperatively. Wound healed well. Histopathology report confirmed lymphangioma. And no recurrence on 6 month postoperative follow up.



Fig 4a



Fig 4 b



Fig 4 c



Fig 4 d

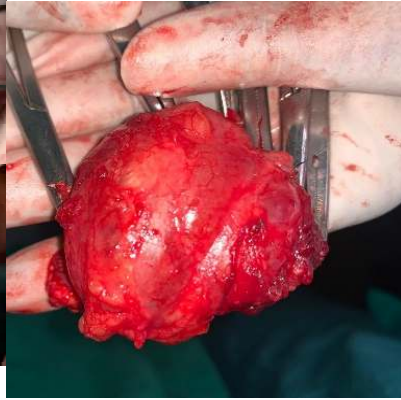


Fig 4e

DISCUSSION

There are many differentials for cystic lesions of Neck ranging from branchial cyst, thyroid cyst, thyroglossal cyst, dermoid or epidermoid cyst, to metastatic squamous cell carcinoma. Lymphangiomas are considered rare and occur mainly in childhood⁽⁵⁾. Because of its very low incidence, most surgeons are unable to gain much personal experience with this intriguing lesion⁽⁶⁾.

Lymphangiomas are benign congenital tumor of lymphatic origin, with cystic spaces lined with true endothelium, classified as microcystic (capillary lymphangiomas), macrocystic (cavernous lymphangiomas) and cystic hygromas^(7, 8). Cystic hygromas were first described by Redenbacher in 1828. The name "Cystic Hygroma" was devised by Werner in 1834^(9, 10, 11). They are composed of cysts and sinuses, containing eosinophilic acellular lymph fluid

The most widely accepted theory about the development of cystic lymphangioma is that they arise from sequestrations of the primitive embryonic lymph sacs⁽⁵⁾. 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⁽⁶⁾. However

there was no history of the same in our patients.

These relatively uncommon malformations present as painless mass cystic to palpation, transilluminant generally involving the head and neck region⁽¹²⁾. As with in all our cases. All our cases were males above the age of twelve, although most of the studies suggest no gender predilection. The most common documented site is the neck. Particularly in the posterior triangle⁽⁴⁾. All our four cases involved different regions of head and neck. Two cases involved the posterior triangle. In case 3 the neck lesion extended in the subclavicular plane to the axilla and chest. And in the other two cases it involved the anterior triangle; however case 2 had extension till nasopharynx.

In children cervical lesions can cause dysphagia and airway obstruction, however, this is rare in adults⁽¹³⁾. There were no obstructive symptoms in any of our patients. Lymphangiomas may remain static or involutes, but in some cases they may increase in size, especially after internal haemorrhage or infection, can grow rapidly potentially leading to life-threatening airway compromise or obstruction. They also have tendency to infiltrate into and around muscles, vital nerves and vessels⁽⁶⁾

The diagnosis is usually made on clinical grounds, but CT and MRI scanning will

more accurately determine the size, the exact anatomical location, its relationship with important structures and aid the surgical planning⁽¹⁴⁾. CT scan was done in all our cases. MRI done in the case which had extension upto axilla. It is essential to evaluate intrathoracic extension of cystic hygroma, which might be seen in 10% of cases, and this is done by USG/CT/Magnetic Resonant Imaging (MRI)⁽¹⁵⁾ There is no consensus in literature concerning the use of Fine needle aspiration cytology (FNAC) to diagnose these lesions, However there are a few reported cases where FNAC proved to be an important diagnostic tool⁽¹⁶⁾. FNAC done in all the cases. suggestive of lymphangioma except in case 2 which was just reported as a cystic lesion. FNAC together with imaging proved to be beneficial. Despite all the advanced imaging techniques, the diagnosis of adult lymphangiomas remains a challenge. A correct diagnosis is ensured only by histopathological examination of the surgical specimen.

There can also be intraoperative surprises. As in one of our case (case 4) there was sublingual extension of the lesion, hence mimicking a plunging ranula intraoperatively, however later proved histopathologically as lymphangioma. Such Rare instances were cases of sublingual lymphangioma mimicking a ranula has also been reported⁽¹⁷⁾.

Though many methods of treatment like aspiration, sclerotherapy have been described over the years. Surgical excision remains the treatment of choice, but it is challenging and it should be therefore, best undertaken by experienced surgeons in specialist centres. The goal should be complete excision with good cosmesis⁽⁹⁾. Although benign, histology must be undertaken for affirmative diagnosis. In adult patients, this neoplasm can even switch to squamous cell carcinoma^(8, 18). Hence the need for its early intervention

Surgery can be helped by the injection of tissue blue into the lymphatic spaces⁽¹⁹⁾.

Though All our cases underwent appropriate surgical excision without the injection of tissue blue. Total tumor removal is the method of choice if it can be achieved. Subtotal removal is defensible in extensive tumor⁽¹⁸⁾. Partial removal of lymphangiomas is proved unreliable and is considered adequate only for lymphangiomas involving tongue, pharynx, and larynx. In two of our cases total removal was not possible, in case 2 and 4 parts extending into pharynx and to the floor of mouth was marsupialized respectively.

DE serres had proposed a staging system which includes laterality ("uni" or "bi") in addition to relationship to the hyoid ("infra" or "supra") as a reliable way of predicting outcome in patients with lymphatic malformations of the head and neck. All our 4 cases were unilateral. Two cases which extended to suprahyoid region required marsupialisation⁽²⁰⁾

Intraoperatively all the cases had variable extension and different ease of surgery. One case had intraoperative complication of nerve injury (suprascapular nerve) which was identified on table, and repaired by plastic surgeons. The patient had undergone physiotherapy sessions. Complete surgical removal with no recurrence till date along with normal shoulder function was achieved with the help of multidisciplinary team work.

