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Surgical excision as primary treatment modality for extensive cervicofacial lymphatic malformations in children

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ABSTRACT

Objective: There has been much recent focus on sclerotherapy treatment of lymphatic malformations with OK432. Surgical treatment however can have a number of advantages, including complete curative excision. The aim of this study was to evaluate the results of surgical excision as the primary (first) treatment for this condition. This group includes a number of children with very extensive disease as well as some with smaller lesions.
Methods: Prospectively collected database with additional information from medical records of children with cervicofacial lymphangiomas treated over 10 years at a tertiary paediatric referral centre. For this study only children who underwent surgical excision as the primary treatment modality were included.
Results: Total of 118 children with lymphatic malformations were treated under the care of the senior author over a 10 year period. Of these 53 patients, who underwent surgical excision as the primary treatment modality for cervicofacial lymphatic malformations were included in this study. Forty-one patients who underwent sclerotherapy as the initial treatment were excluded. Also excluded were 6 patients who underwent thoracic surgery and 18 who were treated conservatively. The majority of the patients (41, 77.3%) underwent only a single surgical procedure (36 – surgical excision, 5 – laser excision). At the first follow up after the primary surgery, the result was complete resolution of symptoms in 29 patients, near complete resolution in 13 patients (together 79.3%) and partial response in 11 (19.7%) patients. Twenty-three patients with disease localised only to the neck, all (100%) had a complete or near complete resolution of the disease after the primary surgery. Complete/near complete response was achieved in 98% cases with macrocystic disease, regardless of the location. Minor complications occurred in 11.3% patients. No permanent nerve weaknesses occurred.
Conclusion: Cervicofacial lymphatic malformations in children should be managed in a multidisciplinary setting. Surgery remains a very important treatment modality. The majority of patients (80%) in this study had complete or near complete resolution with one surgical procedure. Isolated neck lesions have the best outcomes (100% resolution in this study). Patients with macrocystic disease, achieved complete or near complete resolution in 97% of cases, regardless of the location. Some children with extensive disease will need multiple treatments. Surgical excision as the primary treatment modality in selected cases is safe and reliable technique and has good aesthetic and functional outcomes in experienced hands.

1. Introduction

Lymphatic malformations are low flow vascular anomalies, the incidence being 1:5000 at birth [1]. These comprise 6% of all paediatric soft tissue tumours [2]. Morphologically these consist of cyst like dilated vascular channels filled with lymph [3]. Lymphatic malformations are classified according to cyst size into macrocystic (few cysts which are at least 2 cm^3 in size), microcystic (numerous small cysts which are less than 2 cm^3 in size) or mixed [4,5]. Lymphatic malformations usually present as slow growing soft swellings in the head neck region [6]. They can rapidly increase in size as a result of bleeding or infection, or may rarely decrease in size spontaneously [7]. Lymphatic malformations can be asymptomatic or may affect breathing or swallowing, depending on their size or location.

The treatment modalities for lymphatic malformations can be divided into active observation, sclerotherapy [8–10], surgical excision [11] or a combination of these. Surgical excision has the advantage of usually a single procedure to excise the disease. It offers potential complete eradication of the disease and a lifetime cure. Sclerotherapy will always leave residual malformation.

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Review of the literature on this topic revealed many articles on surgical treatment as a part of multimodality treatment or as a salvage treatment for sclerotherapy failure, but only a few articles reporting on results of surgery as the only treatment. The aim of this study was to evaluate the results of a large series of cervicofacial lymphatic malformations in children treated by surgical excision as the primary (first) treatment at a tertiary paediatric referral centre.

2. Methods

This study was conducted at the department of paediatric otolaryngology at Great Ormond Street Hospital, London. All children with lymphatic malformations under the care of the senior author (BEJH) from January 2001 to December 2010 were evaluated. The data was collected from a prospectively collected database with supplemental information from medical records. These patients were managed in a multidisciplinary setting. The diagnosis was made on clinical basis and confirmed with MRI scan in all these patients. The data collected included the anatomical sites involved by the disease, age at the first intervention, need for tracheostomy and the outcomes. These patients were offered surgery, sclerotherapy or a combination of treatment on the basis of symptoms, anatomical sites involved and the nature of the lesion (microcystic or macrocystic). The final decision for primary treatment modality was taken as per parental preference and the team advice. For this study only children who underwent surgical excision as the primary treatment modality were included. The aim of surgery was complete excision for some lesions and subtotal excision for others. Results are therefore expressed in terms of symptom resolution (consistent with sclerotherapy results) rather than completeness of excision. Also excluded were children with intrathoracic extension of the lymphatic malformation that required surgical intervention by thoracic surgeons. The data was collected on the nature and the extent of the surgery.

All the operations were performed by the senior author (BEJH). The aim was to achieve complete resection of the disease where possible. A subtotal excision was performed if it was felt that complete excision may lead to neurological complications or functional impairment. The patients were followed up on a regular basis and the need for further surgical treatment or sclerotherapy was decided on individual basis for the residual or recurrent disease. The data was also collected on the total number of procedures that a patient underwent, complications encountered and the final outcomes. The outcome was recorded as complete response, near complete response or partial response. Near complete response was defined as majority of the disease cleared with minimal residual malformation. Partial response was defined as improved with significant ongoing malformation.

3. Results

On reviewing the records, a total of 118 children with lymphatic malformations were treated at paediatric otolaryngology department at Great Ormond Street Hospital, under the care of the senior author over a 10 year period. Of these patients 53 underwent surgical excision as the primary treatment modality for cervicofacial lymphatic malformations (Fig. 1). Forty-one patients who underwent sclerotherapy as the initial treatment were excluded. Also excluded were 6 patients who underwent thoracic surgery and 18 who were treated conservatively. For the purpose of this study 53 children (27 male, 26 female) with cervicofacial lymphatic malformations were included (Table 1). The age at the time of initial surgery varied from 3 days to 10 years. Of these 19 children were operated under 12 months of age and 46 children under 6 years of age at the primary surgery. In this group, 12 had bilateral neck disease and the rest 41 unilateral. Only 19 patients had the lesion isolated to the neck area. Amongst the rest, the disease was very extensive involving parotid gland in 13 patients, laryngo-pharynx in 4 patients, oral cavity (floor of mouth/tongue) in 16 patients and the submandibular region in 10 patients. Parapharyngeal space was involved in 10 and retropharyngeal space in 2 patients. There was involvement of face in 4 patients. Many of these patients had involvement of multiple sites. As per the MRI findings the disease was macrocystic in 31 patients, microcystic in 9 patients and mixed in 13 patients.

Surgical excision was the primary (first) treatment modality in all these patients. Horizontal skin crease incisions were generally used, with modifications as per the individual pathology. Fifteen (28.3%) of these patients had a tracheostomy before the surgical excision, of which 7 have been decannulated. Inspite of the extensive nature of the lesions, majority of the patients (41, 77.3%) underwent only a single treatment (36 – surgical excision, 5 – laser excision). The remaining 12 patients underwent treatment more than once (twice for 5 patients, thrice for 3, four times for 3 patients and five for one patient). Laser excision was performed for the lesions primarily involving the tongue/tongue base area, using a

Fig. 1. Pre-operative photos of patient with cervicofacial lymphatic malformation.