CASE REPORT

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SCLEROTHERAPY TO A LARGE CERVICOFACIAL VASCULAR MALFORMATION: A CASE REPORT WITH 24 YEARS’ FOLLOW-UP

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Abstract: Background. Large craniofacial venous malformations frequently cause significant cosmetic and functional problems. Treatment of the lesions early in life helps to avoid these problems. We present a case of a large cervicofacial venous malformation.

Methods. The patient was treated with a 5% benzyl alcohol solution of sodium morrhuate. The treatment was begun when the patient was 3 months of age, with a total of 23 injections.

Results. The lesion had completely disappeared by age 10. No major complications were observed except a superficial tissue loss on the temporal region. It was healed by conservative treatment. There were no recurrences during the 14-year follow-up.

Conclusions. It is difficult to conclude that this type of treatment is the best choice for patients with venous malformation on the basis of a single case. However, intralesional sclerotherapy should be kept in mind as a savior treatment option in cases of large venous malformations in anatomic regions that present challenges to both the surgeon and the patient. Intralesional sclerotherapy can be performed without serious complications if the sclerosing agent is selected and injected cautiously.

Keywords: venous malformation; sclerotherapy; head and neck
the lesion may be normal to deep blue color depending on the amount of dermis invasion. These malformations can be localized as a solitary lesion or can involve extensive areas of the body. They are usually located on the face, limbs, or trunk and can occur in internal locations, such as oronasopharynx, brain, bladder, spinal cord, skeletal muscle, and bones. They are found in several vascular syndromes (eg, Klippel Trenaunay, Maffucci, blue rubber bleb nevus, and familial cutaneomucosal venous malformations). Visual superficial varicosities may also be present. Phleboliths, which are characteristic and distinguishing features of low-flow venous malformations, may be detected on plain radiographs.

Large craniofacial venous malformations often cause significant cosmetic and functional problems. Treatment of the lesions early in life avoids these complications. Sclerotherapy with various agents has been a primary treatment for venous malformations.

We present a patient with large extensive craniofacial venous malformation treated only with intralesional sclerotherapy with sodium morrhuate, which was started at 3 months of age and repeated until the patient was 10 years old. The patient was followed up another 14 years after the completion of treatment.

**CASE REPORT**

A 3-month-old girl was referred to our clinic with a large head and neck mass on the left side (Figure 1). According to the history provided by the girl's parents, the mass was apparent at birth. The pregnancy was uneventful, and there was no family history of vascular malformations or other congenital anomalies. On physical examination, a large blue mass on the left cervicofacial area was seen (Figure 2). There was not any palpable thrill on the mass. The mass was soft and nonpulsatile. Slow refilling could be detected after compression was relieved. Examination also showed that the lesion was likely to enlarge during crying. Telangiectatic lesions were present over the mass. The patient had strabismus on the ipsilateral eye. The left upper eyelid was also ptotic, with cleft and coloboma-like deformity (Figure 3). Plain radiographs showed phleboliths in the mass. We were unable to perform a CT scan or MRI, because these modalities were unavailable in our country at that time. The mass was initially diagnosed as cavernous hemangioma.

We planned intralesional sclerotherapy for treatment. A 5% benzyl alcohol solution of sodium morrhuate (Varicocid; Chemishe fabric Kreussler, Germany) was used as the sclerosing agent. Sodium morrhuate is obtained from the fatty acids in the liver of morrhua. Treatment was started when the patient was 3 months of age. Each injection was performed with the patient under general anesthesia. One milliliter of solution was administered into the channel of the mass as soon as blood was seen in the syringe. Localized edema and redness occurred because of the inflammatory effect of sodium morrhuate after the injection. The edema resolved in 4 weeks. (The family members were counseled for this reaction.) In the first year, the injections were repeated every 2 months. In the following years, the injection intervals were 3 months, 4 months in order, and then twice yearly injections. The total number of injections was 23, at a dose of 1 mL per injection. No major complications were encountered. When the patient was 8 years old, a superficial tissue loss measuring approximately 2 × 3 cm developed on the temporal region; this lesion healed with conservative treatment within...
3 weeks. We completed the treatment when the patient was 10 years of age. The mass completely disappeared in 10 years. Strabismus healed as well. Although the blepharoptosis gradually decreased, it needed to be corrected with the levator plication technique when the patient was 15 years old. Strabismus gradually improved but did not completely resolve; thus, ophthalmic surgeons performed a corrective operation on the patient when she was 17 years of age. No recurrences were detected during the 11-year follow-up period (Figures 4–6).

**DISCUSSION**

A vascular malformation is composed of dysmorphic vessels that are believed to result from faulty embryologic development. These malfor-
motions are subcategorized as arterial, capillary, lymphatic, or venous disorders, depending on their predominant channel type. These lesions are also classified according to their rheologic behavior. The capillary, lymphatic, and venous malformations are slow-flow lesions, whereas the arteriovenous malformations are high-flow lesions.

