CHAPTER 3

Cystic Hygroma

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INTRODUCTION

Lymphangiomas are benign masses with multinodular cysts of different sizes and contents. Microcysts are less than 1 cm in diameter; macrocysts are greater than 1 cm in diameter and tend to be less invasive, less numerous, and less difficult to remove. Both microcysts and macrocysts may contain blood and/or lymph, a consequence of similar lymphatic and vascular embryology. In general, microcysts are more likely to contain blood and macrocysts more likely to contain lymph. Macrocysts that contain lymph are also called cystic hygromas and they are subsumed in the general category of lymphatic malformations.

The risks of expectant management include infection, progressive growth and disfigurement, extension into previously uninvolved areas, dysphagia, airway compromise, and erosion into vascular structures. Asymptomatic cysts in the premature or small-for-dates child may await growth and development of the infant. For the majority of patients there is no need to defer excision.

The determination of a lymphangioma's size and character is based on location, clinical examination and investigation. Some regions tend to have typical lesions: for example, reddish lesions in the base of the tongue are typically microcystic with a significant vascular component; soft boggy masses in the superficial neck or axilla – sometimes with a bluish hue – are often macrocysts with lymph. The best investigations to determine cyst contents is either a T2-weighted gadolinium-enhanced magnetic resonance imaging (MRI) or needle aspiration of the dominant cyst. Lymph is straw-coloured; thin bloody fluid may occur when a lymphatic cyst is enlarged by a ruptured blood vessel. Abundant dark or red blood indicates a significant vascular component. Viscid yellow-clear fluid from an intra-oral lesion may signal a ranula, deriving from salivary tissue. Depth of invasion and an estimate of the structures involved is best determined by MRI scanning. Rarely, a neck lesion may extend to the anterior mediastinum and compress the trachea. Spontaneous enlargement may occur following an upper respiratory tract infection; spontaneous regression is rare although sometimes follows local infection

Figure 3.1

General anaesthesia is used and blood made available if the lesion appears vascular on pre-operative screening. If lesions are close to important motor nerves, one may use a nerve stimulator and interdict use of musculoskeletal blocking agents.

Pre-operative planning will usually demonstrate a safe plane of attack and may set expectations with regard to a complete excision or a debulking operation. Loupe magnification is often helpful, as is a bipolar cautery when working close to nerves or vital structures. Microvascular lesions tend to infiltrate tissue planes, are more likely to bleed and have a high rate of recurrence. Macrocystic lesions tend to spread along fascial planes and around neurovascular structures. Intra-operative rupture decreases the likelihood of complete resection, which averages 50%. Any residual cystic tissue will increase the likelihood of recurrence. Because this is not a malignant lesion, it is seldom necessary to sacrifice essential local structures. It is commonly necessary to place a closed suction drain, particularly when the lesion is incompletely excised. For the most common (cervical) lesions, a transverse skin crease incision extending the length of the mass is placed in Langer's lines. A first-generation cephalosporin is used peri-operatively.

Figure 3.2

If the lymphangioma demonstrates dermal infiltration, an ellipse of skin is removed. Otherwise, generous sub-platysmal skin flaps are raised. The external jugular vein and ansa cervicalis are not considered essential and may be sacrificed.

Figure 3.3

Dissection of cervical lesions begins at the superior margin of the mass, near the ramus of the mandible. Upward reflection of the facial artery and vein allow the precise visualization necessary to preserve the marginal branch of the facial nerve. Bipolar cautery may be used and optical magnification is often helpful.

Figure 3.4

The dissection proceeds medially, lifting the cyst from the surrounding alveolar tissue.

It may be necessary to divide the middle thyroid vein and artery as the carotid sheath is approached. Deep dissection frequently involves the contents of the carotid sheath and sometimes the following nerves: vagus, spinal accessory, hypoglossal, sympathetic trunk, phrenic and the brachial plexus.

