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Article

VASCULAR MALFORMATION CASES PROFILE IN RSUP DR. KARIADI CENTRAL-GENERAL HOSPITAL SEMARANG BETWEEN 2020-2023

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ABSTRACT

Background: Vascular malformations are abnormalities of the vascular system that arise from improper development during embryogenesis. These vascular anomalies are present at birth and grow proportionally to the size of the child. They have complex clinical presentations and are often misdiagnosed. There are few reports on epidemiology data of these anomalies, and this paper aim to contribute on vascular malformations descriptive epidemiological data.
Method : Patients with vascular malformations who were registered in RSUP Dr. Kariadi Central-General Hospital between January 2020 and December 2023 were included in this paper. All data were collected retrospectively from medical records.
Results : The gender distribution is dominated by male then followed female with the frequency of 51.2% and 48.8% subsequently. Age 1-20 years was the most dominant age range, contributing to 63.2% of the cases. The most commonly found type of vascular malformations are low-flow malformations with venous malformations (69.6%) being the most common and followed by lymphatic malformations (16%). Surgical treatment was done on 52% of the cases, followed by non-surgical (39.2%) and combination of both surgical and non-surgical methods (8.8%). The most common anatomical site was the craniofacial area (56.8%) followed by the limbs area (28.8%).
Conclusion: Both diagnostic and therapy remain a challenge on vascular malformations. By fully understanding about these vascular anomalies, the proper diagnostic can be made hence the proper treatment.

Keywords: *Vascular malformation; Profile; Epidemiology*

Latar Belakang: Malformasi vaskular merupakan kelainan sistem vaskular yang timbul akibat perkembangan yang tidak sempurna selama embriogenesis. Anomali vaskular ini sudah ada sejak lahir dan tumbuh proporsional dengan pertumbuhan anak. Gambaran klinisnya kompleks dan seringkali salah didiagnosis. Laporan mengenai data epidemiologi dari anomali ini masih terbatas, sehingga penelitian ini bertujuan memberikan kontribusi pada data epidemiologi deskriptif malformasi vaskular.
Metode: Pasien dengan malformasi vaskular yang terdaftar di RSUP Dr. Kariadi Semarang pada periode Januari 2020 hingga Desember 2023 diikutsertakan dalam penelitian ini. Seluruh data dikumpulkan secara retrospektif melalui rekam medis.
Hasil: Distribusi jenis kelamin didominasi oleh laki-laki (51,2%) kemudian perempuan (48,8%). Rentang usia 1–20 tahun merupakan kelompok terbanyak, mencakup 63,2% kasus. Jenis malformasi vaskular yang paling sering ditemukan adalah *low-flow malformations*, dengan malformasi vena sebagai tipe terbanyak (69,6%), diikuti malformasi limfatik (16%). Penatalaksanaan bedah dilakukan pada 52% kasus, penatalaksanaan non-bedah pada 39,2%, serta kombinasi keduanya pada 8,8% kasus. Lokasi anatomi tersering adalah regio kraniofasial (56,8%), diikuti ekstremitas (28,8%).
Kesimpulan: Diagnosis dan terapi malformasi vaskular masih menjadi tantangan. Dengan pemahaman yang lebih komprehensif mengenai anomali vaskular ini, penegakan diagnosis yang tepat dapat dilakukan sehingga memungkinkan pemberian terapi yang sesuai.

Kata Kunci: *Malformasi vaskular; Profil; Epidemiologi*

Conflicts of Interest Statement:
The author(s) listed in this manuscript declare the absence of any conflict of interest on the subject matter or materials discussed.

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INTRODUCTION

Vascular malformations are abnormalities of the vascular system that arise from improper development during embryogenesis. Although these anomalies are congenital, they often remain unnoticed and may only become apparent later in life as they grow in proportion to the individual.[1] Vascular development occurs in two phases: first, during vasculogenesis, where blood vessels originate from embryonic cells, and second, during angiogenesis, when the primitive vascular system grows and matures into arteries and veins. Any disruptions during early angiogenesis, typically between 4 and 10 weeks of gestation, can affect vascular development and lead to vascular malformations.[2]

Mutations in two key pathways; the RAS-MAPK and PI3K/AKT/mTOR pathways, result in upregulated signalling and increased angiogenesis. MAP2K1 mutations in the RAS-MAPK pathway are linked to extracranial arteriovenous malformations and fistulas. Mutations in the RASA1 gene (also part of the RAS-MAPK pathway) are associated with combined capillary-arteriovenous malformations and Parkes-Weber syndrome, which is characterized by limb overgrowth alongside capillary and arteriovenous malformations. Mutations in TIE2/TEK (part of the PI3K/AKT/mTOR pathway) have been identified in venous malformations and blue rubber bleb nevus syndrome. Additionally, PIK3CA gene mutations (in the PI3K/AKT/mTOR pathway) are associated with lymphatic malformations, venous malformations, and PIK3CA-related overgrowth syndromes such as CLOVES and Klippel-Trenaunay.[1]

