Temporal Space Lymphatic Malformation in a 15-Year-Old Adolescent: An Extraordinary Case

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Abstract Lymphatic malformations—previously called lymphangiomas or cystic hygromas—are regarded as non-malignant primary disorders of the lymphatic system. They appear predominantly in infants and children, with 90 % of cases being diagnosed by the age of 2 years. Also, they constitute an infrequent entity, accounting for 5 % of all benign tumors in infants and children. In adults they are extremely rare. In the present article we present an extraordinary case of a lymphatic malformation that ensued in the temporal area of a 15-year old adolescent.

Keywords Lymphatic malformation · Lymphangioma · Cystic hygroma · Temporal space · Surgical management

Introduction

Lymphatic malformations (LMs), previously called lymphangiomas or cystic hygromas, are now classified as low flow vascular malformations [1]. They are regarded as a non-malignant primary disorder of the lymphatic system,

however the exact mechanism of their pathogenesis has not been adequately elucidated yet [2]. LMs appear predominantly in infants and children, with 90 % of cases being diagnosed by the age of 2 years [3, 4]. Also, they constitute an infrequent entity, accounting for 5 % of all benign tumors in infants and children [3, 5]. In adults they are extremely rare [6]. In the present article, we describe an extraordinary case of a LM that ensued in the temporal area of a 15-year-old patient.

Case Presentation

A 15-year-old adolescent male was referred to our outpatient department, due to a swelling at the left temporal area. The swelling had appeared 4 days earlier and had gradually grown since then. The patient was otherwise healthy and reported no recent injury or infection. Clinical examination showed a non-fluctuant, compressible, and mildly tender mass, located superiorly to the zygomatic arch, measuring approximately 4 × 3 cm (Fig. 1). The patient exhibited no trismus and was afebrile. The rest of the clinical examination was non-contributory. A laboratory work-up with complete blood cell count, biochemical panel and inflammatory markers was performed. The biochemical panel included: urea, creatinine, conjugated and unconjugated bilirubin, aspartate and alanine aminotransferases, alkaline phosphatase, and γ -glutamyl-transferase, while the inflammatory markers assessed were C-reactive protein (CRP) and erythrocyte sedimentation rate (ESR). All the above turned out to be within normal limits.

One would begin the investigation of such a case with an ultrasound scan. However, the exact extent of such a lesion, along with the possible invasion to adjacent tissues and most importantly a potential brain involvement can be

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Fig. 1 Photograph of the patient. A swelling at the left temporal area, measuring about 4×3 cm, is depicted

adequately assessed by a computed tomography or a magnetic resonance imaging scan (MRI). Since we had to deal with a soft tissue lesion an MRI was selected as the best modality. The described lesion had demonstrated potential of rapid growth, so if a brain involvement was present, it would be a case of emergency. Given these, and after neurosurgical consultation we decided to immediately proceed with a contrast-enhanced MRI scan, omitting an ultrasound. The MRI obtained, on T1 sequence, revealed a mass of soft tissue, occupying the space between the temporal fascia and the temporalis muscle. The mass appeared well-circumscribed with no sign of invasion to the adjacent tissues, contained septa and demonstrated heterogeneous signal intensity. Namely, there are areas of low, mixed with areas of high signal intensity (Fig. 2).

The list of differential diagnosis of a temporal swelling includes the rough categories of an infection, a neoplasm, a cystic lesion and other tumor-like lesions. As far as the present case is concerned, an infection alone seemed unlikely, because the clinical and radiological investigation revealed no potential infectious focus. Additionally the lesion had rapidly gained significant size, thus if it had been of infectious origin (e.g. an abscess), it would have caused more severe symptoms, fever and elevation of white blood cells count and laboratory inflammation markers. None of these was present. On the other hand, no neoplasm, cystic or other lesion alone could probably gain

Fig. 2 MRI image, on T1 sequence, showing a mass of soft tissue, occupying the left temporal space

significant size within only 4 days. On the basis of this analysis, the most probable diagnosis would be a pre-existing lesion, which rapidly enlarged triggered by a sub-clinical infection. Actually, this is frequently the first manifestation of many cystic and tumor-like lesions of the head and neck. This hypothesis was verified by the MRI, which showed a mass with benign imaging features. As the lesion appeared to be solid, a cystic lesion was rendered unlikely, therefore the diagnoses of a benign tumor or a tumor-like lesion comprised the greatest probability. Entities belonging to these two categories and also being compatible with the age of the patient were: muscular tissue tumors such as the rhabdomyoma or the leiomyoma, tumors of the fatty tissue such as the lipoma, peripheral nerve tumors such as the schwannoma, a vascular malformation, and finally a LM.

