Parapharyngeal Cavernous Lymphangioma - A case report

[1]Mohammed U, [2]Iseh KR, [1]Sahabi SM, [2]Aliyu N, [1]Abdullahi K

[1]Departments of Histopathology and [2]ENT, Usmanu Danfodiyo University Teaching Hospital, Sokoto. Nigeria.

Correspondence: *Mohammed U, Email:umarsambas@yahoo.co.uk*

BACKGROUND

Lymphatic malformations of the head and neck, also known as lymphangiomas or cystic hygromas, are a diverse group of lesion. They represent benign hamartomatous tumours of lymphatic vessels with a marked predilection for the head, neck and oral cavity. Cavernous or microcystic lymphangioma, however, is composed of small lymphatic spaces and poses a therapeutic dilemma by its propensity to cause airway and feeding difficulties and by its tendency to recur despite extensive surgery.

Case report: *We present JB a 6 year old pupil who presented to clinic with 4 years history of recurrent sore throat. There was no dryness of the tongue, peri-areolar swelling, epistaxis nor snoring. On examination there was fullness in the right lateral wall of the oropharynx pushing towards the midline, with soft palatal bulging at the same site displacing the uvula to the left. The overlying mucosa appeared normal with no hyperemia or exudate. Other investigations were within normal limits. Intraoperatively there was a huge firm parapharyngeal mass (mainly muscular) involving the right side of the soft palate, as well as the superior, middle and part of the inferior constrictor muscle. Assessment of a parapharyngeal tumour was made with differentials of (soft palatal tumour or tonsillar tumour). The excised tissue was submitted for histology. Histological diagnosis of cavernous lymphangioma was made.*

Conclusion: *Lymphangioma of the parapharyngeal space is a rare entity. It is difficult to diagnose purely on clinical examination, needing extra attention as it is deeply located in the neck spaces which contain neurovascular structure. Preoperative imaging plays a crucial role in identification of the lesion because it can help avoid complications during surgical exploration.*

Keywords: Parapharyngeal, lymphangioma, benign, cystic hygroma

INTRODUCTION

Lymphatic malformations are congenital malformations of the lymphatic system. Cystic hygroma also known as cystic lymphangioma was first described in European literature by Redenbacher in 1828.[1]They consist of channels and cystic spaces of varying size and result in the accumulation of fluid, often beneath the skin.

Embryologically, they are thought to originate from the sequestration of lymphatic tissue during the development of lymphatico-venous sacs, which then fail to communicate with the remainder of the lymphatic or venous system. Later on the sequestered lymphatic tissues dilate, which results in the cystic morphology of the lesions.[1, 2]

Two-thirds of all the reported cases are found in the cervico-facial regions and axilla. Other less common sites are mediastinum, groin and below the tongue. Occasionally, these malformations occur in the liver, spleen, kidney and intestine.

Omental cyst in omentum and mesenteric cyst

in the mesentery of intestine represents parallel lesions at these locations.[3,4] Lymphatic malformations can be congenital or acquired. Acquired lesions generally arise from obstructions of the lymphatic system.[5] Trauma or infection is generally considered responsible for their pathogenesis.[6]

Indications for treatment include recurrent infection, cosmetic disfigurement and compression of local structures such as the airway, blood vessels, or upper gastrointestinal tract.[5]

CASE REPORT

JB is a 6 year old pupil who presented to our clinic with a 4 year history of recurrent sore throat, about 4 episodes per year, usually associated with odynophagia and dysphagia (to solids) and high grade intermittent fever, with no chills nor rigors. There was also an associated history of a non-pulsating swelling in the right aspect of the throat, but no history of contact bleeding, dryness of the tongue, peri-areolar swelling, epistaxis, snoring nor excessive sneezing. On general examination JB was not in obvious respiratory distress, pale nor cyanosed. On examination of the oral cavity/oropharynx, the lips appeared normal with fair orodental hygiene. There was fullness in the right lateral wall of the oropharynx pushing towards the midline with soft palatal bulging at the same site displacing the uvula to the left, while the overlying mucosa appeared normal with no hyperemia nor exudate, as well as being firm, non-tender with no contact bleeding .

Assessment of parapharyngeal tumour was made with differentials of soft palatal or tonsillar tumour. Full blood count and serum electrolytes and urea were within normal ranges.

Operative findings were those of an enlarged right level Ib lymph node, an enlarged right submandibular gland, and a huge right parapharyngeal mass (mainly muscular and firm) involving the right side of the soft palate, the superior, middle and part of the inferior constrictor muscle. Through both external and internal approaches, a right sided per-styloid neck dissection with right submandibular gland and right parapharyngeal mass excision biopsy was done.

