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Treatment of lymphatic malformations: a more conservative approach

Dror Gilony[a,d,*], Michael Schwartz[b,d], Thomas Shpitzer[c,d], Raphael Feinmesser[c,d], Liora Kornreich[b,d], Eyal Raveh[a,d]

[a]*Department of Otolaryngology, Schneider Children's Medical Center of Israel, Petach Tikva, Israel*
[b]*Department of Imaging, Schneider Children's Medical Center of Israel, Petach Tikva, Israel*
[c]*Department of Otolaryngology-Head and Neck Surgery, Rabin Medical Center, Beilinson Campus, Petach Tikva, Israel*
[d]*Sackler Faculty of Medicine, Tel Aviv University, Tel Aviv, Israel*

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Abstract

Background/Purpose: Lymphatic malformation is a benign disfiguring lesion of the neck and face in children. This study investigated the application and outcome of different modes of treatment.

Methods: The medical files of all children with lymphatic malformation of the head and neck attending a tertiary medical center in 1999 to 2010 were reviewed. Findings were compared by treatment: surgery, OK-432 sclerotherapy, or observation.

Results: The study group included 46 patients, most (65%) with macrocystic disease. Twenty were treated by OK-432 sclerotherapy, and 15, by surgery; 11 (with minor disfigurement) were observed only. Mean follow-up time was 2.4 years. Complete removal or complete response to treatment was achieved in 67% of the surgery group and 45% of the OK-432 group; fair results (>50% reduction in swelling) were achieved in 20% and 50%, respectively. Sclerotherapy failure did not interfere with subsequent surgery. Complete spontaneous regression occurred in 5 patients under observation only.

Conclusions: OK-432 sclerotherapy is associated with good aesthetic results in children with lymphatic malformation. Observation alone is sometimes sufficient. Surgery should be reserved for cases requiring a histologic diagnosis, microcystic disease, patients with an urgent clinical problem (eg, airway obstruction), and sclerotherapy failures.

Lymphatic malformation is a relatively uncommon benign lesion consisting of masses of abnormal lymphatic channels. It most commonly involves the head and neck. The reported incidence is 1 of every 2000 to 4000 live births [1]. Ninety percent of lymphatic malformations are diagnosed before the age of 2 years. The clinical course is variable. The lesions may fluctuate in size, particularly after upper respiratory tract infection, trauma, or inflammation [2].

Although surgery is considered the mainstay of treatment [3], complete excision is frequently impossible owing to the risk of damage to vital structures and the often infiltrative nature of the lesion, with a lack of definite borders to distinguish it from normal tissue. OK-432 (picibanil) sclerotherapy is becoming an accepted alternative, especially for macrocystic disease [2]. OK-432 is an immune modulator

* Corresponding author. Tel.: +972 3 9253367; fax: +972 3 9253273.
E-mail address: drorgil@zahav.net.il (D. Gilony).

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derived from low-virulence *Streptococcus pyogenes*. It stimulates an inflammatory response that causes local inflammation, resulting in regression of the lesion. Its main advantages compared with other sclerosants, such as ethanol or bleomycin, is a lack of major complications and absence of perilesional fibrosis, which allows for subsequent surgery in cases of failure [4]. Because lymphatic malformations are often mainly an aesthetic problem, some patients are observed only, without intervention.

The purpose of the present study was to investigate the application and outcome of the different modes of treatment of lymphatic malformations mainly of the head and neck.

1. Methods

A retrospective design was used. The files of all patients with lymphatic malformation who attended our tertiary medical center in 1999 to 2010 were reviewed for diagnostic workup, mode of treatment, findings at surgery when performed, follow-up, and pathology. Patients with mixed vascular and lymphatic lesions and patients lacking satisfactory follow-up were excluded from the analysis. The study was approved by the institutional Helsinki Committee.

The diagnosis of lymphatic malformation at our institute is based on the typical clinical finding of facial or cervical mass or swelling with confirmation by magnetic resonance imaging (MRI). Fine needle aspiration is not part of the diagnostic process. OK-432 sclerotherapy was introduced as a therapeutic option in our center in 2003. It has since been found to be unfeasible for microcystic lesions (<1 cm diameter) and is currently limited to macrocystic lesions. Surgery is the treatment of choice for patients with microcystic disease or suspected airway compromise and for sclerotherapy failures. Patients with only minor disfigurement and no functional deficit are initially observed and referred for treatment according to symptoms over time.

Sclerotherapy is performed with ultrasound guidance under general anesthesia. The fluid content is partially removed from the fluid-containing spaces, which are then injected with OK-432 (1-3 kilo equivalents). Follow-up clinical and ultrasound examinations are performed after 6 weeks. Additional treatment is recommended for remnant macrocysts, if detected. In cases of failure, follow-up MRI is considered before surgery.

For the present study, the results of treatment were categorized as follows: complete—complete removal of the lesion by surgery or no visible residual mass on clinical inspection after conservative treatment; fair—subtotal removal of the lesion by surgery or a more than 50% reduction in lesion size on clinical inspection after conservative treatment; and poor—only partial removal by surgery or a less than 50% reduction in lesion size on clinical inspection after conservative treatment [5,6]. The study patients were divided by mode of treatment, and the outcome was compared among the groups.

2. Results

Eighty-four patients were diagnosed with lymphatic malformation at our center during the 12-year study period, of whom 46 met the inclusion criteria. These patients were divided into 3 groups by type of treatment: surgery, OK432 sclerotherapy, and observation. Table 1 shows the patient ages, follow-up times, radiologic findings, and treatment results by group. There was no statistically significant association of outcome with radiologic findings or mode of treatment (Table 2).

2.1. Surgery

Fifteen patients underwent a total of 21 operations. These included 9 patients treated before sclerotherapy became available in 2003 and 6 patients who were treated later.

Table 1 Age, follow-up time, radiologic findings, and outcome by mode of treatment in 46 patients with lymphatic malformation

Parameters	Surgery (n = 15)	OK-432 sclerotherapy (n = 20)	Observation (n = 11)
Patient age			
Children/adults	14/1	18/2	11/0
Age of children, range (mean)	2 mo to 10 y (3.6 y)	5 m to 10 y (3.3 y)	1 mo to 14 y (3.75 y)
Follow-up time, range (mean)	0.5-12 y (2.6 y)	0.5-5 y (2.1 y)	0.5-7 y (2.8 y)
Radiologic findings (%)			
Macrocyst (>1 cm)	53%	70%	82%
Microcyst (<1 cm)	13%	5%	0%
Mixed	33%	25%	18%
Treatment results			
Complete removal	67%	45%	45% (spontaneous regression)
Fair (>50% reduction)	20%	50%	55% (minor symptoms)
Poor (<50% reduction)	13%	5%	0% [a]

[a] Patients who had no improvement were usually referred to therapy and therefore were not included in the observation group.