Lymphangiomas of the head and neck in children

D.L. Grasso, G. Pelizzo, E. Zocconi, J. Schleef

ENT Department, Pediatric Surgery Department, IRCCS “Burlo Garofolo” Children’s Hospital, Trieste, Italy

Summary

Lymphangiomas are rare benign congenital tumours, involving both the head and the neck and causing obstructing symptoms in the upper airways as well as aesthetic anomalies. In recent years, sclerosing therapy with OK-432 has become the treatment of choice in the management of these lymphatic malformations. Nonetheless, surgery still seems to be the therapy advocated for resolution of symptoms. Herein, three cases of lymphangiomas involving the head and the neck are described and a review of the English scientific literature is outlined.

Key Words: Head and neck • Lymphangioma • Children • Diagnosis • Treatment

Introduction

Lymphangiomas are rare congenital benign lesions occurring mainly in the head, neck and oral cavity. They consist in localized centres of abnormal development of the lymphatic system. Three theories have been proposed to explain the origin of this abnormality. The first suggests that a blockage or arrest of normal growth of the primitive lymph channels occurs during embryogenesis, the second that the primitive lymphatic sac does not reach the venous system, while the third advances the hypothesis that, during embryogenesis, lymphatic tissue lies in the wrong area. Lymphangiomas are classified as microcystic (capillary lymphangiomas), macrocystic (cavernous lymphangiomas) and cystic hygromas according to the size of the lymphatic cavities incorporated. Bill and Summer, in 1965, presented the concept that cystic hygromas and lymphangiomas are variations of a single entity and that its classification is determined by its location in the head and neck. A commonly used classification classifies these lesions into capillary lymphangioma or lymphangioma simplex, cavernous lymphangioma, and cystic lymphangioma or cystic hygroma. When a lymphangioma is confined to fairly dense tissue, such as the tongue, it presents as a cavernous lymphangioma, but when it develops in the relatively loose fascia of the neck, a cystic lesion occurs. These three types are frequently found together in a same patient, depending on the severity of the disease. Cystic hygromas, however, account for approximately 90% of the lymphangiomas in the head and neck region. Other common sites, outside the head and neck, include the axilla, shoulder, chest wall, mediastinum, abdominal wall, and thigh. Surgical resection still remains the best treatment for lymphangiomas; other treatment options, such as sclerotherapy have been proposed as an alternative to reduce the impact and complications of surgery. Various products, such as sodium morrhuate, dextrose, tetracycline, doxycycline, bleomycin, ethibloc and OK-432, have been used as sclerotherapeutic agents. Apart from OK-432, the other agents were reported to cause perilesional fibrosis and thus to complicate eventual surgical excision. The present report focuses on 3 cases of lymphangiomas of the head and neck in children, the therapeutic options adopted and a review of the English scientific literature.

Case report 1

Prenatal diagnosis of neck mass

At birth, presence of cystic lymphangioma involving both sides of the neck and extending to the floor of the mouth. Macroglossia. Sclerosing therapy was started from the first month of life with Ethibloc® (Ethicon GmbH, Hamburg-Norderstedt, Germany) medium injection amount of 2 ml (alcoholic solution of zein), but results were not successful. Sclerosing therapy with OK-432 was performed. Concentra-
tion of OK-432 was 0.01 mg/dl. After fluid aspiration from the lesion, the same amount of OK-432 was introduced, up to a maximum of 20 ml. Injections were performed, with ultrasonography (US) guidance, under general anaesthesia and following informed consent from the parents.

At 18 months old, during sclerosing therapy with OK-432, respiratory distress appeared. Tracheotomy was performed. After two months, submandibular and bilateral functional neck dissection was performed. Tracheotomy was removed after one year. Today, macroglossia and prognathism are present. No recurrence of disease in the previous area has been found. Lymphangiomatosis, at the tongue base, is still present.

Case report 2

Prenatal diagnosis of left submandibular mass

Tongue protrusion and mild face asymmetry were present. Extrinsic tracheal compression was revealed by computed tomography (CT), with no dyspnoea. Sclerosing therapy was started from the first month of life with OK-432. Concentration of OK-432 is 0.01 mg/dl. After fluid aspiration from the lesion, the same amount of OK-432 was introduced, up to a maximum of 20 ml. Injections were performed with US guidance under general anaesthesia and with informed consent from the parents. At 6 months of life, rapid enlargement of the tongue occurred which required nasogastric feeding. Respiratory distress required tracheotomy. Submandibular neck dissection was performed. Mild facial palsy appeared after surgery. At 18 months of life, no resolution of tongue enlargement has been obtained. Surgical reduction of the tongue was performed. Improvement in general conditions was observed. Tracheotomy was removed. Today, only mild dynamic facial palsy is present. No feeding problems. Speech therapy is followed by the patient as the only treatment.

Case report 3

No prenatal diagnosis of neck mass. At birth, a mass was present in right neck region. In the following weeks, the neoplasm became enlarged. MRI revealed the presence of a lesion with inner segmentations in the right neck which dislocated the carotid artery and the right internal jugular vein. A neck dissection was performed with removal of a large multi cystic mass. Histopathology revealed a multi cystic hygroma. Three months later, relapse of disease with a single cystic lesion. Sclero-therapy was performed with OK-432. Concentration of OK-432 was 0.01 mg/dl. After aspiration of fluid from the lesion, the same amount of OK-432 was introduced (maximum 20 ml). Injections were performed, under US guidance and general anaesthesia with informed consent from parents. At present, no relapse or disease is present.