DISCUSSION

Nearly half of all lymphatic malformations are diagnosed in the head and neck regions, and frequently pose treatment dilemmas for the surgeon. This is largely due to the variety of anatomic structures within or bordering the parapharyngeal space that may account for many types of lesions that involve this area.[7,8] Lymphatic malformations range in size from small, asymptomatic masses to massive, disfiguring lesions.[2] The index case presented without any obvious neck swelling, which was why some investigations like ultrasound scan of the neck was not requested nor fine needle aspiration cytology carried out to aid diagnosis before any surgical procedure was done on the patient. Three types of lymphangioma have been described: the superficial multicystic type; the deep cavernous type; and the cystic hygroma.

The index case was of the deep cavernous type as evidenced by the histology report. However, lymphangiomas actually represent a single type of defect in lymphatic development manifesting with different degrees of severity. Generally speaking, lymphangiomas are less common than hemangiomas.[9] Most of those occurring in the head and neck area (50-65%) are present at birth, while 90% are clinically apparent at 3 years of age; the majority of these are the deep cavernous type.[10] The index case had been

having symptoms pointing towards the lesion from earlier in life, likely from around two years of age, occurring in the head and neck area and on the operating table it was found to be deep seated and histologically of the cavernous type. The most common presentation is that of a painless soft mass that gradually enlarges and then remains static over a long period. Although occasional enlargement and then shrinkage occurs, a residual mass remains.[11] The index case presented with a 4 year history of swelling in the right aspect of the throat which was painless, with occasional increases in size but never shrinking since its onset. The usual presentation of a cystic hygroma apparent at birth is a painless mass with worries and queries from the parents about the lesion. The other modes of presentations are related to the complications or effects of cystic hygroma, such as respiratory distress with difficulty in feeding as in the index case i.e. odynophagia and dysphagia to solids. The patient also presented with high grade intermittent fever, no chills nor rigors, and a sudden increase in the size as well as infection in the lesion.[12]

Ultrasound scan usually shows a multicystic lesion with internal septations, while no blood flow is detected on Color-Flow Doppler. The latter is particularly helpful in detecting the different vascular characteristics of mixed vascular malformations and it also differentiates hemangiomas from vascular malformations.[13] The index case did not benefit from ultrasound scan because of the unusual presentation of the condition and the clinical finding were highly suggestive of a parapharyngeal tumour with differential of soft palatal or tonsillar tumour.

Computed tomography scan (CT scan) and magnetic resonance imaging (MRI) delineate extension and show relation with other anatomical structures better than ultrasound scan. CT scan shows multicystic, homogeneous, non-invasive lesions with low attenuation. MRI is particularly useful for malformations that involve muscle and whilst T1-weighted images are helpful in locating lesions, a T2-weighted scan can readily distinguish them from normal muscle. Cross-sectional imaging is also helpful in measuring how much of the lesion is close to the great neck vessels, particularly when surgery is being planned.[14] These benefits from CT scan and magnetic resonance imaging (MRI) were temporarily not available at the time of patient's presentation and patient could not afford these services in other centers which were far away from this centre.

Histopathology revealed thin connective-tissue stroma separating cystically dilated spaces lined by a single layer of benign endothelial cells which was consistent with cystic hygroma (Fig I and II).

Current treatment methods for lymphatic malformations include surgery, sclerotherapy and laser therapy, or a combination thereof. Non-surgical conservative treatments include radiotherapy, electro coagulation, cryotherapy, ligation and embolization.[14] This patient benefited from surgery only because this was the only available option in this centre.

Although lymphatic malformations are benign lesions, spontaneous regression is rarely seen and in only 12.5-15.0%. Therefore, observational monitoring may be appropriate in some cases. However, frequently a regression is usually followed by recurrence in most cases. Surgery remains the treatment of choice in well-localized lesions with the least recurrence rate when complete excision is possible.[12,15]This patient was discharged home and is doing well and is regular on follow up as the chances of recurrence was explained to the patient's parent especially in an incompletely excised lesion like his own.

CONCLUSION

Lymphangioma of the parapharyngeal space is a rare entity. It is difficult to diagnose purely on clinical examination, and needs extra attention as it is deeply located in the neck spaces which contain neurovascular structures. Preoperative imaging plays a crucial role in the identification of the lesion because it can help to avoid complications during surgical exploration.