Figure 3.1

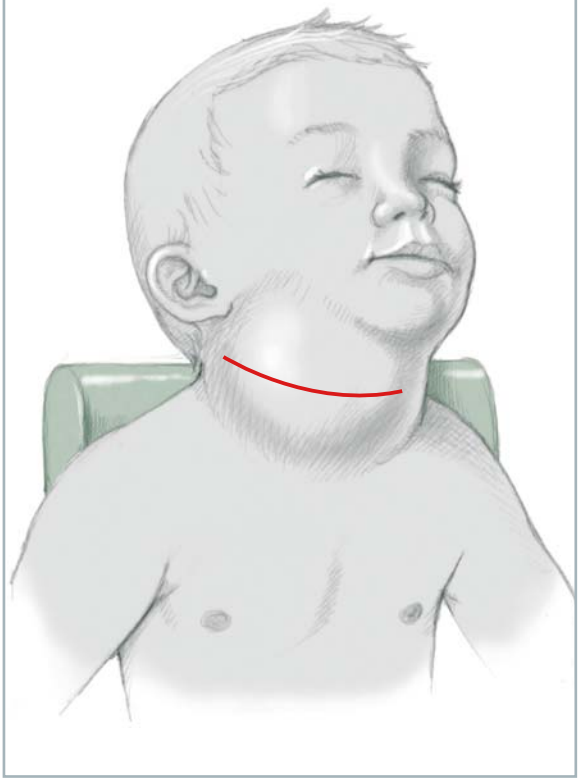


Figure 3.2

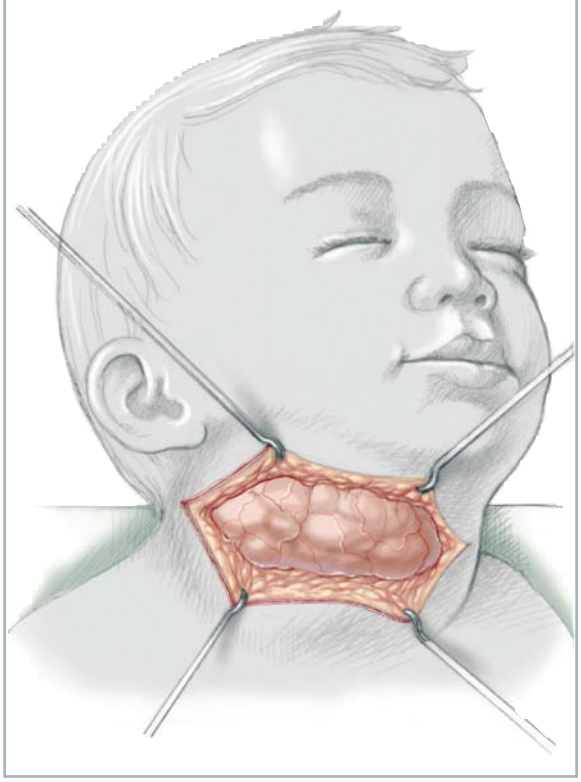


Figure 3.3

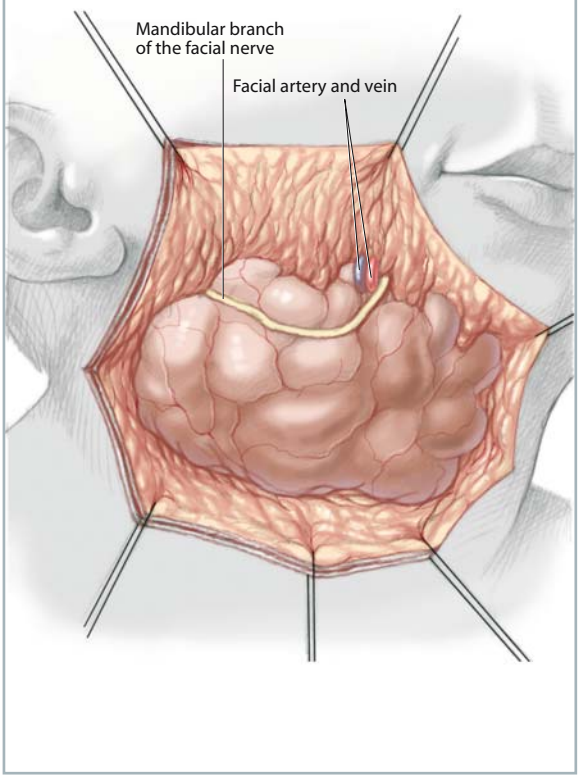


Figure 3.4

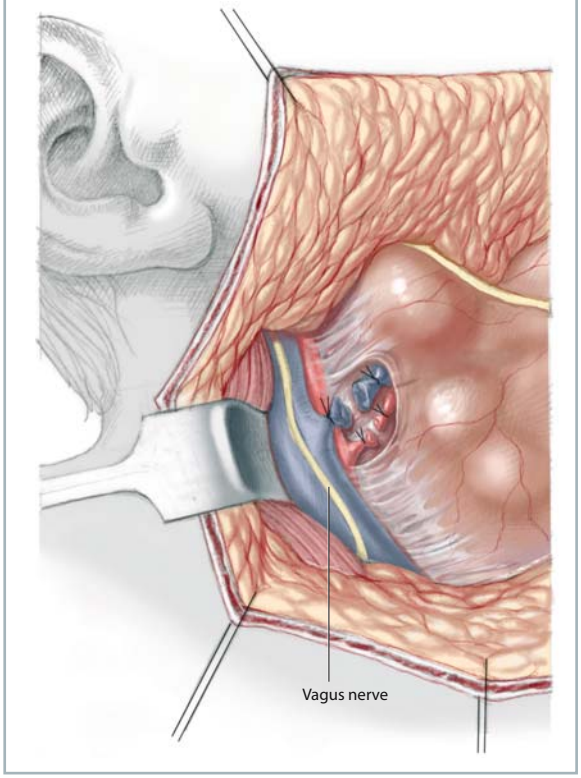


Figure 3.5

Care is taken to preserve the hypoglossal nerve as it passes through the bifurcation of the carotid artery. The mass must then be freed from the hyoid bone and submandibular gland. It is rarely necessary to remove the submandibular gland en bloc with the mass, sacrificing the facial artery. The mass may be adherent to the brachial plexus in the floor of the anterior triangle or the spinal accessory nerve as it courses through the posterior triangle. Extension of the lymphangioma under the clavicle may lead to axillary or mediastinal involvement (requiring sternotomy if the lesion proceeds deeply). Combined masses may be delivered either above or below the clavicle.

Figure 3.6

The platysma is re-approximated with fine absorbable sutures and the skin closed with subcuticular sutures of similar material. Closed suction drainage is used for most lesions.

Figure 3.5

