Isolated Laryngeal Lymphangioma: A Rarity

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Abstract Lymphangiomas are uncommon congenital lesions of the lymphatic system, most often located in the head and neck. Most of these lesions manifest in children less than 2 years of age. Involvement of the larynx in isolation is rare and only a few cases have been reported so far. We report the case of a thirteen-year-old girl who presented with a swelling in the supraglottis. Excision of the mass was done after tracheotomy. Histopathological diagnosis was lymphangioma of the larynx. After a follow up of 15 months the patient is free of recurrence with all functions of larynx being normal.

Keywords Lymphangioma · Larynx

Introduction

The lymphatic system forms during the sixth week of embryonic development from paired jugular sacs that begin as clefts in the cervical mesenchyme. Lymphangiomas are congenital malformations of the lymphatic system. They commonly present in the head and neck region, with over 90% of lesions manifesting before 2 years of age. The onset of the lesion after 2 years of age is rare. Although the head and neck region is the most common site of presentation, isolated laryngeal lymphangioma is extremely rare as reported by Seven et al. [1]. Laryngeal sites are usually in continuity with foci of disease in the neck. Reported incidence of involvement of larynx in a children's series of head and neck lymphangiomas reviewed by Cohen and

Thompson [2] was 14–22%. We report the case of a thirteen-year-old girl, who was diagnosed with an isolated lymphangioma in the supraglottis. She was managed successfully by an endoscopic approach and is free of recurrence in the follow up period of 15 months.

Case Report

A thirteen-year-old girl presented to the outpatient department with complaints of foreign body sensation in the throat, progressive change in voice and difficulty in swallowing for 3 months. She also complained of difficulty in breathing on lying down at night. On examination of the girl, the voice was muffled and there was no stridor. The neck, nose and oropharyngeal examination was normal. On indirect laryngoscopy, there was a smooth globular mass in the right side of larynx, just below the tip of epiglottis, obscuring view of the cords partially. The site of origin of the mass could not be ascertained.

A MRI of the neck was obtained. It revealed a large, well-demarcated, broad based polypoidal soft tissue mass along the right aryepiglottic fold, abutting on the posterior surface of epiglottis. It measured $27 \times 22 \times 28$ mm in size and was causing effacement of the pyriform sinus, as seen in Figs. 1 and 2.

The impression of a benign lesion of the larynx was made and we took up the patient for excision of the mass. Tracheostomy was done because of anticipated difficulty in intubation. On direct laryngoscopy we found a globular cystic mass arising from the aryepiglottic fold, and false cord on the right side, abutting on the laryngeal surface of the epiglottis as seen in Fig. 3. The mass was slit open using a sickle knife. It was cystic with multiple septae and was filled with yellowish fluid. The mass was deroofed and

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Fig. 1 MRI sagittal section showing soft tissue mass in supraglottic area

Fig. 2 Axial section showing mass occupying the airway

Fig. 3 Direct laryngoscopic picture showing globular mass close to the epiglottis, obscuring view of endolarynx

excision was done, assisted by radiofrequency coagulation. All laryngeal structures were preserved and haemostasis was ensured.

The histopathology of various sections showed unremarkable stratified squamous epithelium lined mucosa with submucosa containing numerous glands. Multiple cystically dilated channels lined by flat attenuated endothelium were seen in the underlying tissue. The channels were containing lymph and lymphocytes. Few lymphoid aggregates were also seen between the vessels as seen in Fig. 4.

The patient was decannulated after 24 h in the post-operative period. The recovery was uneventful. The patient has been asymptomatic and all laryngeal functions are normal after a follow up period of 15 months.

Discussion

Lymphangiomas are uncommon congenital benign malformations of the lymphatic system. Cohen and Thompson [2] in their extensive review of pediatric lymphangioma in 160 patients, found involvement of the larynx in ten patients. Lymphangiomas involving the larynx are either extension of the cervicofacial lesions or they extend to involve the neck and other areas. Orvidas and Kasperbauer [3] in their review of pediatric lymphangiomas of head and neck, found the submandibular area to be most commonly affected (37%) and the parotid gland as the second most common site (31%). Isolated laryngeal lymphangioma is a rare form of the lesion [2, 4], with only a few cases reported in the literature. Lymphatic malformations confined to the larynx occur more often in Caucasians than African Americans [2]. Kenton et al. [5] reported the case of a neonate with lymphangioma of the larynx, who presented with stridor and progressive respiratory failure. They gave a review of seven cases of laryngeal

Fig. 4 Histopathological picture showing lymphoid aggregates

lymphangioma reported so far in the literature. Only two of these seven cases presented in young adults, the remainder were all reported in less than 2 years of age.

Lymphangiomas present with symptoms depending upon the site of involvement and the size of the mass. Cystic hygromas of the neck may at times be so extensive so as to lead to symptoms of airway obstruction. Lymphangioma localized to the larynx present with symptoms similar to other benign tumours of the larynx such as change in voice, airway obstruction and stridor if it is extensive disease. Claros et al. [6] reported the case of a 16 year old girl with Down's syndrome. She presented with progressive dyspnea and had to undergo supraglottic partial laryngectomy for a lymphangioma involving the epiglottis. A case of a 37-year-old lady is reported [1], who presented with symptoms suggestive of acute epiglottitis. After resolution of her acute symptoms, she was found to have a mass arising from the epiglottis, excision biopsy of which revealed it to be a lymphangioma. It was emphasized that if an infection occurs in a case of a lymphatic malformation, symptoms can rapidly worsen and lead to complications like airway obstruction. A case has been reported in a thirteen-year-old girl with lymphangioma restricted to the larynx, who presented with acute airway obstruction in the absence of any other symptomatology [7].

Lymphangiomas are diagnosed on the basis of history, examination findings and radiological investigations. Magnetic resonance imaging can demarcate the mass from the surrounding tissues and can give the extent of the lesion. Contrast enhanced CT scan and ultrasound of the neck can also be helpful in investigating these lesions.

Various treatment options of lymphangiomas, namely surgical excision; laser debulking, sclerotherapy and corticosteroids have been outlined [3]. It is suggested that the desire for complete excision must be weighed against the need to keep vital structures intact. 75% patients in a series of sixty seven children with lymphangiomas of the head and neck [3] underwent surgical excision. It was found that lesions involving the lip, hypopharynx and/or larynx, tongue and floor of mouth had high rates of recurrent or persistent disease. If the larynx is involved tracheotomy is almost always required [2]. The surgical laser is considered particularly suitable for a lesion in the larynx because it allows precise ablation and recontouring of the tissue so

that the functions can be preserved [4]. Sclerosing agents are under investigation for laryngeal lymphangiomas but since they act by causing inflammatory reaction, their use can result in damage to adjacent structures. Radiotherapy is not effective in treatment of these lesions [7]. As there have been no controlled trials which have compared the various treatment modalities, the management should be decided on the basis of the extent of the lesion and the site of the lesion. Even with complete excision the recurrence rate of these lesions is high. The predicted prognosis is poor if there is extensive infiltration of upper airway and oesophageal structures as seen by Hartl et al. [8].

In our case we were able to excise the lesion without causing any damage to surrounding vital anatomical structures of the larynx, using an endoscopic approach.

We reported this case because of the unusual site of involvement; in isolation from rest of the head and neck and unusual age of presentation. Although rare in occurrence, the otolaryngologists should keep in mind the diagnosis of lymphangioma in the larynx amongst other benign lesions.

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