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Vascular Malformations of the Head and Neck in Children

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Abstract. *Background/Aim: Vascular malformations are congenital abnormalities that result from disturbances in the embryologic development of the vascular system. A retrospective study at a single institution was performed to determine the localization and treatment patterns for vascular malformations in children. Patients and Methods: A total of 198 pediatric patients were identified. Age at diagnosis and presentation, sex, localization, diagnostics, and therapy were described. Results: The most common diagnosis was lymphatic malformation (LM, 58.6%), followed by venous (VM, 31.8%) and arteriovenous malformation (AVM, 4.5%). The mean age at diagnosis was 2.2 years, while the mean age at presentation at our hospital was 7.2 years. The sex ratio showed a female predominance (1.44:1), which was most evident in children with AVM. The neck, cheek/parotid gland and oral cavity were the most predominant locations. Half of the patients required at least one intervention at our hospital. Especially, CM and LM were managed by watch-and wait, whereas lymphovenous malformation (LVM) and AVM were most often treated. Treatment differed between the various malformation types, the most common used treatment was conventional surgery followed by laser therapy. In case of treatment, the average number of procedures in our hospital was 1.58 for VM, 1.53 for LM, 1.33 for AVM, and 1.0 for LVM. Conclusion: In children with vascular malformations interventional treatment is often necessary, in many cases more than one treatment step is needed. Correct identification of the malformation type is important for optimal treatment and appropriate care of patients with vascular malformations.*

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Vascular anomalies are a heterogenous group of diseases ranging from self-limited, isolated, innocuous lesions to lifelong, disfiguring, and even life-threatening disease. They are classified by the International Society for the Study of Vascular Anomalies (ISSVA) based on physical findings, clinical presentation, and biologic characteristics into vascular tumors, which are caused by abnormal endothelial cell proliferation, and non-neoplastic vascular malformations (1). The precise pathogenesis of vascular malformations is still unknown and misdiagnosis of patients with vascular malformations is common (2-4). An exact terminology is important for differentiating vascular malformations from vascular tumors like hemangiomas to prevent ineffective or even adverse therapy. Vascular malformations are typically present at birth and grow proportionally to the patient, although they may not become clinically evident until later in life. In contrast to hemangiomas, they do not regress spontaneously but rather tend to progress over the patient's lifetime (5). Vascular malformations are classified based on their vascular composition (arterial, venous, lymphatic, or combined) and flow dynamics (high or low flow) and include capillary malformation (CM), venous malformation (VM), lymphatic malformation (LM), arteriovenous malformation (AVM), and their combinations of which lymphovenous malformation (LVM) is the most common (1). The symptoms are related to localization and size. Low flow malformations present more frequently in smaller children than high flow lesions, the latter often not becoming symptomatic until teenage years. In the head and neck region, vascular malformations often involve multiple contiguous anatomic spaces and encase critical neurovascular structures, which makes therapeutic interventions rather difficult. In cases where surgical intervention is impossible due to infiltrative growth into encompassing structures, depending on the type of malformation sclerotherapy or

Table I. *Number, sex, mean age at diagnosis and presentation of the patients with vascular malformations.*

	Number of patients (%)	Male (%)	Female (%)	m:f ratio	Mean age at diagnosis	Mean age at presentation
LM	116 (58.6%)	55 (47.4%)	61 (52.6%)	1: 1.11	2.6 years	6.2 years
VM	63 (31.8%)	20 (38.7%)	43 (68.3%)	1: 2.15	0.9 years	9.0 years
AVM	9 (4.5%)	2 (22.2%)	7 (77.8%)	1: 3.5	6.3 years	9.9 years
CM	5 (2.5%)	2 (40.0%)	3 (60.0%)	1: 1.5	2.0 years	2.2 years
LVM	5 (2.5%)	2 (40.0%)	3 (60.0%)	1: 1.5	0.8 years	9.0 years

LM: Lymphatic malformation; VM: venous malformation; AVM: arteriovenous malformation; CM: capillary malformation; LVM: lymphovenous malformation; m: male; f: female.

Table II. *Distribution of the localization of the vascular malformations in the head and neck area (in some patients more than one area was affected).*

