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Invited Article

Interdisciplinary Management of Head and Neck Vascular Anomalies: Clinical Presentation, Diagnostic Findings and Minimalinvasive Therapies

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ABSTRACT

Objectives: Vascular anomalies are included in the 30 000 rare diseases worldwide affecting less than 5/10 000 people. Depending on their morphology and biological properties, they can cause varied disorders with organ involvement. Almost 60% of vascular anomalies have a predilection for the head and neck region in children. Clinical and scientific effort to establish interdisciplinary management concepts for vascular anomalies is increasing worldwide.

Methods: Especially in the head and neck region, clinical impairment and organ dysfunction is associated with cosmetic issues that may represent a physical and psychological issue for the patient. Correct diagnosis, based on clinical presentation and symptoms, is a prerequisite for appropriate therapy, ranging from conservative management to a spectrum of minimally invasive treatment options. We searched PubMed for German and English language published data until December 2016 with focus on clinical studies, review articles and case reports on vascular anomalies with a focus on the head and neck region.

Results: The last ISSVA update in 2014 has contributed to a better understanding of vascular anomalies, classifying them in vascular tumors and vascular malformations. The predominant representatives of vascular tumors are congenital and infantile hemangiomas. Infantile hemangiomas have the ability of spontaneous regression in more than 80%. Patients with symptomatic growing hemangiomas with ulcerations, bleeding complications and restriction of hearing, swallowing disorder, impairment of vision, or cosmetic dysfigurement require treatment. Therapies include oral propanolol, transcatheter embolization and surgery. Vascular malformations tend to progress with patientś age and are subdivided in slow flow and fast flow lesions. Symptomatic slow flow lesions, e.g. venous and lymphatic malformations, benefit from percutaneous sclerotherapy. Fast flow lesions, as arteriovenous malformations, are rare but undoubtedly therapeutically the most challenging vascular anomaly. Depending on location and size, they may require multiple transcatheter embolization procedures for successful occlusion of the AVM.

Conclusions: This review provides knowledge on the current ISSVA classification of vascular anomalies, their clinical presentation, diagnostic evaluation and minimally invasive therapy options to encourage the establishment of a comprehensive interdisciplinary management for head and neck vascular anomalies.

1. Introduction

Vascular anomalies comprise of congenital vessel disorders which can be associated with soft tissue and organ involvement. The main representatives are vascular tumors and malformations which can already be clinically apparent in the newborn and might be associated with considerable symptoms [1]. The exact incidence of vascular anomalies, a rare disease that mainly affects children and young adults worldwide, is unknown [2]. In approximately 5 cases per 10 000 individuals a vascular anomaly requiring treatment is diagnosed.

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Almost 60% of vascular anomalies in young patients have a predilection for the head and neck region due to unkown reasons [3–5]. Patients can suffer from ulcerations and bleeding complications combined with restriction of hearing, swallowing disorder, and impairment of vision that require treatment. Associated cosmetic disfigurement in the head and neck region has to be addressed, too. Treatment includes pharmacotherapy with oral propanolol, minimalinvasive percutaneous sclerotherapy and transcatheter embolization, and rarely surgery.

A prerequisite for appropriate therapy, which usually consists of an interdisciplinary multimodal approach, is correct diagnosis of the underlying vascular anomaly. In 1982 a fundamental classification system for vascular anomalies was established by Mulliken and Glowacki. In 1992 it was modified by the International Society for the Study of Vascular Anomalies (ISSVA) [6–8]. The updated and currently available ISSVA classification is well accepted internationally and offers clinicians and researchers an important guideline for diagnosis and appropriate therapy of vascular anomalies avoiding incorrect and often confusing nomenclature [9].

The aim of this review is to describe a comprehensive interdisciplinary management approach for head and neck vascular anomalies, based on the current ISSVA classification of vascular anomalies, their clinical presentation in the head and neck region, radiological diagnostic evaluation tools and treatment options.

2. Materials and Methods

We searched PubMed for German and English language published data until December 2016 with focus on clinical studies, review articles and case reports on vascular anomalies with a focus on the head and neck region. Diagnosis was based on the current ISSVA classification of vascular anomalies.

