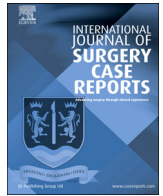




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Extensive parapharyngeal lymphangioma in a adult—Case report and review of literature

Pradeep Balineni^{a,*}, Venkata Pavan Kumar C.H.^a, Debaraju Reddy E.^a, Janani S. Reddy^b^a Department of Surgery, SLG Hospitals, Hyderabad, India^b Department of Surgery, Panimalar Hospitals, Chennai, India

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ABSTRACT

INTRODUCTION: Lymphangiomas are benign swellings most commonly seen in paediatric population. Head and neck are the common sites of presentation rarely can also be seen other sites as axilla, chest, abdomen, liver, spleen. Even though many different modalities have come up surgical excision remains the main stay of treatment.

CASE REPORT: A 24 year male patient presented with complaints of a swelling the neck since 2 months, which was 8 × 5 cms, soft, cystic and transilluminant in the anterior triangle of the neck. MRI study showed features suggestive of a para pharyngeal lymphangioma. Patient had undergone surgical excision through trans mandibular trans cervical approach for extra cranial part and for intra cranial part has got OK-432 sclerosant therapy.

DISCUSSION: Lymphangiomas are benign, hamartomatous swellings. They are mostly noticed due to their large size causing disfigurement, dyspnoea or dysphagia. They are diagnosed prenatally by using ultrasound scanning. MRI is the superior imaging technique to diagnose lymphangiomas. Surgical excision is the mainstay of treatment, other treatment modalities as sclerotherapy, laser excision, radiotherapy, unroofing, aspiration or steroids are also used.

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1. Introduction

Ever since its first description in European literature by Redenbacher in 1828 [1] there were many reported cases of cystic hygroma in paediatric population, and rarely in adults. Lymphangiomas make up to 6 % of the benign swellings in patients younger than 20 years [2]. Head and neck are the most common sites of lymphangioma; however para pharyngeal lymphangiomas are exceedingly rare. Other less common sites are axilla, mediastinum, groin and abdominal cavity [1,3]. There are three theories which are popular regarding the aetiology of the lymphangioma which are arrest of the normal growth of primitive lymph channels, lymph channels not reaching up to the venous system and finally which emphasizes lymph channels getting laid in wrong areas during embryogenesis [4].

They mostly present within 2 years of age as a asymptomatic neck mass. The mainstay of treatment for these lesions remain to be surgical resection, other modalities of treatment are aspiration, steroids, sclerotherapy, radiotherapy, laser excision and cauterization. Here we report a rare case of a parapharyngeal lymphangioma

in an adult which is rare site and age group of presentation. This case report is reported in lines with the scare guidelines [5].

2. Case report

A 24 years male presented to the surgical department with complaints of a swelling in the neck since 2 month, and features of upper respiratory tract infection. Patient did not notice any swelling prior to this episode and had no history of pain or fever. Patient had no features of respiratory obstruction or difficulty on swallowing. On examination patient had a 8 × 5 cms soft, cystic, non-tender swelling which is transilluminant in the anterior triangle of the neck, swelling had reduced mobility and does not move on deglutition (Fig. 1).

Ultrasound examination of neck revealed a large multi loculated cystic swelling with thin internal septations, MRI imaging showed heterogeneous lesion which was low intensity on T1 and high intensity on T2 in the Para pharyngeal space extending below along the anterior border of the sternocleidomastoid muscle up to supra-sternal level (Fig. 2) suggestive of a lymphangioma.

Patient had undergone a transcervical surgical excision of the cervical part of lymphangioma up to the superior border of sub mandibular gland (Figs. 3 and 4 with intra lesional picibanil (OK 432) injection to the intra cranial part in view of not disturbing any vital structures for excision of the intracranial part. Post oper-

* Corresponding author at: Flat no 73, Srr Ridge Villas, Nizampet Road, Bachupally, Hyderabad, Telangana, 500090, India.

E-mail address: pradeep052191@gmail.com (P. Balineni).



Fig. 1. Showing swelling.

actively patient was started on liquid diet followed by soft diet on post-operative day 1 and was discharged on post-operative day 2. Post-operative pathological report showed multi-lobulated, thin walled, lymph containing sac (Fig. 5). Patient had no complications or recurrence after 1 year of follow up period.

3. Discussion

Lymphangiomas are benign, hamartomatous, congenital malformations of the lymphatic system [3]. Incidence of these lesions is reported differently in various studies ranging from 1.2 to 2.8 per 1000 new births and 1 in 750 spontaneous abortions, Gupta et al., reported it to 1 per 1000 new births and Nicholls et al., reported it to be as 1 in 6500 live births [3]. 80–90 % of cases present with in the first 5 years of age mostly in first 2 years of life, with no gender or racial discrimination [1,4].