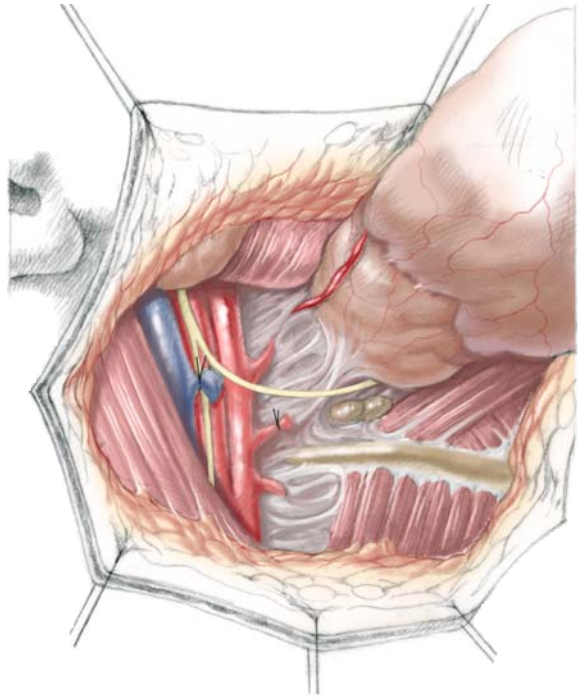
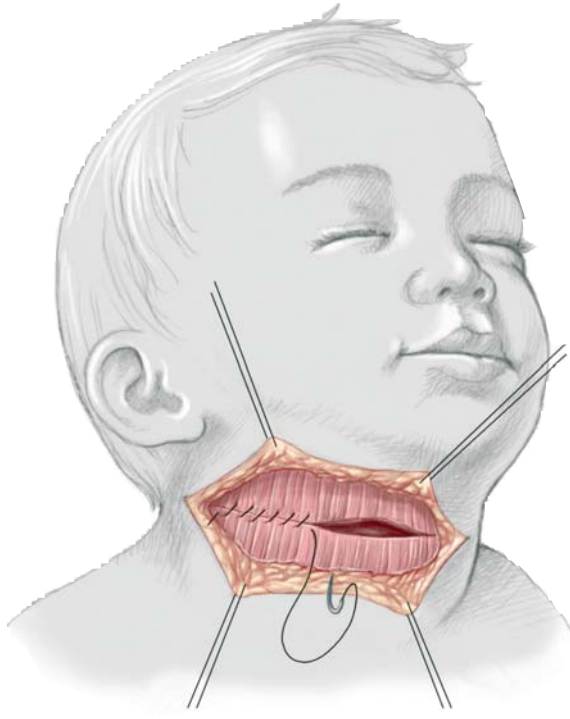


Figure 3.6



CONCLUSION

Feeding resumes when the infant is awake and alert. Extensive intra-oral dissection may temporarily impair swallowing and delay the onset of oral feeds. Drain removal may take days or weeks and is dictated by the daily drainage volume. Antibiotics are administered daily from 1 to 3 days.

In cases of partial resection, recurrence typically occurs within a year of surgery. Lymph leaks and nerve injuries are minimized by the use of bipolar diathermy. Rarely, lymph leaks may require re-exploration when drains are inadequate or removed early.

Excision is the current gold-standard therapy. There are several reports of successful use of sclerosing agents such as OK-432 or bleomycin in lymphangiomas. This appears to be effective mainly in macrocystic lesions.

An exciting advance in the management of fetuses with a high probability of upper-airway obstruction at birth due to a giant cervical lymphangioma, is the development of the ex utero intrapartum treatment (EXIT).

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