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Transoral robotic surgery for adult parapharyngeal lymphangioma: A case report

TORS for adult PPS lymphangioma

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Abstract

BACKGROUND

Lymphangiomas are a group of benign swellings which are commonly seen in the paediatric age group. The most common sites of presentation is in the head and neck region , less commonly seen in axilla , chest , liver , spleen *etc.* The ideal modality of treatment has always been surgical excision irrespective of the site and age group. But with the advent of minimally invasive surgical techniques, it is now possible to perform excision of parapharyngeal space lesions with minimal morbidity and good clearance

CASE SUMMARY

42 year old male patient who presented with complains of difficulty in swallowing and had undergone surgery twice outside where in Transcervical approach was attempted to remove the parapharyngeal mass which had failed. MRI scan demonstrated a 6x5x4 cm left parapharyngeal mass. He underwent transoral robotic surgery for the excision of the parapharyngeal mass lesion and had an uneventful post-operative recovery period.

CONCLUSION

Lymphangiomas are hamartomatous swellings which are benign in nature. The presenting symptoms of the patient with large parapharyngeal mass includes dysphagia, dyspnoea and neck swelling. Clinicoradiological evaluation is of utmost importance in such cases to determine the relation adjacent vital structures and also to determine the approach the tumour. With the advent of robotics in oncology, transoral robotic excision is one of the best approaches to perform such a surgery provided , patient, surgeon and anatomical factors favour the same.

Key Words: lymphangioma; parapharyngeal space lesions; Transoral robotic surgery for PPS mass , case report

Dabas S, Menon NN, Ranjan R, Gurung B, Shukla H, Sharma AK, Tiwari S, Sinha A, Bhatti SS, Sangal R. Transoral robotic surgery for a rare case of adult parapharyngeal lymphangioma. *World J Otorhinolaryngol* 2022; In press

Core Tip: This is an article which portrays adult parapharyngeal space lymphangioma which is a rare case. It also emphasizes on the role of minimally invasive techniques like transoral robotic surgery for the excision of parapharyngeal space tumours which has made surgical excisions less morbid for the patients and gives good clearance of disease.

INTRODUCTION

Cystic Hygroma also known as cystic Lymphangioma, jugular lymphatic obstructive sequence and hygromacollcysticum was first described in 1828 by Redenbacher^[1]. Cystic hygroma is also known as. Only 6% of benign lumps in patients less than 20 years are attributable to lymphangiomas^[2]. Parapharyngeal space is an extremely rare subsite for lymphangiomas in head and neck. Other sites include axilla, mediastinum, groin and abdominal cavity^[3].

Different etiological and pathogenic factors have been described as causal, including traumatic, infectious and chronic inflammatory factors, as well as alterations in embryological development^[4]. In adults, late proliferation of cellular nests of the lymphatic system may appear due to stimulation caused by trauma or infection^[5].

Lymphangiomas in the head and neck region commonly develop as an asymptomatic neck mass around 2 years of age.

The imaging modality preferred for lymphangiomas include Ultrasound [USG] supplemented by CT and MRI scans for obtaining more information about the structural features, internal as well as peripheral enhancement patterns^[6].

The most common treatment modality includes surgical excision of the mass. The other treatment modalities include aspiration, sclerotherapy, steroids, laser and radiotherapy.

This case describes a rare case of adult parapharyngeal space lymphangioma which was managed with transoral robotic surgery.

CASE PRESENTATION

Chief complaints

42 year old male patient presented to our outpatient department with chief complaint of difficulty in swallowing both solids and liquids for the past 2 months.

History of present illness

Patient had complain of dysphagia for past 2 months with no associated odynophagia , respiratory difficulty or neck mass. He gives history of undergoing Transcervical surgery twice outside for the same issue but which failed to remove the concerned pathology. No surgical records of the previous surgeries were available at the time he presented to us. He did not complain of any neck swelling or respiratory difficulty.

History of past illness

Patient had no history of any significant co morbidities.

Personal history

No significant history noted.

Family history

No significant history noted.

Clinical examination

Vitals recorded were as follows , temperature 36.5 degree Celsius , pulse rate 88/ minute , blood pressure 130/80 mmHg and respiratory rate of 18/minute. On clinical

examination, a submucosal bulge was noted in left tonsillar area. No swelling was however palpable in the neck.

Laboratory examinations

Routine blood investigations were done as part of pre-operative work up which were within normal limits.