Postoperatively all our cases received antibiotics and in case 3 which extended to floor of mouth oral Glycopyrrolate given postoperatively in order to reduce salivation thus preventing orocutaneous fistula formation. There are reported cases of wound infections, respiratory infection post excision hence antibiotic prophylaxis is necessary.

CONCLUSION

Lymphangioma being an uncommon pathology in adults needs specific emphasis. Though there is no gender predilection described in literature, all our cases were males. Our case series also helps

in concluding that head and neck lymphangiomas may arise spontaneously in adults even without significant history of trauma or exertion. Though classically described as being common in the posterior triangle, our cases had varied anatomy ranging from a small mass confined to the posterior or even the anterior neck triangle to a one that can even extent to the pharynx or uptill the axilla. Histopathology is crucial to its diagnosis as it can mimic a variety of conditions. Though complete surgical excision is the treatment of choice, the surgical accessibility to its complete removal might not always be possible due to its anatomical location, and it often remains a challenge for the surgeon to remove the mass intoto preserving the vital structures around.

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REFERENCES

1. Childress ME, Baker CP, Samson PC. Lymphangioma of the mediastinum; report of a case With review of the literature. *J Thorac Surg* 1956; 31:338–348
2. Dowd CN. XI. Hygroma cysticum colli: its structure and etiology. *Ann Surg* 1913; 58:112–132
3. Studies, B. C., Aydin, S., & Go, M. (2015). A Giant Lymphangioma on the Neck. 26(4), 323–325. *The Journal of Craniofacial Surgery* Volume 26, Number 4, June 2015
4. Sherman, B. E., & Kendall, K. (2001). Case Reports A Unique Case of the Rapid Onset of a Large Cystic Hygroma in the Adult. 22(3), 206–210.
5. Kraus J, Plzák J, Bruschini R, Renne G, Andrlé J, Ansarin M, Betka J. Cystic lymphangioma of the neck in adults: a report of three cases. *Wiener klinische Wochenschrift*. 2008 Apr 1; 120(7-8):242.
6. Jack E Brooks M.D. Cystic hygroma of the neck *The Laryngoscope*/Volume 83, Issue 1, January 1973
7. Sichel Jy, Udassin r, Gozal D, Koplewitz BZ, Dano i, Eliashar r. OK-432 therapy for cervical lymphangioma. *Laryngoscope* 2004; 114:1805-9
8. Grasso DL, Pelizzo G, Zocconi E, Schleef J. Lymphangiomas of the head and neck in children. *Acta otorhinolaryngologica italica*. 2008 Feb; 28(1):17.
9. Azim MT, Hussain SM, Mughal MA. Primary Supraclavicular Cystic Lymphangioma in an Adult: A Rare Presentation. *Journal of the College of Physicians and Surgeons—Pakistan: JCPSP*. 2019 Jun; 29(6):S11-2.
10. Nadour K, Moujahid M. Cervicothoracic cystic lymphangioma: about a case. *Pan Afr Med J* 2016; 25:189.
11. Bhatia V, Taksande R, Sondankar D. Cystic Hygroma in an Adult Male: A Diagnostic Challenge. *Iran J Pathol* 2015; 10: 310-3.
12. Valletti PA, Brucoli M, Boffano P, Benech A, Toso A, Dell’Era V, Garzaro M. A single center experience in the management of head and neck lymphangiomas. *Oral and Maxillofacial Surgery*. 2020 Mar; 24(1):109-15.
13. Mathew M, Dil SK. Adult lymphangioma—a rare entity: a report of two cases. *Turkish Journal of Pathology*. 2012 Jan 1; 28(1):80-2.
14. Ricard Simo And Jean-Pierre Jeannon. Benign Neck Disease Watkinson J, Gilbert R. Stell & Maran's textbook of head and neck surgery and oncology. 5th edition, London Hodder & Stoughton Ltd, 2012, Chapter 12:218
15. Morley SE, Ramesar KC, Macleod DA. Cystic hygroma in an adult: A case report. *J R Coll Surg Edinb* 1999; 44:57-8.
16. Permeet K Bagga et all Cervical cystic lymphangioma in an adult ,diagnosed on FNAC *Journal of cytology* Vol.26, No.4, October- December, 2009, pp. 164-165,
17. Marco Nisi et al Sublingyal Lymphangioma mimicking a Ranula: A

- case report. proceedings mdpi journal 2019; 35, 36
18. Berry JA, Wolf JS, Gray WC. Squamous cell carcinoma arising in a lymphangioma of the tongue. *Otolaryngology Head Neck Surg* 2002; 127:458-60.
 19. Riechelmann H, Muehlhaff G, Keck T, Mattfeldt T, Rettinger G. Total, subtotal, and partial surgical removal of cervicofacial lymphangiomas. *Archives of Otolaryngology– Head & Neck Surgery*. 1999 Jun 1; 125(6):643-8.
 20. Lianne M De Serres et al Lymphatic Malformations of the Head and Neck A Proposal for Staging Arch Otolaryngol Head Neck Surg. 1995; 121:577-582
 21. Ameh EA, Nmadu PT. Cervical cystic hygroma: pre-, intra-, and post-operative morbidity and mortality in Zaria, Nigeria. *Pediatric surgery international*. 2001 Jul 1; 17(5-6):342-3

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