

Venolymphatic Malformations: A Pediatric Case Study and Overview

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Abstract

Introduction: Venolymphatic Malformations are rare abnormalities that form in utero and are typically diagnosed within the first few years of life. They are diagnosed by imaging, with MRI being the gold standard and Ultrasound as the next option. Symptoms can range from none to large areas of swelling in the face and neck and are the most common areas where these malformations are found. Treatment may vary according to the severity of the malformations and may include observation, sclerotherapy, as well as surgery.

Objective: Report on the diagnosis, treatment and outcome of a patient with Venolymphatic Malformations.

Methods: Case report of a patient with facial and orbital disfigurement due to Venolymphatic Malformations, who was diagnosed at three years old through MRI, US and treated with two separate sclerotherapy treatments.

Results: After each treatment, the patient's malformations decreased and/or remained stable.

Conclusion: The sclerotherapy was successful at halting the growth of the malformations and reduced the swelling.

Keywords: Venolymphatic Malformation, sclerotherapy, bleomycin, congenital vascular disorders, MRI imaging of venolymphatic malformations.

Introduction

Venolymphatic malformations, or VLMs (previously referred to as lymphangiohemangioma or hemangiolympangiomas) are rare types of deformities which are comprised of venous malformations combined with lymphatic vessels that can also include arteries and capillaries [1]. Venolymphatic malformations are abnormal congenital defects that result from an interference in the embryologic vascular system development. In some patients, they may not be seen until a year or more into the child's life [2,3]. The malformations continue to grow as the child grows. Their increase in size is due to lymph flow, bleeding within the lesion, and inflammation. [1,4]. Furthermore, malformations can grow rapidly with pregnancy or during puberty and at those times tend to have more symptoms [5]. One of the most common features of VLMs is that the venous channels have a nonexistent muscular wall or very thin walls due to the developmental error in blood vessels during the embryonic phase [5]. In comparison to arteriovenous malformations, or AVMs, which are abnormal connections between arteries and veins, venolymphatic malformations are slow-flow lesions, meaning the vascular flow is much slower [4]. Interestingly, the malformations are at .1 to 2% of the population [6]. Moreover, they are much more common in females with a 9:1 female to male ratio. Venolymphatic malformations are commonly found in the head and neck regions of the body but have been also found in the leg, heart intestine, heart or lung spaces. Sadly, the malformations can affect the patient's appearance by causing visible changes to the skin resulting in aesthetic concerns. Additionally, VLMs can cause serious health issues depending on where they are located. Airway obstruction can be a main concern if a VLM is located on the head or neck [4].

Symptoms

Venolymphatic malformations are abnormal developments to the structure of veins and lymph vessels. Due to these vessel abnormalities and lack of lymphatic fluid drainage, patients can experience a wide range of health issues, such as swelling, pain,

infection, and deformity where the fluid can build-up [4]. One of the main symptoms commonly seen are masses on the head or neck appearing as swollen areas that may or may not be painful. When present on the face, there is swollen asymmetrical facial distortion and displacement. Additionally, the lack of drainage to the venous and lymph fluid in the malformations can lead to serious infections such as cellulitis due to the fluid pooling and buildup. These infections could require prolonged courses of antibiotics. Additionally, raised dark red marks may appear on the skin, called lymphangiomas, which are a mixture of lymphatic fluid and blood that have accumulated within the abnormally formed vessels. Also, there is a possibility of abnormal bone growth to the jaw and teeth due to increased blood flow and pressure to the bone [4]. Furthermore, size and location play a large role in the severity of symptoms. For example, smaller VLMs may cause only minor symptoms, while larger VLMs can lead to many more issues [2]. Additionally, the location of VLMs can create greater risk of complications. For example, if a VLM is close to the orbit, it can be swollen, possibly eye redness, and globe displacement from the lack of lymph fluid being drained in the malformation, while if there is a VLM on an extremity, the symptoms may not be as severe [7,8].

