The Journal of Laryngology & Otology, E2, 1 of 3.

doi:10.1017/S0022215107005695
Printed in the United Kingdom

Online Only Clinical Record

Lymphangioma of the larynx

B Vaishali, A Nambiar*, R Indudharan

Abstract
Lymphangiomas are uncommon congenital lesions of the lymphatic system. Most of these lesions present in infancy or early childhood with a swelling in the head and neck region. We report a case of adult lymphangioma of the larynx. We also discuss the presentation, diagnosis and management of this tumour, and we present the current debate on sclerotherapy versus surgery for these tumours.

Key words: Lymphangioma; Larynx

Introduction

Cervical lymphangioma is a rare vascular malformation usually reported in children, rarely in adults. Usually, the clinical features plus computed tomography or magnetic resonance imaging (MRI) findings can suggest the diagnosis. Definitive diagnosis is based on post-operative histology. Despite the benign nature of these lesions, surgical management is difficult, especially for cavernous lymphangioma, because of its tendency to spread along vital structures and the subsequent high incidence of recurrence.[1] Long-term follow up is required.

We present a case of lymphangioma of the larynx in an adult, which posed a diagnostic dilemma and failed to respond to sclerotherapy.

Case report

A 26-year-old woman presented with a history of swelling on the right side of the neck since birth, for which she had undergone surgery at the age of 10 years. Details of this procedure were unavailable. The swelling had reappeared at the age of 16 years, but the patient had been asymptomatic until the age of 23, when the swelling had started to increase in size and the patient had begun to experience dyspnoea on exertion.

The patient was admitted to her local hospital, where she underwent direct laryngoscopy. A huge, vascular mass was seen occupying the larynx. In view of the increased vascularity and strategic position of the mass, the lesion was considered inoperable and the patient was referred to a higher centre for sclerotherapy.

After thorough evaluation, the patient underwent preliminary tracheostomy and thereafter underwent 10 sclerosant injection sessions performed by a thoracic surgeon. There was only a slight regression of the lesion following sclerotherapy. As surgery was thought to be life-threatening, the patient was advised to undergo repeated sclerotherapy whenever the swelling enlarged.

The patient presented to us six months after sclerotherapy, with a swelling in the neck and persistent dyspnoea on climbing a few steps.

On examination, there was swelling on the right side of the neck, which increased on deep expiration and phonation. Flexible laryngoscopy revealed a pinkish, irregular mass involving the right arytenoid and aryepiglottic fold, with sluggish movement of the right vocal fold. The left vocal fold was normal and mobile.

The patient underwent a T2-weighted, fat-suppressed MRI. This showed a well defined, vascular mass involving the right supra-glottic region, extending both medial and lateral to the thyroid lamina and also encroaching into the parapharyngeal space (Figure 1a). Magnetic resonance angiography did not reveal any feeder vessel.

Based on these findings and investigations, a clinical diagnosis was made of cavernous haemangioma of the larynx with extralaryngeal spread, and surgery was planned.

A preliminary tracheostomy was performed and the tumour was explored via a transcervical approach. Extensive fibrosis was encountered, probably due to the previous sclerotherapy. On dissection, there was a spongy, vascular mass lying over the lamina of the thyroid cartilage, extending over and above its upper border, invading the thyrohyoid membrane and encroaching into the paralaryngeal spaces. The mass was dissected out and a rent in the pyriform fossa was closed primarily. Intra-operative direct laryngoscopy confirmed complete removal of the intralaryngeal mass.

The post-operative period was uneventful. The patient's tracheostomy was closed on the 10th post-operative day and the patient was discharged from hospital.

Histopathology

The sections of the dissected specimen showed large, dilated, irregular-shaped, vascular channels lined by bland, flattened, widely spaced endothelial cells embedded in a fibroblastic, collagenous stroma. Some of the channels showed an incomplete layer of smooth muscle. Small collections of lymphocytes were seen scattered focally in the stroma and within the vascular spaces. A few of the lumina showed flocculent, eosinophilic precipitates of proteinaceous lymphatic fluid and haemorrhage. The lesion

From the Departments of ENT and *Pathology, Amrita Institute of Medical Sciences and Research Center, Kochi, Kerala, India.