Lymphangiomas are usually classified as capillary lymphangioma (simplex), cavernous lymphangioma, and cystic hygromas a term first used by Wernher in 1843 [3,6]. Those swellings which develop in dense tissues present as cavernous lymphangioma and those in loose tissues develop as cystic swelling [4]. Cystic hygroma are the most common type of lymphangioma accounting up to 90 %, Bill and summer in 1965 proposed the concept that both cystic hygroma and lymphangioma are variations of a single entity [2,4,6]. They are also classified based on the size of cyst as microcystic with cysts of size less than 2 cms and macrocystic with cysts of size more than 2 cms [3].

Mostly lymphangiomas develop in the neck (up to 80 %) region followed by axilla and extremities, other rare sites of presenta-

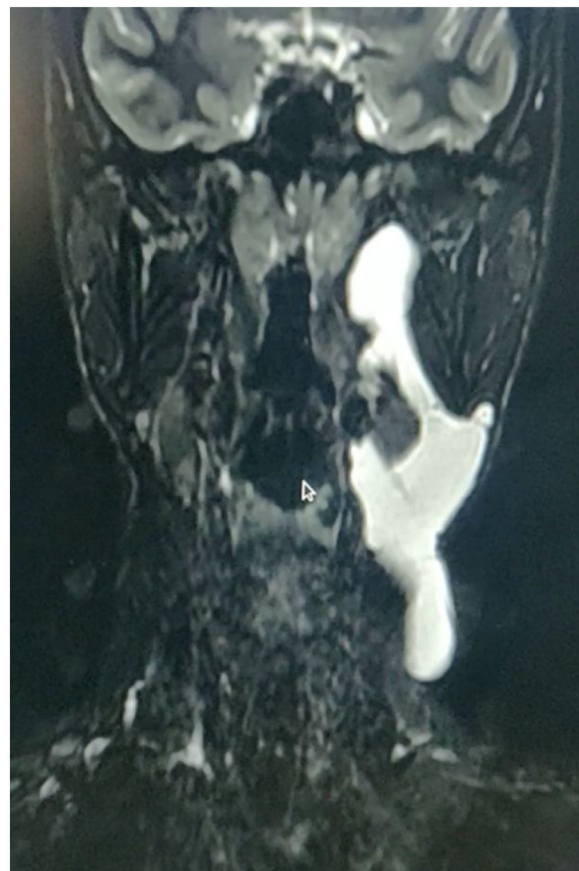


Fig. 2. MRI image showing lesion.

tion are liver, spleen, kidney, intestine and etc3. Around 85 % of cervical lymphangiomas are unilateral [3]. They mostly present as asymptomatic mass of neck and are recognized due to the massive size [4]. The common symptoms of presentation are due to obstruction to the respiratory tract causing respiratory distress in around 11–27 % [2,4,7] and difficulty in swallowing, and pain due to infection or haemorrhage which are the most common complications [1,4]. Although they are benign and have no malignant potential [1] they grow and compress the nearby structures and infiltrate the vital structures, which is also a poor prognostic sign [2]. Clinically they are large, cystic, non-tender and transilluminant swellings unless they are complicated by haemorrhage and infection [1,3,6]. These are known to be associated with trisomies of 13,18,21, Turner syndrome, Noonan syndrome, aortic coarctation hypoplastic left heart syndrome, pentology of Cantrell, Apert syndrome, Cornelia deLange syndrome, foetal alcohol syndrome, Fryns syndrome, lethal multiple pterygium syndrome, limb hypertrophy, Pena Shokeir syndrome and hydrops [1,6].

Pre-operative imaging is essential to know the origin and the extent of the lesion. Differential diagnosis would be lymphangectasia, hemangioma, lipoma. Lymphangiomas can be diagnosed prenatally by ultrasound [3]. They are seen as thin walled multiloculated cystic swelling on ultrasound. Computed tomography (CT) is better compared to ultrasound on which they are seen as low attenuation masses depending on the nature of cystic content with enveloping effect around the trachea, oesophagus, and great vessels; they also have an enhancing rim after intravenous contrast [3,7]. CT also helps in determining the extent of the swelling and differentiates it from nearby vital structures. MR imaging is superior in identifying these lesions on which they show up as low signal intensity to muscles on T1 and high intensity to fat on T2 [3,6,7].