Venous malformations are the most common vascular abnormality, especially in the head and neck region. They are usually present at birth. They grow proportionately with the body. Spontaneous enlargement may be seen in response to trauma, infection, or hormonal changes, all of which may cause thrombosis, ectasia, or development of new arteriovenous communications. They almost never regress, and they often expand. Histologically, venous malformations are collections of abnormal vessels with normal endothelium and normal mast cell count. They have a wide spectrum of clinical presentation, from simple varicosities and telangiectasias to complex anomalies involving several tissue planes (as in our case). They are also found in several vascular syndromes (eg, Klippel-Trenaunay, Mafucci, blue rubber bleb nevus, multiple familial glomangiomias, and familial cutaneomucosal venous malformations).

Most venous malformations can be diagnosed by a careful history and physical examination. Symptoms vary depending on the size and the localization of the lesion. Patients often note firmness and discomfort in the early morning, presumably as a result of stasis and microthrombosis within the lesion. Plain films demonstrate characteristic phleboliths and distortion of nearby bony structures.

Ultrasonography is a simple noninvasive method to distinguish a slow-flow vascular malformation from fast-flow malformation. CT is used primarily to confirm calcifications and to define skeletal changes. MRI is considered the best way to document the rheologic characteristics and anatomic extent of a venous malformation. MRI also helps to differentiate low-flow lesions from high-flow lesions. Unfortunately, we were unable to perform CT and MRI scans in our case 24 years ago. Plain x-ray films (the only radiographic images that we have) showed phlebitis in the mass. Our initial diagnosis was cavernous hemangioma. However, after a new classification proposed by Mulliken in 1982, we reassessed the initial diagnosis. After reevaluation of the history, physical examination, and x-ray findings, we considered the diagnosis to be low-flow (venous) vascular malformation instead of cavernous hemangioma.

The treatment options for venous malformations vary by the size and location of the lesion. The surgical aim is total removal of the lesion, but this cannot be done in every case because of anatomic and functional limitations. Surgical removal of lesions that especially occupy the head and neck may lead to devastating aesthetic and functional consequences.

A variety of techniques have been used over the years in the treatment of venous malformations, specifically, surgical excision, intralesional sclerotherapy, magnesium or copper implantation, radiotherapy, electrocoagulation, cryotherapy, lasers, compression, and steroids. Small asymptomatic lesions can be managed conservatively. The patient should be counseled to avoid trauma and to anticipate possible changes in the lesion at puberty or pregnancy. Small venous malformations can be completely excised or directly injected with sclerosant without the need for radiographic studies. Small superficial venous malformations can be easily treated with a neodymium–yttrium aluminum garnet (Nd:YAG) laser. Large symptomatic or deforming craniofacial venous malformations require sclerotherapy and, in many cases, surgical resection 3 to 4 weeks later, before recanalization occurs.

Many sclerosants have been tried in the past, including boiling water, sodium salicylate, sodium
morrhuate, quinine, silver nitrate, iron, or zinc chloride. Concentrated (95% to 100%) ethanol, sodium tetradeyl sulfate, and polidocanol are in current use. Absolute ethanol is the most destructive sclerosant, and, therefore, it is assumed that it has the lowest recurrence rate. Alcohol injection produces severe pain; thus, general anesthesia is required. Moreover, transient or even permanent neuropathy is a possible complication because of the agent’s neurolytic effect. Hemoglobinuria can be seen secondary to intralesional hemolysis.

The most common complication after sclerotherapy is cutaneous or mucosal blistering. Full-thickness cutaneous necrosis is seen in only a small group of patients. These ulcers are managed conservatively with dressing and antibiotic ointment. Thrombophlebitis and thromboembolism are other postsclerotherapeutic complications. Allergic reactions also have been reported after sclerotherapy. Hence, careful monitoring is mandatory during sclerotherapy, especially with ethanol. Other, less destructive sclerosant agents should be used for superficial venous malformations and venous malformations in close proximity to major nerves to minimize the risk of ulceration and neuropathy.

Sodium morrhuate, also known as detergent sclerosant, is a fatty acid obtained from the liver of codfish. It damages the endothelial cell membrane and eventually leads to endosclerosis. Sodium morrhuate has been successfully used as a sclerosing agent for the treatment of venous malformations. Zhao et al. recently reported that obvious swelling occurred in the local region after intralesional injection of sodium morrhuate, causing dysfunction of the injected sites and in some cases ulceration and tissue necrosis in the injected areas. We used 5% benzyl alcohol solution of sodium morrhuate in our case. We did not see any major complications except a superficial tissue loss that developed on the patient’s temporal region and that recovered in 3 weeks with conservative treatment. Local edema of the injected site was the main problem for the patient and resolved spontaneously in 10 days.

It should be kept in mind that injection of sodium morrhuate to the intraoral or pharyngeal venous malformation and large venous malformation of the neck may result in serious airway obstruction. Tracheotomy or prolonged postinjection intubation may be indicated in these patients. Although large edema and ecchymosis were seen, we did not encounter airway obstruction in our case.

CONCLUSION
Despite having various complications, sclerotherapy is a common method of treatment in low-flow (venous) malformations. Although we achieved satisfactory results from the treatment of this patient with intralesional injection of sodium morrhuate (Varicocid), it is difficult to conclude that this type of treatment is the best choice in all cases of venous malformation on the basis of a report of the single case. However, intralesional sclerotherapy should be kept in mind as a savior treatment option in cases of large venous malformation occupying an anatomic region that is challenging to both the surgeon and the patient. Intralesional sclerotherapy can be performed without serious complications if the sclerosing agent is selected and injected cautiously.

REFERENCES