Vascular malformations are categorized into slow-flow and fast-flow lesions based on their hemodynamic characteristics and the types of vessels involved. Slow-flow lesions include veins, lymphatics, capillaries, or a combination of these. In contrast, arteriovenous malformations are classified as fast-flow lesions due to the presence of an arterial component.[3]

Venous Malformations

Venous malformations (VM) are the most prevalent type of vascular malformations and can develop anywhere in the body, including in visceral areas.[4] Clinically, they present as soft, compressible masses featuring bluish membranes or mucous surfaces, lacking any thrill or pulsation.[3] VMs consist of veins and venules of varying sizes, lined by a single layer of endothelial cells. They typically lack adequate valves, leading to blood flow stasis and inflammation, which can result in the formation of phleboliths. These phleboliths occur in nearly 50% of VM cases and are considered pathognomonic, serving as a key diagnostic feature in imaging studies for VMs.[1,2]

The primary symptom of VMs is pain, which can arise from various factors, including local compression, congestion, thrombosis, or even bleeding into nearby structures.[4] Diagnosis primarily relies on clinical history and physical examination, but diagnostic imaging can help confirm the diagnosis. Ultrasound and color Doppler are first-line tools for this purpose due to their non-invasive nature and availability. VMs typically present as low-flow lesions, often appearing hypoechoic and with phleboliths observed in up to 16% of cases.[1,4]

Treatment indications for VMs are similar to those for other vascular malformations and include functional or aesthetic limitations, as well as any impact on the patient's quality of life.[5] In many cases, VMs can be managed conservatively with compression therapy to address local swelling and pain, along with aspirin for thrombosis prevention. However, VMs in the cervicofacial area can have a significant aesthetic impact and may sometimes necessitate active treatment.[3] Conservative approach includes the use of compression garments, anti-inflammatory medications, and proper wound care. While surgery is seldom the first-line treatment, it may be considered in specific cases, such as ligating efferent veins to enhance sclerotherapy outcomes, removing residual VMs after sclerotherapy, excising sclerotherapy-resistant lesions, or addressing localized lesions that can be completely removed.[1]

Sclerotherapy and embolization are alternative treatment options that involve injecting chemical agents into the vascular malformation to thrombose and obliterate the vessels, effectively inactivating the malformation. These minimally invasive treatments have become the preferred choice. The two techniques differ in their mechanisms: embolizing agents block the vessels, while sclerosing agents damage

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the vascular endothelium, leading to inflammation, clotting, and fibrosis. Sclerotherapy is commonly seen as the primary treatment option. While absolute ethanol is effective for large, extensive VMs, it should be used carefully because of the risks of nerve damage, skin necrosis, and systemic toxicity. Other sclerosants that may be employed include 3% sodium tetradecyl sulfate (STS) and bleomycin.[1]

Lymphatic Malformations

Lymphatic malformations (LM) are characterized by vascular channels or vesicles filled with lymphatic fluid, lined by a single layer of endothelial cells, leading to abnormal lymphatic flow. About 75% of LM cases are found in the cervicofacial region, while the remaining 25% occur in the trunk and extremities. Clinically, these lesions appear as soft, compressible masses with a reddish-brown tint to the skin. They are usually asymptomatic but can cause significant mass effects. LMs are further categorized into three types based on the size of the lymphatic chambers: macrocystic (>2 cm), microcystic (<2 cm), and mixed.[1]

Macrocystic and microcystic lymphatic malformations (LM) can be distinguished by their histological characteristics. Macrocystic LM features large interconnected lymphatic cysts, whereas microcystic LM is made up of smaller lymphatic channels that may interdigitate with surrounding tissue elements. Mixed-type LM contains both macrocystic and microcystic components histologically.[4] Lymphatic malformations (LMs) can develop in any area of the body, but they are most commonly seen in the head and neck, axilla, mediastinum, retroperitoneum, and perineal region. Most cases of LM can be diagnosed based on clinical history and presentation. On colour Doppler, shows absence of flow within the cystic structures, although the septa often show signs of vascularity.[6] Surgery is primarily designated for microcystic forms, while sclerotherapy is usually preferred for macrocystic lymphatic malformations. Ethanol or bleomycin are employed as embolizing agents, especially in the head and neck area.[3]

Capillary Malformations

Capillary malformations (CM) impact the capillaries located in the papillary or reticular dermis, with the number of abnormal vessels diminishing at greater depths. They were historically known as "port-wine stains" before the term CM became widely used.[4] Clinically, capillary malformations typically present as macular stains that are pink or purple in color. CMs account for only 0.5% of cases of vascular malformations, with the majority located in the ophthalmic and maxillary divisions of the trigeminal nerve distribution in the face.[1] These lesions frequently occur alongside other clinical syndromes and anomalies, such as Sturge-Weber syndrome, Proteus syndrome, encephaloceles, and Cobb syndrome, among others.[4]