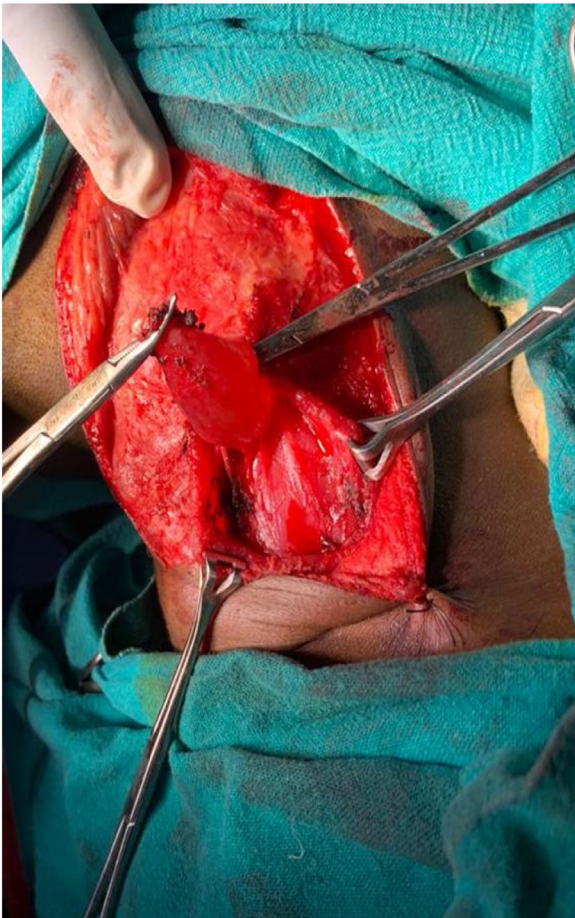


Fig. 3. showing lesion over sternocleidomastoid.



Fig. 4. showing posterior bed of lesion with submandibular gland.

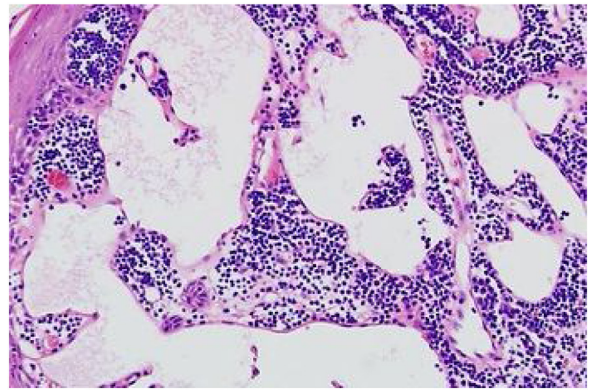


Fig. 5. Pathology slide of lymphangioma.

Pathologically they are multilobulated, fluid filled sacs and thin walled with one or two endothelial layers containing fluid being milky, sero sanguineous or straw colour⁹. Indications of treatment are large sizes, disfigurement, recurrent infections, dysphagia or respiratory distress. Although instances of spontaneous regression are also reported to be 15 % in the literature [6,8]. Mainstay of treatment would be surgical excision of the cyst [9,10], and the other modalities are laser excision, radiotherapy, sclerosant injection, unroofing, aspiration or steroids [3,7]. Sclerosants can be injected both intra lesional (in macrocystic type) or peri lesional, different type of sclerosants used are OK-432, bleomycin, monoclonal antibody, boiled water, quinine, sodium morrhuate, urethane, doxycycline, iodine tincture, tetracycline and nitromin1 [3,4].

Fever, local swelling and hyperemia may be noted due to the adverse effects of sclerotherapy⁷. Okazaki et.al recommended OK-432 treatment alone for single macrocystic type, and pre surgical injection of OK-432 for microcystic type [3,4,6]. Main advantage of using OK-432 is the absence of perilesional fibrosis [4,11]. Zarod et.al has also demonstrated good results by using sclerotherapy with dehydrated alcohol injection [7].

The type and approach for surgical excision depends on the type and location of the cyst. Transcervical transmandibular approach would give a complete access to the parapharyngeal space [1]. Patients may have persistent airway and difficulty in swallowing post surgically due to mucosal edema, enlargement of internal lymphangioma or loss of neural innervation to pharynx [4]. Classification of lymphangioma described by Schuster et al. provide an insight of the post-surgical results and complications, they are type 1 which had no or minimal effect on contour of neck, type 2 are smaller than a line drawn at the lateral border of the head, type 3 extends this line, type 4 extends beyond midline of the body [4]. Recurrence is usually noticed after 1 year of subtotal excision due to regeneration of the lymphatic channels [7]. Recurrence rate of the lesion depends on the complexity of the lesion and also the completeness of excision, in case of simple lesions which are completely excised recurrence rate is almost 0%, in case of complex lesions with partial excision it is 10–27 % and for complete excision it is 50–100 % [3,4]. Gutierrez et.al demonstrated good results with fibrin adhesive treatment in cases of recurrences, some authors also reported notable effect with streptococcal injections in recurrence [2,7].

4. Conclusion

Lymphangioma is a benign swelling mostly in the head and neck regions of paediatric population. They are presented due to the obstruction effect of their massive size or complicated due to infection or haemorrhage. MRI is the better choice of investigation for these lesions over USG or CT. Surgical excision is the mainstay of

treatment though other modalities of treatment are available. OK-432 is a better agent for intralesional sclerotherapy as it has low peri lesional fibrosis.

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Exempted from ethical committee but, informed and written consent from the patient is obtained.

Consent

Informed and written consent was obtained from the patient

Author contribution

Dr. Pradeep Balineni – Data collection, Manuscript structure.
Dr. Pavan Kumar – Data collection, Manuscript writing.
Dr. Debaraju Reddy - review of literature, manuscript analyses.
Dr. Janani Reddy – review of literature, Manuscript writing.

Registration of research studies

- 1 Name of the registry:
- 2 Unique identifying number or registration ID:
- 3 Hyperlink to your specific registration (must be publicly accessible and will be checked):

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Declaration of Competing Interest

The authors report no declarations of interest.

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