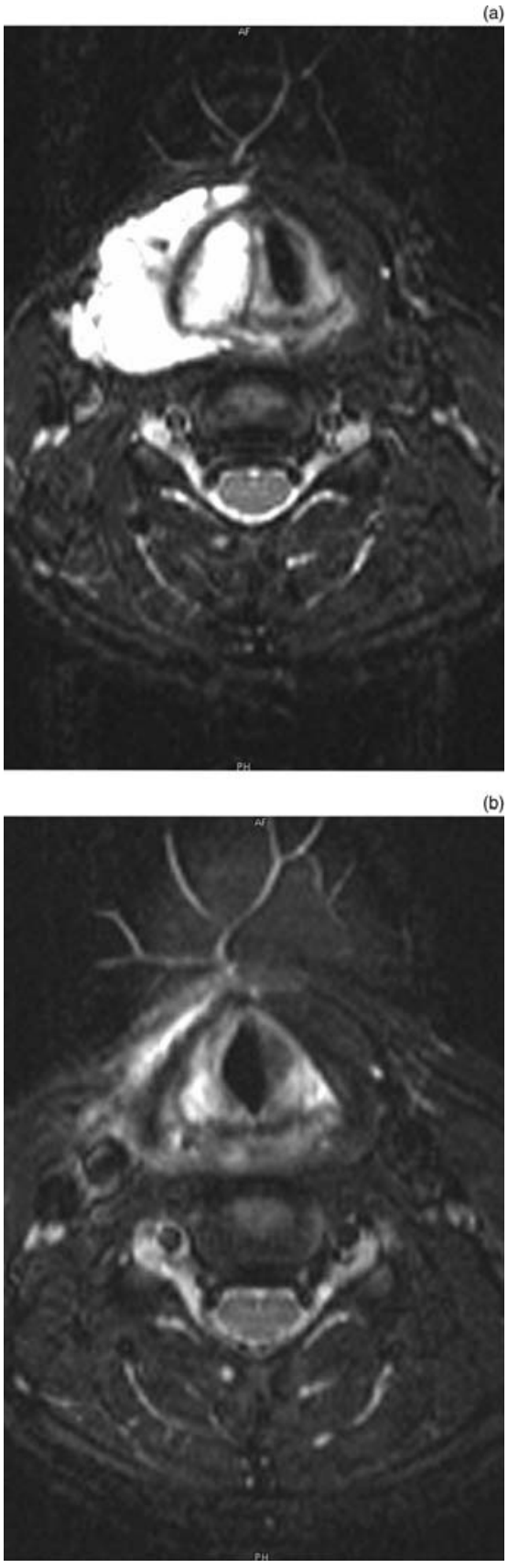

FIG. 1
(a) Pre- and (b) post-operative magnetic resonance imaging appearances of cavernous lymphangioma of the larynx. Note the vascular lesion involving both sides of the lamina of the thyroid cartilage.

was seen to infiltrate into the skeletal muscle and extended up to the surgical margins of excision. A diagnosis of cavernous lymphangioma was made (Figure 2).

Periodically repeated laryngoscopies confirmed the complete removal of the mass. The mobility of the right vocal fold, which had been restricted following surgery, gradually returned. A repeat MRI showed no evidence of any residual mass (Figure 1b). Twenty-four months after surgery, the patient remained asymptomatic.

Discussion

Lymphangioma is a rare, congenital anomaly of the lymphatic system, which usually presents in childhood. The majority occur in the head and neck, particularly in the posterior triangle. There are very few reports in the literature documenting the presence of lymphangioma in the larynx.

The aetiology and pathophysiology of this lesion are still in question. It is thought to arise from sequestration of portions of the primitive embryonic anlage, or as areas of localised lymphatic stasis caused by congenital blockage of regional lymphatic drainage. These lymphatic lesions may be divided into three morphological types – capillary, cavernous and cystic – of which cystic is more common in the neck. Capillary lymphangiomas are composed of capillary-like lymphatic vasculature, cavernous lymphangiomas are composed of dilated lymphatic channels with or without an adventitial layer, and cystic lymphangiomas contain multilocular cysts.[1]

Our patient was diagnosed as having a cavernous lymphangioma. Histologically, cavernous lymphangioma and cystic hygroma are very similar, being composed of dilated, very thin-walled, vascular spaces lined by flattened endothelium and filled with eosinophilic, proteinaceous material. Thin strands of fibroconnective tissue separate these spaces. Scattered lymphoid aggregates are also found, occasionally in the form of germinal follicles, and wisps of smooth muscle fibre may also be present. Sometimes, erythrocytes may also be present focally within these spaces, usually as the result of surgical trauma.[2] A pre-operative diagnosis of lymphangioma was difficult in our case due to the clinical appearance and radiological findings.