Imaging examinations

A fiberoptic laryngoscopy (FOL) was performed showing a bulge in the region of the left lateral oropharyngeal wall with significant narrowing of the airway. An MRI scan was performed which showed a 6x5x4 cm lobulated heterogeneous lesion involving left lateral oropharyngeal wall, parapharyngeal space and peritonsillar area (Figure 1). The lesion compressed and displaced the left palatine tonsil anteroinferiorly, laterally reached the deep lobe of parotid gland, abuts medial pterygoid muscle anterolaterally and indents longus coli muscle posteriorly, compressing the external carotid artery. Post-surgical changes were noted in the region of the left sub mandibular gland and anterior to the sternocleidomastoid muscle.

FINAL DIAGNOSIS

The main differential diagnosis were vascular malformation and lymphoreticular malformation.

TREATMENT

The patient was planned for upfront transoral robotic surgery [TORS] after obtaining anaesthesia clearance. A written and informed consent was taken from the patient for the procedure to be performed. The procedure was done under general anaesthesia. FK retractor was placed and da Vinci Xi robotic system was docked. 3 arms of the robot were used for performing the surgery which contained the camera, bipolar and monopolar cautery. After giving incision in the peritonsillar area, layer by layer dissection was done

of the mucosa, submucosa and constrictor muscles with adequate hemostasis at each step. A well encapsulated lesion was identified in the left parapharyngeal space which was excised (Figure 2) in toto and specimen sent for histopathological examination. They were no intra or peri operative complications. The final histopathological report showed a multilobulated lesion with lymph containing sac and the report was of lymphangioma (Figure 3).

OUTCOME AND FOLLOW-UP

Post operatively the patient was kept in the hospital for 3 days and was put on antibiotics and analgesics. For the initial 1 week, patient was on Ryle's tube feed. Post operatively, patient came for first follow up to the outpatient department on day 7, FOL was done which showed a healed wound. Patient's speech and swallowing was evaluated by our speech and swallowing specialist. The speech of the patient was normal and he swallowing was also unimpaired. He was then started on liquids followed by soft diet. Normal diet was started on day 15 after surgery. Patient has been on regular follow up since then for the past 6 mo and has no signs of residual disease, recurrence or any other post-operative complications.

DISCUSSION

Lymphangiomas are defined as benign, congenital and hamartomatous lesions which develop due to abnormalities in the lymphatic system. 1.2 -2.8 per 1000 new births is the incidence noted in literature^[4]. First 5 years of life show the highest incidence of lymphangiomas [80-90%]^[7].

Histologically , lymphangiomas are classified into capillary, cavernous and cystic hygromas^[8]. Based on size it is divided into microcystic [<2cm] and macrocystic [>2cm]^[3]. Bhayya *et al* classified them as lymphangioma simplex, cavernous lymphangioma, cystic lymphangioma and benign lymphangioendothelioma^[9]. In 1995, de Serres *et al* proposed a classification based on anatomical location: Based on anatomical location de Serres et al

[1995] proposed a classification system which was as follows – Class I indicates unilateral infrahyoid lesion ; Class II, unilateral suprahyoid lesion ; Class III, unilateral combined supra and infrahyoid lesion ; Class IV, bilateral suprahyoid lesion and Class V, bilateral combined supra and infrahyoid lesion^[10].

80% lymphangiomas develop in the neck and 85% are unilateral^[4]. Most common presentations include swelling in the neck, respiratory distress, difficulty swallowing and pain secondary to infection or haemorrhage^[7]. Although lymphangiomas are benign lesions with no malignant potential, occasionally it can enlarge to compress and invade the adjacent structures. The latter is a feature of poor prognosis in advanced cases^[2]. Clinically, they usually appear as painless and cystic lumps, which are transilluminant in nature. They are associated with pain only when they are complicated with infection or haemorrhage^[3].

Lymphangiomas are associated with various congenital syndromes which include the following - ¹ trisomies of 13,18,21, Ullrich Turner syndrome, Noonan-Ehmke syndrome, aortic coarctation , hypoplastic left heart syndrome, Cantrell Haller Ravitsch syndrome, Apert syndrome, Brachmann-de Lange syndrome, foetal alcohol spectrum disorders, Fryns syndrome, Escobar syndrome, limb hypertrophy, fetal akinesia deformation sequence and hydrops^[8].