3. Results

3.1. Pathophysiology and Clinical Presentation

Vascular anomalies are subdivided in vascular tumors and malformations. They can be distinguished by their pathophysiology and morphology (Table 1) [10]. The predominant representative of vascular tumors, characterized by excessive angiogenesis, based on endothelial cell proliferation, are congenital and infantile hemangiomas [11]. They frequently occur in the head and neck region (65%), followed by chest and trunk (25%) and upper or lower extremities (10%) [12]. Congenital hemangiomas have attained their full size at birth and may show fast or no regression. Infantile hemangiomas arise weeks to months after birth and are the most common benign tumors of the infant, showing spontaneous regression in more than 80% and often do not need therapy [13,14]. They express the immunhistochemical marker Glut-1. Only enlarging symptomatic hemangiomas require treatment, especially when neighboring organs, like the aerodigestive tract, hearing and vision are impaired [11–13].

Vascular malformations are characterized by defective vessel-maturation with a varying degree of mesenchymal tissue proliferation, including dermal, subcutaneous, fatty and bone tissue [15]. Depending on the vessels involved and flow characteristics, they are divided in venous (VM 70%), lymphatic (LM 15%), arterio-venous (AVM 6%) and capillary (CM 9%) malformations with slow-flow (VM, LM, CM) or fast-flow properties (AVM) [16–18]. Vascular anomalies progress with patient's age, never regress and require treatment when symptomatic.

The ISSVA update of vascular anomalies in 2014 represents a comprehensive and widely acknowledged classification [9,17,18]. It is an integral part of the clinical work-flow in vascular anomaly centers and allows to diagnose and treat patients based on symptoms, clinical findings and associated syndromes (Table 1) [17,19].

The clinical presentation of head and neck vascular anomalies is versatile. Initially superficial hemangiomas may appear as raspberry colored birthmarks or an enlarging reddish discoloration of the skin. Enlarging hemangiomas with organ involvement can cause ulcerations, bleeding, impairment of hearing or vision, chewing or swallowing disorders and airway obstruction (Fig. 1A) [20,21].

Venous malformations, the most common vascular malformation, can enlarge extensively, become a palpable discolored mass with local blood stasis and cause painful thrombophlebitis (Fig. 1B) [22].

Macrocystic lymphatic malformations (cysts > 1 cm) are diagnosed in the head and neck region in almost 60%, followed by chest wall, axilla and extremities in 30% (combined macro- and microcystic) and visceral involvement in 10% (often microcystic) [23]. They are mostly located superficially and may cause pain after local hemorrhage or infection. Swelling in the head and neck region can be associated with chewing disability, dysphagia and obstructive sleep apnea. Currently treatment with Sirolimus in diffuse lymphatic malformations in neonates and children is being evaluated with good clinical response and

Table 1
Compendium of the ISSVA Classification of Vascular Anomalies.

Vascular Tumors			Vascular Malformations		
Benign Tumors	Borderline Tumors	Malignant Tumors	Simple	Combined	Associated with Other Anomalies
- Infantile Hemangioma - Endothelial Cell Proliferation - GLUT-1 Marker positive	Hemangio-endothelioma	Angio-sarcoma	Venous Malformation (VM) Blue rubber bleb nevus Syndrome Glomovenous malformation	CM + VM CM + LM CM + AVM LM + VM	Klippel-Trénaunay-Syndrome
Congenital Hemangioma Excessive Angiogenesis with Capillary Lobules GLUT-1 Marker negative Fully developed at birth	Others	Others	Lymphatic Malformation (LM) Macrocystic Microcystic Mixed Cystic	CM + LM + VM CM + LM + AVM CM + VM + AVM	CLOVES Syndrome
Tufted Angioma			Capillary Malformation (CM) Teleangiectasia Nevus Simplex Others	CM + LM + VM + AVM	Sturge-Weber-Syndrome
Spindle Cell Hemangioma			- Arterio-Venous Malformation (AVM) -		Parkes-Weber-Syndrome
Epitheloid Cell Hemangioma Others			Arterio-Venous Fistula (AVF) Hereditary Hemorrhagic Teleangiectasia (HHT)		Others

Abbreviations: CM, capillary malformation; VM, venous malformation; LM, lymphatic malformation; AVM, arterio-venous malformation.