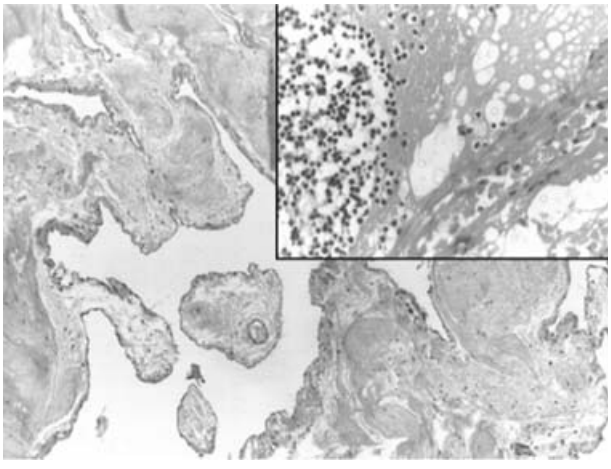

FIG. 2
Histopathological appearance of cavernous lymphangioma of the larynx, showing irregular, dilated channels with widely spaced endothelial lining (H&E; ×100). Inset shows lumina containing lymphocytes and proteinaceous lymph (H&E; ×400).

https://doi.org/10.1017/S0022215107005695 Published online by Cambridge University Press

On review of the literature, we found little difference in the radiological and histopathological features of haemangioma and lymphangioma. A traumatised lymphangioma may resemble a cavernous haemangioma, and, conversely, a haemangioma devoid of erythrocytes may resemble a lymphangioma. Furthermore, the two processes can coexist.

The irregular vascular lumina, lymphoid aggregates and inapparent, widely spaced endothelial cells help distinguish lymphangioma with intraluminal haemorrhage from true haemangioma. Lymphatic endothelial cells are much less reactive for vascular endothelial markers such as factor VIII-related antigen, CD31 and Ulex europaeus lectin than their blood vascular counterparts.[3]

There are no independent therapeutic or prognostic implications in distinguishing between lymphangiomas and haemangiomas. Clinically, the extent of the lesion is the most significant factor affecting prognosis.[2]

Various treatment modalities have been suggested in the literature, using sclerosants such as OK-432 (OK-432 is a lyophilised mixture of a low virulence strain (Su) of group A Streptococcus pyogenes incubated with benzylpenicillin. It is a potent immunostimulant. It is also a sclerosant used universally for treatment of angiomatous malformations.) (Picibanil, Chugai Pharmaceuticals Co. Ltd, Tokyo, Japan)[4], bleomycin,[5] ethanol and acetic acid. The use of radio frequency ablation has also been described, with good results.[6] Where facilities are available, magnetic resonance guided neodymium: yttrium aluminium garnet laser induced interstitial thermotherapy has been proven as a safe mode of treatment. This can be safely used for deep tumours, with preservation of vital structures. Real time monitoring of tissue temperature with thermo-sensitive sequences allows controlled coagulation necrosis.[7] However, surgery remains the therapeutic option of choice. Long-term follow up is mandatory in order to promptly treat any recurrence.

Sclerotherapy obviates the need for invasive primary surgery and is safe, with a high cure rate especially with agents such as OK-432. However, sclerotherapy may have local or systemic complications and requires prolonged treatment and regular follow up; the cost is high and suitable agents must be available. Surgery may be the better option, depending on the site of the lesion. This was the situation in our case, in which the lesion was obstructing the airway, requiring a long-term tracheostomy in a young woman. The potential for recurrence exists no matter what treatment modality is chosen. However, if complete surgical clearance can be achieved, the risk of recurrence will be greatly reduced, or removed. In addition, surgery is a single stage procedure, compared with the multiple sessions of sclerotherapy required. The hazards of surgical exploration include damage to vital structures and uncontrolled haemorrhage if feeder vessels are not identified and controlled pre-operatively.

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Address for correspondence:
Dr R Indudharan,
Professor,
ENT Dept,
Amrita Institute of Medical Sciences,
Kochi, Kerala,
India 682026.

Fax: 95 484 2802020
E-mail: drindudharan@aimshospital.org

Dr R Indudharan takes responsibility for the integrity of the content of the paper.
Competing interests: None declared