Imaging is an important diagnostic modality for evaluation of the location, site of origin and relation to adjacent structures vital to understand the exact origin, location and extent of lymphangioma. On ultrasound done during the antepartum period, lymphangiomas appear as thin walled multiloculated cysts. CT scan is better compared to USG for imaging lymphangiomas. In CT scans, they appear as low attenuation masses with regular margins and an enhancing rim on intravenous contrast. On T1 weighted images, they appear low signal compared to muscles and on T2 weighted images , they appear hyperintense compared to fat^[4,6].

50% of parapharyngeal space tumours have a salivary cause, 20% have a neurogenic root, and 30% are benign and malignant lymphoreticular lesions, metastatic lesions, carotid body tumours and lipomas. Amongst neurogenic tumours, Schwannomas arising from the vagus nerve, presenting as post styloid compartment lesions are the most common^[11]. On imaging, homogenous lesions seen displacing the internal carotid or common carotid artery anteriorly is suggestive of schwannoma arising from vagus nerve or sympathetic trunk^[12]. Lipomas appear as hyperintense lesions on T1- and T2-weighted images with internal septations^[13]. Vascular tumours are generally more conspicuous on T2-weighted images. Pleomorphic adenomas are usually characterized by the smooth and regular, non infiltrating margins with cystic or necrotic areas^[14]. Paragangliomas have a characteristic “salt and pepper” appearance on MRI scan^[15]. Dermoid and epidermoid cysts are differentiated from other similar lesions as they often appear as unilocular masses with homogeneous hypoattenuating fluid on CT scan^[16].

The selection criteria employed for treatment of lymphangiomas include large size, disfigurement, recurrent infections, dysphagia or respiratory distress. Literature shows spontaneous regression in 15% cases^[8]. Surgery forms the mainstay of treatment^[17]. Other options for treatment include laser excision, radiotherapy, sclerosant injection, unroofing, aspiration or steroids^[4].

The decision on choosing the type of approach required for surgery depends on the site and nature of the lesion. Traditionally, the transcervical transmandibular approach was observed to give good exposure of the parapharyngeal space^[3]. The main disadvantages of this approach include, the dysphagia and persistent respiratory difficulties which the patients face that is secondary to the post-operative mucosal edema and loss of pharyngeal innervation^[7].

Traditionally, surgeons believed that in case of lesions extending to parapharyngeal space complete surgical excision was difficult to obtain without damaging nervous and

vascular structures and thus they used to resort to alternatives like drainage and sclerotherapy with different substances such as tetracycline, bleomycin and triamcinolone and radiofrequency ablation^[18]. Complications and associated morbidity can include infections, tissue necrosis, cranial nerve lesions, vascular thrombosis and even endocrine disorders^[19]. But now, with the advent of robotic surgery these disadvantages have been overcome and it is possible to obtain 100% clearance of lesions in parapharyngeal space without injuring any neural or vascular structure^[20].

A surgical algorithm was proposed for by Chu *et al* in 2017 for access to parapharyngeal space^[21]. (Figure 4)

TORS should be preferred for well circumscribed lesions in the pre- or post-styloid compartment^[21] as suggested by Chu *et al*. Transoral robotic surgery was opted in our case as it gives a magnified and high resolution view, along with motion scaling and tremor filtration which allows for delicate dissection around the tumour capsule. This approach is also less morbid compared to the other traditional approaches and reduces the chances of complications like swallowing problems, trismus, auriculotemporal syndrome, first bite syndrome, marginal mandibular and hypoglossal nerve palsy. Cosmesis achieved with this approach is also excellent compared to the transcervical approaches. The other advantages of using robotic surgery in this case include minimal blood loss, shorter hospital stay and early initiation of oral diet for the patient. Post operatively our patient had no infections, wound dehiscence and we were able to start orally by day 7.

Recurrence is usually noted in the first year following incomplete excision of lymphangiomas due to regeneration of the lymph vessels^[22]. Besides completeness of excision, recurrence rate is based on the type and complexity of the mass. Simple lesions which are completely excised have 0% recurrence rate whereas complex lesions with

partial excision have a recurrence rate of 10–27 % and in those with complete excision, it is 50–100%^[4, 7].

CONCLUSION

Lymphangiomas are benign hamartomatous lesions which rarely present in the parapharyngeal space. Large size, dysphagia, respiratory distress, pain due to recurrent infection and haemorrhage are the main indications warranting treatment. MRI is the ideal imaging modality. Transoral robotic surgery is one of the best approaches to parapharyngeal space lesions with favourable anatomy, with minimal morbidity and excellent clearance.

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