Diagnosis of capillary malformations (CMs) relies solely on clinical features, with no need for imaging studies. Ultrasound examination of these lesions typically reveals no abnormalities and can help distinguish them from other vascular malformations. Capillary malformations (CM) are treated with tunable flashlamp pulsed-dye laser therapy. This method works by having hemoglobin absorb the laser light, which is then transformed into heat, causing the coagulation of blood vessels. Through a process called selective photothermolysis, the treatment effectively targets the lesions while protecting adjacent tissues.[4]

Arteriovenous Malformations

Arteriovenous malformations (AVMs) account for 3-20% of all vascular malformations and are considered the most complex type. They are characterized by a 'nidus,' a vessel that establishes an abnormal connection between the arterial and venous systems.[1] Clinically, AVMs may present as soft tissue swellings that are red or bluish in color, slightly compressible, and pulsatile with a palpable thrill. They are usually painless but may be associated with frequent episodes of bleeding.[2]

Ultrasound imaging is the first-line diagnostic tool, providing information about the anatomical extent of vascular malformations and distinguishing between fast-flow and low-flow types. For low-flow vascular malformations like VM and LM, magnetic resonance imaging (MRI) is the most appropriate follow-up. MRI is useful for assessing the extent of the vascular malformation and its relationship with surrounding structures. In the case of fast-flow lesions, digital subtraction angiography (DSA) is the gold standard for confirming arteriovenous shunting, evaluating the angioarchitecture of the nidus, and determining the specific drainage patterns.[2]

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Surgical resection can be considered for symptomatic lesions, but it is especially challenging in the case of AVMs due to their complex network of vascular connections. Treatment for AVMs centers on obliterating the nidus, with sclerotherapy and embolization being the primary options to enable safer intraoperative resection and minimize blood loss.[7] When considering the resection of an AVM, the main objective is disease control, since these lesions are seldom curable. Indications for intervention include ischemic pain, recurrent ulcerations, bleeding, or disrupted cardiac function.[8]

METHOD

Patients with vascular malformations who were registered in RSUP Dr. Kariadi Central-General Hospital between January 2020 and December 2023 were included in this paper. All data were collected retrospectively from medical records. The primary outcomes of this paper were the data of vascular malformations in terms of age, gender, vascular malformations subtype, and the treatment done to the patient. The data is presented in tabular form then processed using SPSS with output results, then analyzed by the author. Because there were no patients' identifications published, ethical clearance and informed consent were not deemed necessary.

RESULTS AND DISCUSSION

Four years data from 2020 to 2023 we collected a total of 125 patient's data, with the most case found in year 2022. The gender distribution is dominated by male then followed female with the frequency of 51.2% and 48.8% subsequently. Age 1-20 years was the most dominant age range, contributing to 63.2% of the cases. The most commonly found type of vascular malformations are low-flow malformations with venous malformation (69.6%) being the most common and followed by lymphatic malformations (16%). Surgical treatment was done on 52% of the cases, followed by non-surgical (39.2%) and combination of both surgical and non-surgical methods (8.8%). Due to the most common anatomical site being in the craniofacial area (56.85) followed by limbs (28.8%), hence surgical approach was chosen. Functional and physiological aspects are taken into consideration with the treatment chosen for the patient.

Table 1. *General features of 125 patients with vascular malformations*

		Frequency (f)	Percentage (%)
Case per year	2020	26	20.28
	2021	23	18.4
	2022	39	31.2
	2023	37	29.6
	Total	125	100
Gender	Male	64	51.2
	Female	61	48.8
	Total	125	100
Age (year)	<1	13	10.4
	1-20	79	63.2
	21-40	28	22.4
	41-60	3	2.4
	>60	2	1.6
	Total	125	100
Types	Low-flow	109	87.2
	Fast-flow	16	12.8
	Total	125	100
Treatment	Surgical	49	39.2
	Non-surgical	65	52
	Surgical and non-surgical	11	8.8
	Total	125	100

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Table 2. *Specified vascular malformation features and the treatment given*

		Frequency (f)	Percentage (&)
Location	Craniofacial	71	56.8
	Trunk	10	8
	Limbs	36	28.8
	Multiple	8	6.4
	Total	125	100
Low-flow	Vein	87	69.6
	Lymphatic	20	16
	Capillary	1	0.8
	Lymphatovenous	1	0.8
Fast-flow	Artery	3	2.4
	Arteriovenous	13	10.4
	Total	125	100
Non-surgical	Sclerosant agent	49	39.2
Surgical	Excision	41	32.3
	Mass reduction	23	19.7
Surgical and non-surgical	Sclerosant and excision / mass reduction	11	8.8
	Total	125	100

CONCLUSION

Both diagnostic and therapy remain a challenge on vascular malformations. By fully understanding about these vascular anomalies, the proper diagnostic can be made hence the proper treatment.

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