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Lymphatic malformations: A proposed management algorithm

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ABSTRACT

Objective: The aim of this study was to develop a management algorithm for cervicofacial lymphatic malformations, based on the authors' experience in managing these lesions as well as current literature on the subject.

Study design and methods: A retrospective medical record review of all the patients treated for lymphatic malformations at our institution during a 10-year period (1998–2008) was performed. Data collected: age at diagnosis, location and type of lesion, radiologic investigation performed, presenting symptoms, treatment modality used, complications and results achieved.

Results: 14 patients were identified. Eight (57%) male and six (43%) female. There was an equal distribution between the left and right sides. The majority (71%) of cases were diagnosed within the first year of life. The majority of lesions were located in the suprahyoid region. The predominant reason for referral was an asymptomatic mass in 7 cases (50%) followed by airway compromise (36%) and dysphagia (14%). Management options employed included: observation, OK-432 injection, surgical excision and laser therapy. In 5 cases (36%) a combination of these were used.

Conclusion: Historically surgical excision has been the management option of choice for lymphatic malformations. However due to the morbidity and high complication rate associated this is increasingly being questioned. Recent advances in sclerotherapy e.g. OK-432 injection have also shown significant promise. Based on experience in managing these lesions as well as current literature the authors of this paper have developed an algorithm for the management of cervicofacial lymphatic malformations.

1. Introduction

Lymphatic malformations (LMs) are rare benign tumours that result from localized congenital malformations of the lymphatic system. These lesions are diagnosed during infancy in the overwhelming majority of cases and most often present as an asymptomatic mass in the cervicofacial region [1]. The management of LMs remains a challenge. Surgical excision has traditionally been the first line of treatment however due to the close proximity of vital structures this often leads to incomplete excision and recurrence, or damage to vital structures e.g. cranial nerves [2]. More recently the use of OK-432 has gained popularity especially in the management of macrocystic lesions [3]. The authors reviewed the records of all cases treated for LMs at our institution during a 10-year period. Whilst the majority of cases were managed surgically, promising results were obtained with observation alone as well as with OK-432 injection. Based on experience gained in the management of these lesions as well as current literature on the subject the authors of this paper have formulated a proposed management algorithm for LMs.

2. Patients and methods

After obtaining local ethics committee approval, all the patients diagnosed and treated for LMs at our institution between 1998 and 2008 were identified. A retrospective review of these patients medical records were performed focussing on age at diagnosis, location of lesion, type of LM, radiologic investigation performed, presenting symptoms, treatment modality used, complications encountered and results achieved.

3. Results

A total of 14 patients were identified (Table 3). The diagnosis was based on clinical examination and radiologic investigations. Eight (57%) patients were male and the remaining six (43%) female. Two cases (14%) were identified during the prenatal period, five (36%) at birth and three (21%) during the first year of life. The remainder of cases (29%) showed some variation regarding age of presentation with a range of 3–7 years of age. There was an equal distribution between the right and left sides. The location of these lesions was predominantly surpahyoid (86%), with 4 cases

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Table 1
Treatment modalities used.

Treatment modality	Cases
Surgery	5
Ok-432 injection	3
Observation	3
Ok-432 followed by surgery	2

suffering from extensive disease encompassing both the supra and infrahyoid areas. In 2 of these cases there was significant mediastinal extension of the tumour. Only 1 case of exclusively infrahyoid LM was treated at our institution during the 10-year period. The most commonly affected area was the submandibular region affecting 5 cases (36%). Six cases had combined macro- and microcystic disease and 2 cases suffered from predominantly microcystic disease.

The predominant reason for referral to our service was an asymptomatic mass (50%), the remaining patients presented with either airway compromise (36%) or dysphagia (14%). In all cases the diagnosis of LM was confirmed with radiologic investigations. Magnetic resonance imaging (MRI) was the most commonly used modality and was employed in 71% of cases. Other investigations included computed tomography (CT) and ultrasound (US) scanning.

Treatment modalities employed consisted of observation, OK-432 injection, surgery or a combination of these (Table 1). All cases of microcystic disease of the oral cavity were managed with surface potassium–titanyl-phosphate (KTP) laser therapy in order to control symptoms. Of the seven patients treated with surgical excision, four had been operated on at other hospitals prior to referral to our institution. Major complications of treatment included a single case of facial nerve palsy and another of hypoglossal nerve palsy. Both of these cases had undergone previous surgery and the above complications occurred during revision surgery for recurrent disease. Minor complications included seroma and haematoma formation in 2 separate cases.

