CASE STUDY

Extensive Laryngopharyngeal Cavernous Lymphangioma Causing Upper Airway Obstruction

Linfangioma cavernoso laringofaríngeo extenso causante de obstrucción de las vías aéreas superiores

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Introduction

Lymphangiomas represent malformations of lymphatic system that frequently affect head and neck region. They are usually seen at birth or in early childhood. 1

According to the size of the lymphatic cavities lymphangiomas are classified as microcystic (capillary), macrocystic (cavernous) and cystic hygromas. 2 According to progression of disease staging system was proposed: stage I–unilateral infrathyroid disease, stage II–unilateral suprathyroid, stage III–unilateral supra- and infrathyroid disease, stage IV–bilateral suprathyroid, and stage V–bilateral suprathyroid and infrathyroid disease. 3

For laryngeal and hypopharyngeal site the radiographic differential diagnosis includes: retention cyst, ectopic thyroid, dermoid cyst, thyroglossal duct cyst or hemangioma. The histologic differential diagnosis includes hemangioma, and metastatic papillary cystic thyroid carcinoma. 4,5

Extensive cavernous lymphangioma occurs in 3%–18%. Advanced lymphangiomma can cause airway or feeding difficulties, especially in newborns and young infants. Their outcome is sometimes poor. 6

The aim of this work was to present surgical technique and the outcome for far advanced laryngo-pharyngeal lymphangioma that was previously unsuccessfully treated.

Case Report

Male patient, 42 years old, weighing 142 kilograms was urgently admitted because of advanced stridor, cyanosis, tachypnea and tachycardia. Medical documentation revealed fourteen previous operations in three medical centers during the past 20 years. Some of these operations were endoscopic using CO2 laser, and some by external approach, and the last operation was done four years ago. Patohistologic diagnosis was cavernous lymphangioma.

ENT examination confirmed multiple cervical scars, and extensive tumor formation starting from the tongue base, extending downwards and nearly completely obstructing airways. He was immediately transferred to operating room and urgent tracheostomy was performed. Trachea was significantly dislocated. Immediately after tracheal opening oxygen saturation improved, but it was still 88%, because of poor ribcage expansion and intraabdominal pressure on diaphragm. In order to achieve proper cannula position and

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care, reduction of all subcutaneous pretracheal tissue was made, and trachea was sutured to the skin.

The next day computed tomography was undertaken. It confirmed extensive tumor starting from tongue base, to right hypopharyngeal and laryngeal structures, ending at the level of ventricular folds, and producing subtotal airway obstruction (Fig. 1.).

In order to improve his general and pulmonary condition the patient was placed on pulmonary therapy (parenteral antibiotics and bronchodilatators) and diet with restriction of carbohydrates and fat. This improved his Body Mass Index from 43.3 kg/m² to 37.5 kg/m².

Definitive surgery was performed after one month. Collar neck incision was made; supraglottic laryngectomy, partial right pharyngectomy, and partial tongue base resection were necessary to remove the tumor entirely (Fig. 2.). Direct pharyngeal suture was possible. Postoperative course was uneventful. Pathohistology confirmed cavernous lymphangioma (Fig. 3.).

The patient was decannulated after two weeks. After three months control CT showed adequate airway and absence of tumor (Fig. 4.). Current follow-up exceeds one year, and confirms his excellent medical condition.

Discussion

Different nonsurgical modalities were proposed for treatment of lymphangiomas, such as: radiation, diathermy, and injection of interferon alfa, bleomycin sulfate, triamcinolone acetonide, alcohol solutions and sclerosant OK-432. The success of OK-432 was emphasized in few studies, especially in macrocystic form of disease.¹,² Also, patients with stages I, II and III respond better to treatment.³

Spontaneous partial resolution of lymphangiomas is typical; however surgery is often needed. The goals of surgical therapy are: complete removal of the tumor, preservation of function, reduction of complications and prevention of disfigurement.⁴

Adequate surgical therapy of head and neck lymphangiomas is difficult because of cystic character, thin lining
and characteristic invaginations. Cavernous and microcystic lesions demand a few surgical interventions. Tracheotomy must be performed in one half of the patients, and excision of advanced disease neck dissection, and tongue or pharynx resections should be made. 8–10

However, surgery for extensive tumors is not always successful. Prolonged tracheotomy, laser therapy and partial resection were proposed. It was reported that early surgery can reduce the duration of tracheotomy and improve results. On the other side, neck dissection can block both lymphatic and venous drainage, and increase airway obstruction. So, risks and benefits of lymphangioma surgery must be carefully evaluated. 11–13

Age, symptoms and extent of the disease are important prognostic factors for lymphangiomas. Recurrence rate after surgery of extensive lymphangiomas varies, with values up to 80%. 1,4

We report adult patient who was unsuccessfully treated many times using different approaches. The extension of tumor, and his extreme obesity led to subtotal airway obstruction.

Urgent tracheotomy, with removal of soft tissue mass, enabled sufficient respiration. Adequate medical preparation and extensive surgery made tumor elimination possible. Feeding and respiration were normalized. Sclerotherapy was not feasible in this case because of many previous operations and scars.

This case confirms that extensive surgery, even in far advanced and complicated cases of cavernous lymphangioma can give good results. However, long-term follow-up is needed since many previous operations were performed, and the disease was extensive.

Conflict of Interest
The authors declare no conflict of interest.

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