Discussion

Lymphangiomatous lesions are rare congenital malformations of the lymphatic system that occur throughout the body with greater frequency in the cervicofacial area. Cystic lymphangiomas have been reported, in the literature, to be present in up to 67% of all cystic lymphangiomas. Introduction of prenatal US allows detection of disease in utero, in approximately the same percentage. However, almost all are detected before the age of two years. Histologically, these lesions are composed of dilated lymphatic channels with one or two endothelial layers, with or without an adventitial layer. These dilated lymphatics can vary in size, depending upon the location and surrounding tissues and is the basis for classification. Cavernous lymphangiomas are found in areas, such as the tongue and floor of the mouth. Cystic hygromas, on the other hand, arise from lymphatic tissue in areas where expansion can occur and large multi-
loculated cystic spaces can develop. Depending upon cystic space size, they are classified as: macrocystic, microcystic and mixed. The incidence of lymphangiomas has been reported to range from 1.2 to 2.8 per 1000 newborns. The most prominent sign or symptom of all lymphangiomas is the presence of a mass. The mass may be small and unnoticed at birth only to present later with an upper respiratory tract disorder or incidental trauma at the site. Most lesions, however, are recognized early on account of their size and associated symptoms of respiratory obstruction and problems with feeding, which are the second and third most common presenting symptoms. Difficulty in swallowing results from lymphangiomas extending to involve the oral cavity, oropharynx, and/or the hypopharynx. Isolated tongue involvement can lead to macroGLOSSA with dysphagia and airway obstruction. Airway and swallowing problems may persist after surgery in the neck on account of mucosal oedema, enlargement of internal lymphangiomas, and loss of neural innervation to the pharynx or tongue. In adult patients, neoplasms can switch to squamous cell carcinoma. Diagnosis is not difficult in most cases. The neoplasms are usually characterized by the presence of a soft, compressible, loculated and ill-defined mass, which is usually located in the posterior cervical triangle. The lesions are not attached to the skin or movable across deeper tissues, and readily transilluminate. The anterior triangle of the neck has been indicated as the most common site. Cystic hygroma may be localized in the parotid area, and is the second most common congenital mass of the parotid area. Diagnosis is not difficult in most cases. The neoplasms are usually characterized by the presence of a soft, compressible, loculated and ill-defined mass, which is usually located in the posterior cervical triangle. The lesions are not attached to the skin or movable across deeper tissues, and readily transilluminate. The anterior triangle of the neck has been indicated as the most common site. Cystic hygroma may be localized in the parotid area, and is the second most common congenital mass of the parotid area. US, CT and MRI can be used to define the relationship of the lesion with the neighbouring structures and to help plan surgical strategies. US, which provides size and extension of the lesion, should be performed routinely. The clinical course of the pathology varies from a spontaneously regressing cyst to an aggressively invasive lesion. Spontaneous or traumatic haemorrhage of the cysts is the most common complication of the lesion.

Although various methods have been described, surgical removal of the neoplasm still appears to be the best choice in the treatment of lymphangiomas. Nevertheless, in the neck, the close relationship of the tumour to vital structures requires softer approaches in order to avoid fatal consequences. On the other hand, total removal of the mass is necessary to eliminate the risk of recurrence. To this end, many Authors have described various techniques, including serial aspiration, radiotherapy, sclerosing therapy with steroids, alcohol, bleomycin sulphate, tetracycline, and, more recently, OK-432. The main advantage of OK-432 with respect to other sclerosing agents is the absence of perilesional fibrosis, and intralesional injection of OK-432 has been proposed as the first-line treatment for lymphangioma, for the past decade. In a recent retrospective study, Oka-zaki et al. recommended OK-432 injection therapy alone, for single cystic and macrocystic types, and surgical excision after pretreatment with OK-432, for microcystic and cavernous types. In our series, not all patients showed resolution of symptoms following sclerosing therapy and, indeed, required surgical removal. Non-surgical procedures could be proposed in unresectable and recurrent tumours. Classification of lymphangioma coli has been described by Schuster et al. in order to provide predictive values of post-treatment results and occurrence of complications. They identified 4 types, depending upon volume of the lymphangioma itself, as assessed by prenatal US or clinically. Type I included tumours which had no, or only a minimal, effect on the contour of the neck; type II lymphangiomas are smaller than a line drawn at the lateral border of the head; type III tumours exceed this line; in type IV, the lymphangioma extends beyond the midline of the body. Hygromas involving multiple anatomic sites also recur more frequently than lesions confined to a single location. Some Authors have described cystic hygromas with an infrathyroid or suprahyoid location. These studies show an increased recurrence rate, morbidity, and complication rate in those lesions presenting in the suprahyoid region. While location of the lesion in the cervicofacial area was an important factor for success or failure, the age at which surgery was performed did not seem to affect the outcome. Complications are more common in lesions involving the midline. Recurrence rates vary depending upon the complexity of the lesion and the completeness of excision. Simple hygromas that are completely excised seldom recur. Complex lesions that are completely excised might recur in 10-27%, whereas partially resected lesions may recur in 50-100%. In our patients, recurrence of disease was observed, despite total surgical removal.

**Conclusions**

Lymphangioma of the head and neck are benign neoplasms which are easy to diagnose. Surgical intervention represents the treatment of choice. Lesion extension and involvement of vital structures can reduce, in some cases, the possibility of complete resection. Sclerosing therapy should be considered for recurrences.
References


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