Aspiration of the lesion was not undertaken, because the results of the cytological examination would require 2–3 days, which would cause a delay in the diagnosis and treatment of the patient. Besides, whatever the possible diagnosis suggested by the cytological examination, a biopsy would be mandated to verify it. As the lesion demonstrated benign features and was relatively easily accessible, an excisional biopsy was decided. The lesion was approached via a preauricular incision with temporal extension. The temporal fascia was identified, incised and readily elevated, discovering the lesion, which was a mass lying on the temporal muscle. The mass resembled fat and was non-encapsulated and hypovascular. Additionally, it was loosely adherent to the underlying muscle tissue, therefore a plane of dissection was easily identified

(Fig. 3). The mass was dissected off by means of scissors and digital pressure and no removal of normal tissue was required (Fig. 4).

The histopathological examination of the lesion revealed multiple dilated and irregular channels lined by endothelial cells and surrounded by adipose and fibrous tissue. These findings were compatible with the diagnosis of macrocystic LM (Fig. 5).

The patient presented an uneventful postoperative recovery, with normal function of all branches of the facial nerve. On follow up after 6 months there was no sign of recurrence (Fig. 6).

Discussion

As aforementioned, in 90 % of cases, LMs are diagnosed by the age of 2 years. They may also ensue in older ages, very rarely though. It is believed, that lesions of adults may congenitally present and may not become clinically apparent until adulthood, when they start enlarging. Trauma or infection may act as trigger for the enlargement process [5]. In our case, the patient was 15 years old and the lesion had appeared suddenly, 4 days before his presentation at hospital. However, the patient did not report any injury or respiratory infection. Moreover, laboratory values and inflammation markers were within normal limits. Therefore, we may hypothesize that the enlargement of the lesion was induced by a minor skin infection of the scalp or a subclinical viral infection of the upper respiratory system.

Fig. 3 Intraoperative photograph of the mass

Fig. 4 Photograph of the excised tumor

Fig. 5 Photomicrograph of the lesion, showing multiple dilated and irregular channels lined by endothelial cells and surrounded by adipose and fibrous tissue (Hematoxylin & Eosin 200×)

The typical clinical presentation of a LM includes a non-tender, soft, doughy and transilluminating mass [5, 7]. Our lesion was compatible with this pattern, yet it was mildly tender. According to Naidu and McCalla, over the period 1828–2004, only 91 cases of LMs in adults had been reported [6]. In addition, LMs occur most commonly in the neck [7]. The presented case comprises three extraordinary features. First, the lesion presented in the temporal region. Second, the patient was a 15-year-old adolescent. Third, the lesion had grown very fast, actually within 4 days. Furthermore, the patient had negative history for trauma

Fig. 6 Postoperative photograph of the patient

and infection. This cluster of rarities renders the present case unique.

MRI is the imaging modality of choice for the investigation of a possible LM [5, 7, 8]. The principal advantages of MRI are the better delineation of the anatomical extent of the lesion and its relationship to the adjacent tissues. Contrast enhancement with paramagnetic substance provides additional information about the vascularity of the lesion. On MR images a LM typically presents as a mass with low or intermediate signal intensity on T1 sequence and high signal intensity on T2 sequence. Infrequently a LM may appear hyperintense on T1 sequence, a finding associated with clotted blood or lipid content within the lesion [9, 10]. In the current case, MRI depicted a septated lesion of heterogeneous signal intensity. This imaging pattern may be attributed to the microscopic structure of the presented LM, which comprised dilated channels—producing low signal—coupled with adipose and fibrous tissue—producing high signal on T1 sequence.

LMs are benign lesions, however they rarely resolve spontaneously and they often attain significant size, effecting esthetic deformity and functional disorders and occasionally compromising vital structures of the head and neck [3, 7, 8]. Small and asymptomatic lesions may initially be left untreated, provided that close follow up be ensured [4, 7, 8]. Otherwise, treatment is indicated. The traditional treatment of choice of a LM is surgery, which

provides cure in cases of complete excision of the lesion [7, 11]. Postoperative rates of complications and recurrence as high as 12–33 and 15–53 % respectively are reported [5]. Microcystic lesions do not respect tissue planes. Consequently, they are the most difficult to excise, and thus associated with the highest recurrence rates [12]. Sclerotherapy is advocated as the main alternative to surgery. Several sclerosing agents have been implemented [5, 8, 9, 13]. Their most severe complication is unpredictable and extensive scarring, due to penetration into adjacent tissues. Other conservative modalities, including laser ablation, radiofrequency ablation and cryotherapy have also been introduced, with ambiguous results though [4]. Regarding our case, the lesion demonstrated potential of rapid enlargement and caused esthetic disfigurement to the patient. Therefore, therapy of the lesion was indicated and surgical excision was considered as the optimal modality. A preoperative incisional biopsy was considered unnecessary, given the benign clinical and imaging pattern, coupled with the small size of the lesion.

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