	All vascular malformations n (%)	LM n (% of LM)	VM n (% of VM)	AVM n (% of AVM)	CM n (% of CM)	LVM n (% of LM)
Localization						
Neck	91 (46.0%)	85 (37.9%)	5 (7.9%)	-	-	1 (20%)
Cheek/parotid region	45 (22.7%)	25 (21.6%)	14 (22.2%)	2 (22.2%)	1 (20%)	3 (60%)
Tongue/oral cavity	43 (21.7%)	18 (15.5%)	22 (34.9%)	1 (11.1%)	-	2 (40%)
Perioral/lips	10 (5.1%)	3 (2.6%)	5 (7.9%)	2 (22.2%)	-	-
Orbital region	9 (4.5%)	7 (6.1%)	1 (1.6%)	-	1 (20%)	-
Occipital/temporal	6 (3.0%)	3 (2.6%)	-	2 (22.2%)	1 (20%)	-
Paranasal	3 (1.5%)	1 (0.9%)	2 (3.2%)	-	-	-
Chin	3 (1.5%)	2 (1.7%)	1 (1.6%)	-	-	-
Frontal	2 (1.0%)	-	1 (1.6%)	-	1 (20%)	-
Face	1 (0.5%)	-	-	-	1 (20%)	-
Ear canal	1 (0.5%)	-	-	1 (11.1%)	-	-
Missing information	11 (5.6%)	7 (6.0%)	-	4 (44.4%)	-	-

LM: Lymphatic malformation; VM: venous malformation; AVM: arteriovenous malformation; CM: capillary malformation; LVM: lymphovenous malformation.

embolization are important treatment options (5). This study aimed to describe clinical characteristics, diagnosis, and therapy in a hospital-based cohort of 198 pediatric patients with vascular malformations.

Patients and Methods

All patients with vascular anomalies aged 0-18 years who were admitted to the Department of Otorhinolaryngology over a ten-year-period were enrolled in this retrospective study. All cases were diagnosed based on the ISSVA classification of vascular anomalies (1), according to the clinical appearance, biological behavior of the malformations, and investigations of imaging and pathology in some cases. Specially designed data-collected forms were used to collect the following information: age at diagnosis and presentation, sex, localization, and management.

Results

In total, 198 pediatric patients with vascular malformations were included into this retrospective analysis. The most common diagnosis was LM (58.6%), followed by VM (31.8%) (Table I). The children were aged from birth (0 days) to 18 years, 81 patients (40.1%) were male, and 117 patients (59.1%) were female. The male: female ratio for all vascular malformations was 1:1.44. The female predominance was most evident in AVM patients whereas the male: female ratio was most balanced in LM. The mean age at diagnosis was 2.2 years, while the mean age at presentation at our hospital was 7.2 years. Patients with LVM and VM were the youngest at diagnosis whereas patients with CM were the youngest at presentation (Table I, Figure 1).

Most of the vascular malformations were located in the neck (46.0%), cheek/parotid region (22.7%), and oral cavity (21.7%). Different malformation types showed distinctive distribution patterns as shown in Table II. In 28.7% of the patients imaging was performed at our hospital, mostly magnetic resonance imaging (n=61) and sonography (n=13).

Overall, 26.3% of the patients were pretreated at other hospitals before (Table III). A total of 99 patients (50%)

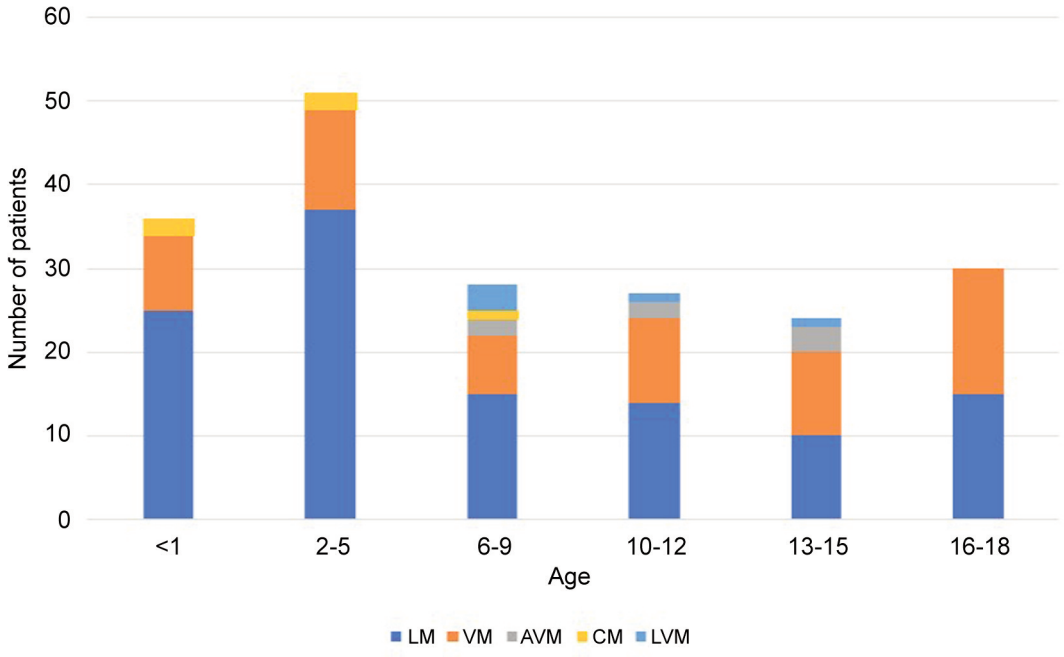

Figure 1. *Age at presentation*.