Diagnosis

Venolymphatic malformations are diagnosed with several different imaging modalities but are usually first found by patient history and clinical characteristics, if present [2]. A CT scan, which is an imaging test that uses x-rays and a computer to create detailed images of the bones and soft tissues of the body, may be the initial route to determine the mass or swelling that has been detected by the physician but is not widely chosen if the malformation is suspected due to this modality producing a large dose of radiation for the patient [4]. Plain radiographs may detect phleboliths which are small, calcified blood clots that can be commonly seen with venolymphatic malformations [4]. Color Doppler Ultrasound, which is a specialized ultrasound that shows blood flow in vessels and organs by converting sound

waves into color-coded images, is used to show the vascular and lymph flow in the malformation. Blockages will be seen in this case study [9]. The most affective modality for evaluating venolymphatic malformations is Magnetic Resonance imaging (MRI), because of its ability to differentiate between venous and lymphatic components [4]. Specifically, with this modality, physicians can identify the shape and size of the malformation, due to MRI's superior soft tissue visualization. Additionally, this can help significantly if surgery is the chosen treatment option.

Treatment

The method of treatment will depend on the location and size of the malformation. It is used to control lesion size, pain, bleeding, loss of function, and compressed nerves [2,3,4]. If there is an absence of pain or other symptoms, observation is the first step. If there are signs of infection, antibiotics, such as amoxicillin, are prescribed to eradicate bacteria from low flow malformations, evidence shows longer than standard courses from 6-24 weeks of treatment may reduce the possibility of the infection recurring [2]. Laser therapy is a selective photothermolysis where laser energy is preferentially targeted at abnormal vessels with very little damage to normal surrounding normal tissues. This is an option for malformations on or close to the skin surface. Sclerotherapy, which seals or blocks off the vessels that are draining, can be used to block the lymph and blood from passing through fibrosis of the lesion. It is considered safe and effective. Bleomycin, a chemotherapy drug, is the most used sclerosing agent. It is highly effective in the treatment of venolymphatic malformations and has been recorded on multiple patient surveys [3,4,6,10,11]. The last option is surgery to remove the malformation if possible. By using imaging, doctors can determine the size and exact location and make sure no other structures are at risk during the surgery first [4].

Case Report

Female patient, 15 years old, was being fostered by a couple who noticed swelling in her right cheek and right eye at the age of 3. She was initially taken to her pediatrician who referred them to an ear nose and throat doctor who recommended they take her to Children's Hospital, which unfortunately had a month waiting period to be seen. That week the couple noticed the swelling in the cheek and eye as well as the eye redness with bulging became so bad that they went to Children's Emergency Room to be seen immediately. The patient was given a maxillofacial and soft tissue neck CT scan with contrast which found multiple densities in the subcutaneous fat over the right side of her face and around her right eye with possible vascular formations but no high flow lesions. The next day she had an Angio-brain orbit MRI with contrast as well as a soft tissue Ultrasound. The orbit MRI showed multiple lesions on the right side of her face and orbit that went into her hard palate that were intra and extra-conal. By the fluid level they diagnosed the lesions to be venolymphatic malformations in the right orbit and in the cheek. (See Figures 1-3).



Figure 1: MRI image of Venolymphatic Malformation in the right eye enhanced with contrast that was found and led to the diagnosis of the disorder.



Figure 2: MRI image of Venolymphatic Malformation in the right cheek enhanced with contrast that was found and led to the diagnosis of the disorder.

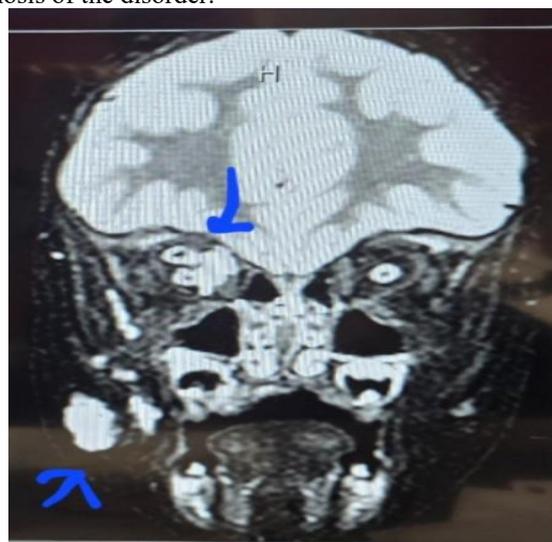


Figure 3: In this image you can see the right orbital Venolymphatic Malformation as well as the one in the right cheek region, both highlighted with contrast during the MRI study. This was pre sclerotherapy.

The right orbit malformation approaches the optic nerve and back of the globe. They waited three months to watch the malformations which increased in size then set a date to an Angio-Percutaneous Sclerotherapy of the right cheek and right orbit. (See *Figure 4*).