Two cases (14%) were lost to long term follow up and were therefore not included in the final outcome measurement. Outcome was defined as excellent in cases with complete resolution of the lesion and no residual cosmetic or functional impairment. In cases with minimal residual or recurrent disease, of little concern, the outcome was classified as good. A patient with recurrent or persistent disease staying stagnant or showing some degree of improvement was rated as fair. Finally patients with severe progressive disease showing minimal or no response to treatment were classified as having poor outcome. Favourable outcomes were obtained in 10 cases (83%). Of these five were classified in the "excellent" and the remainder in the "good" outcome groups. Persistent or recurrent disease was encountered in 4 cases. The most common site of recurrent disease was the tongue (50%).

4. Discussion

LMs once referred to as either cystic hygroma or lymphangioma depending on cyst size is now more commonly divided into macrocystic, microcystic or combined disease [4]. The reported incidence of these tumours in the literature is quite variable, ranging between 4 per 10 000 births in one study [5] and 1 per 16 000 births in another [6]. The overwhelming majority of LMs occur in the cervical region with an increased incidence on the left side. There is no difference in distribution between the sexes with both equally affected. The age at diagnosis is reportedly 75% at birth with 90% of the remaining cases diagnosed by the age of two [7]. Two cases in our series (14%) were diagnosed during the antenatal period. Five cases (36%) were diagnosed at birth and 86% of cases were diagnosed within the first 3 years of life. To date numerous theories have been proposed regarding the embryological aetiology of these lymphatic malformations. These include the centrifugal and centripetal theories, the former proposed by Sabin and the latter by McClure and Huntington, as well as the combined theory proposed by van der Jagt and Kutsuna [3,7]. Regardless of the proposed theory the final result is failure of either the peripheral lymphatics to flow into the jugular sacs or failure of the jugular sacs to reunite with the venous system [7]. It is also widely accepted that LMs can be acquired secondary to surgery, trauma, infections, neoplasms and chronic inflammation [3,7]. The histopathological classification of LMs as proposed by Kennedy in 1989 includes four distinct groups [7]. It is widely accepted however that all these lesions form part of the same disease process [8].

The majority of cases (43%) in our series presented with an asymptomatic mass in the cervicofacial region. The remainder either presented due to airway concerns (36%) or dysphagia (21%). These findings are consistent with previously reported presenting symptoms and signs [9]. In the group that presented with airway compromise, three patients required tracheostomies two of which were emergent. Infective episodes as well as haemorrhage into these cysts are quite common, often leading to rapid enlargement of the cyst with potential airway compromise [10]. This can occasionally, as illustrated in our case series, require an emergency tracheostomy. Current guidelines on the management of these episodes advocate the use of parenteral Gram positive coverage for up to 3 weeks followed by a prolonged course of oral antibiotics [10]. In our case series 64% of patients developed at least one such episode whilst others suffered from recurrent infections, all of which were successfully managed medically. A recent report by Sires et al. [11] showed promising results with use of systemic corticosteroids in the treatment of these episodes in ophthalmic LMs.

In an effort to predict the prognosis of LMs several staging systems have been proposed. Orvidas and Kasperbauer [12] used the variables of functional impairment, cosmesis, number of locations and age at diagnosis in order to formulate a staging system. They demonstrated an increase in persistence as well as complication rate with increasing stage. However the most commonly used staging system was developed by de Serres et al. and published in 1995 [13]. It is based on the anatomical location of the LM and consists of five stages (Table 2). In their paper they described a clear correlation between the stage of the disease and the prognosis as well as associated complication rate. Group 1 patients had a complication rate of 17% compared to 67% in group 3 and 100% in group 5. Clearly demonstrating a progressive increase in complication rate associated with higher staging. In a retrospective study of 22 cases Hamoir et al. [14] applied the proposed staging system and reported findings consistent with those of de Serres et al. In our case series 7% were classified in group 1, 64% in group 2 and 29% in group 3. There were no patients in group 4 or 5. Our results reflect the findings of de Serres as well as Hamoir et al. with a more favourable outcome

Table 2
Staging of LMs.

Stage	Description
1	Unilateral infrahyoid
2	Unilateral suprahyoid
3	Unilateral infrahyoid and suprahyoid
4	Bilateral suprahyoid
5	Bilateral suprahyoid and infrahyoid