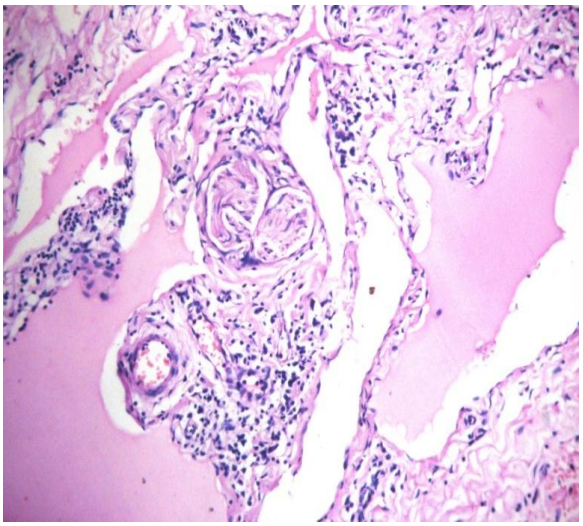

FIG I. cystic spaces with neurovascular bundles and regular lymphocytes. (H & E x100)

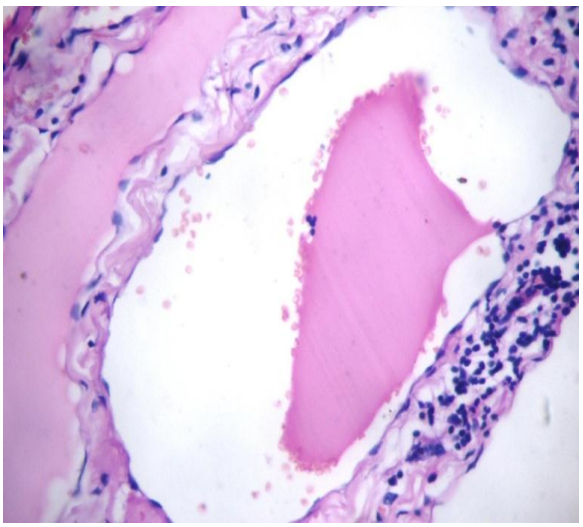

FIG II. Cystic space containing eosinophilc materials including some lymphocytes (H & E x200)

REFERENCES

1. Mehta M. Cystic hygroma- 2 cases. Indian Journal Otolaryngology and Head and Neck Surgery, 2000, 52:3:319-22
2. Ramashankar CP, Nishit KS, Grishi G. Lymphatic malformations: A dilemma in diagnosis and management. Contemp Clin Dent. 2014; 5(1): 119–122.
3. Kocher HM, Vijaykumar T, Koti RS, Bapat RD. Lymphangioma of the chest wall. J Postgrad Med 1995;41:89-90.
4. Priyadershini AR, Mahindra K, Farhana G, Shaliputra PM. Parapharyngeal lymphangioma -a case report Journal of Dental and Medical Sciences. 2015;14 (8 Ver. IV): 21-25
5. Brock ME, Smith RJ, Parey SE, Mobley DL. Lymphangioma. An otolaryngologic perspective. Int J Pediatr Otorhinolaryngol. 1987;14:133–40.
6. Aneeshkumar MK, Kale S, Kabbani M, David VC. Cystic lymphangioma in adults: Can trauma be the trigger? Eur Arch Otorhinolaryngol. 2005;262:335–7.
7. Maran AG, Mackenzie IJ, Murray JA. The parapharyngeal space. J Laryngol Otol. 1984;98:371–80.
8. Batsakis JG, Sneige N. Parapharyngeal and retropharyngeal space diseases. Ann Otol Rhinol Laryngol. 1989;98:320–1.
9. Marx RE, Stern D. Oral and Maxillofacial Pathology: A Rationale for Diagnosis and Treatment. 2nd ed. Chicago: Quintessence Publishing Co; 2003. p. 435.
10. Neville BW, Damn DD, Allen CM, Bouquot JE. Oral and Maxillofacial Pathology. 2nd ed. Philadelphia: WB Saunders; 2002. p. 476.
11. Bauman NM, Giguere CM, Manaligod JM, Sato Y, Burke DK, Smith RJ. Management of lymphatic malformations-If, when and how. Oper Tech Otolaryngol Head Neck Surg. 2002;13:85–92.
12. Perkins JA, Maniglia C, Magit A, Sidhu M, Manning SC, Chen EY. Clinical and radiographic findings in children with spontaneous lymphatic malformation regression. Otolaryngol Head Neck Surg. 2008;138:772–7.

13. Fonkalsrud EW. Lymphatic disorders. In: Grosfeld JL, O'Neill JA Jr, Coran JA, Fonkalsrud EW, Coran AG, editors. Pediatric Surgery. 6th ed. Chicago: Mosby Elsevier; 2006. pp. 2137–45

14. Zhou Q, Zheng JW, Mai HM, Luo QF, Fan XD, Su LX, et al. Treatment guidelines of lymphatic malformations of the head and neck. Oral Oncol. 2011;47:1105–9.

15. Boardman SJ, Cochrane LA, Roebuck D, Elliott MJ, Hartley BE. Multimodality treatment of pediatric lymphatic malformations of the head and neck using surgery and sclerotherapy. Arch Otolaryngol Head Neck Surg. 2010;136:270–6.