Figure 4: The patient at 3 years old before sclerotherapy treatment with right eye and cheek Venolymphatic Malformation diagnosis.

The patient was under anesthesia while 3% of Sodium Tetradecyl Sulfate foam was injected un ultrasound and fluoroscopy guidance into the soft tissue of the right cheek. After a digital subtraction contract injection was put into a lymphatic microcyst with the superomedial quadrant of the right orbit, then an injection of bleomycin under ultrasound and fluoroscopy was placed. Both procedures were successful. (See *Figure 5*).

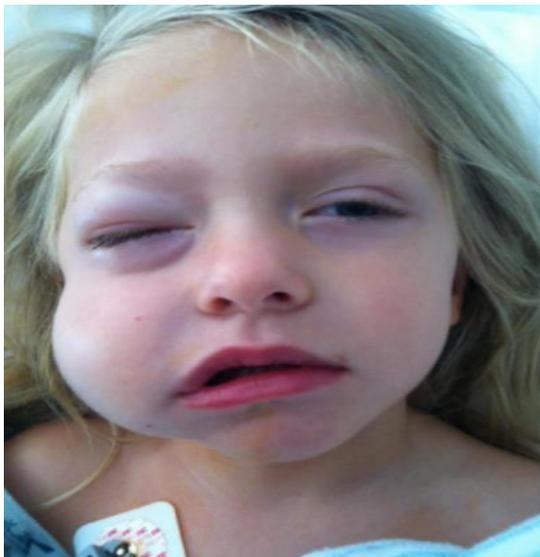


Figure 5: In this image the patient has undergone right cheek sclerotherapy and exhibits swelling and right eye edema.

The patient was released and returned five months later for a follow-up MRI and Ultrasound which showed diminished T2/stir intensity and no enhancement correlating to sclerosis in the cheek and orbit. There was no growth, and they were stable. A year and a half later the patient and family returned to Children's Hospital with swelling symptoms and was given another MRI with contrast and an ultrasound. The results

showed right conjunctival swelling due to the increase in the size of the right orbit venolymphatic malformation. The physician recommended another sclerotherapy treatment and scheduled one in four months. The patient was now 5 years old and under anesthesia a Percutaneous sclerotherapy of the right orbit was performed. Under the guidance of ultrasound, an injection of bleomycin opacified with contrast was advanced on the medial side of the orbit into the retrobulbar intraconal cysts. Multiple C-arm cat scans were obtained during the procedure to guide needle position into the retrobulbar cysts. The procedure was successful, and the patient was noticed to have severe conjunctival edema. Five months later a follow-up MRI showed a decreased caliber of the right orbital malformation and stated it was stable. The parents stated the swelling slowly decreased and as the blood drained from the eye malformation it caused the cheek to bruise.

Ten years have passed, and the patient has now gone through puberty. Unfortunately, the venolymphatic malformations in her right eye and right cheek have started to swell and have become painful. She had an ultrasound and MRI with contrast which revealed the orbital malformation has a slight decrease from her last MRI while the malformations in her cheek have increased. The patient states she has pain in both areas and is scheduled for another sclerotherapy procedure to both areas in a month.

Conclusion

Venolymphatic malformations may require patients to return for treatment if they are symptomatic over their lifetime due to the lymphatic and venous vessels swelling with lack of drainage and forming cysts [4,12]. Percutaneous intralesional sclerotherapy is recognized as a gold standard treatment due to its success and effectiveness. The patient in this case study has had success with her sclerotherapy in both areas of her malformations. As she grows and her hormones change, so do her malformations. The one close to her right orbit is of concern due to it being close to her optic nerve. Her parents and doctors will keep an eye on the development of these malformations and offer her options for treatment.

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Conflict of Interest

The author declares that they have no competing interests.

Dedication

To my stepdaughter, who at a young age was given the diagnosis of Venolymphatic Malformation. I dedicate this article lovingly to you, to remind you that you are more than veins and vessels that are intertwined. Having this diagnosis has brought challenges that only a few can understand, yet you bring a light and smiles to each day. You never complain about your rare condition; you are truly an inspiration and a blessing to know and love. It is my hope that the future brings new ways of ending this disorder and this article be a step in awareness, but also to acknowledge the strength that you display every single day.

With all my love

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