Table III. *Therapy of patients with vascular malformations*.

	Previous therapy n (%)	Invasive therapy at our institution n (%)	Number of therapeutical procedures (n)				
			Conventional surgery	Laser	Sclerotherapy	Embolization	Other
LM	33 (28.4%)	51 (44%)	33	24	10	-	13
VM	14 (22.2%)	38 (50.7%)	14	12	10	-	5
AVM	1 (11.1%)	6 (66.7%)	5	2	-	3	-
CM	0	-	-	-	-	-	-
LVM	4 (80%)	4 (80%)	3	-	-	-	1

LM: Lymphatic malformation; VM: venous malformation; AVM: arteriovenous malformation; CM: capillary malformation; LVM: lymphovenous malformation.

required at least one intervention at our hospital, whereas the other lesions were managed conservatively (Table III). CM and LM were managed by watch-and wait, whereas LVM and AVM were most often treated. Treatment differed between the various malformation types, the most common used treatment was conventional surgery (Table III). In case of treatment, the average number of treatment sessions in our hospital was 1.58 (range=1-5) for VM, 1.53 (range=1-6) for LM, 1.33 (range=1-2) for AVM, and 1.0 for LVM. Long-term outcome often was not documented as the hospital is a center for patients with vascular anomalies, which often come from distant regions.

Discussion

Vascular malformations are a group of diseases that are often associated with lifelong morbidity (6). Although VM are generally more common, in our cohort most of the patients had LM, this may be due to selection of our hospital by the parents. The male: female ratio in the whole cohort was 1:1.44. In all groups a female predominance could be detected. In a review on vascular malformations, a male: female ratio for VM of 1:1.2, for AVM of 1:4, and for LM of 1:1 was reported (7). In the present series, the female predominance was most evident in children with AVM,

however, the patient group with AVMs was rather small. In children with VM there was also a clear female predominance (1:2.15), whereas the sex ratio was most balanced in LM (1:1.11). The reason for the higher prevalence in females is still not understood. The age at diagnosis was the highest in patients with AVM (6.3 years). One reason is that AVM is commonly misinterpreted as hemangioma in small children. In many cases, only the absence of involution of suspected hemangioma or progression of disease leads to the correct diagnosis of AVM. However, proper identification of the type of malformation is paramount for adequate management. The age at presentation at our hospital also was rather high, especially in children with AVM and VM. This is the cause of a high number (26%) of pretreated cases, which often are more complicated to treat due to altered microvasculature of the malformation and the presence of scar tissue.

Doppler ultrasound, computed tomography, and magnetic resonance imaging (MRI) may be helpful in diagnosing vascular malformations; however, in most of the cases the diagnosis is made by clinical characteristics (5). In our cohort, the rate of imaging at our hospital was low, in less than one third of the patients imaging was performed. However, some patents presented with imaging that was already previously performed.

Treatment of vascular malformations is complex and until now, there are no clearly defined treatment guidelines. In many patients the watch-and-wait policy is used, and the challenge is knowing when to intervene. Interventions are typically required to control lesion size, deterioration in function, pain, bleeding, and nerve impairment due to compression (8, 9). Some vascular malformations can also present with challenging hematologic aberrations (10). The treatment and prognosis are primarily dependent upon the velocity of flow within the lesion. Low-flow malformations are often faring far better than high-flow lesions. Surgical management involves excision, laser treatment or both (11). Sclerotherapy and embolization as well as drugs like sirolimus are non-surgical options (12-14). In this series, the most common used options were conventional surgery, laser, and sclerotherapy; however, many patients required multiple treatment sessions and modalities especially patients with VM and LM. In these cases, a complete resection in the head and neck area is often impossible without compromising functional important structures or aesthetics. Therefore, an individualized, multidisciplinary approach to treatment is required.

Conclusion

The diagnosis and management of vascular malformations remains challenging due to their low prevalence and the various clinical presentations and courses of the disease. In children with lymphatic malformations interventional treatment is often necessary, in many cases more than one treatment step is needed. Correct identification of the malformation type is important for optimal treatment and appropriate care of patients with vascular malformations. In order to gain further insight into vascular malformations in children while providing safe clinical care, they should be treated and carefully monitored through prospective clinical trials.

Conflicts of Interest

The Authors declare no conflicts of interest in relation to this study.

Authors' Contributions

SW designed and coordinated the study. LP collected and analyzed the data. LP and SW interpreted the data. LP and SW wrote the first version of the manuscript. JAW reviewed the manuscript. All Authors approved the manuscript.

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