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The Profile of Vascular Anomalies in Pediatric Patients at A Tertiary Referral Hospital in West Java, Indonesia

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ABSTRACT

Introduction: Vascular anomalies consist of a wide spectrum of pathologies, which are classified as either vascular tumours or vascular malformations. It is important for the clinician to be able to diagnose, classify, and manage variants of vascular anomalies. Objectives: We aimed to describe the clinical characteristics, further examinations, and treatment recommendations of vascular anomalies in pediatric patients at the Pediatric Dermatology Outpatient Clinic of Dr. Hasan Sadikin General Hospital, Bandung. Methods: We conducted a descriptive retrospective study of all patients with vascular anomalies at the Pediatric Dermatology Outpatient Clinic of Dr. Hasan Sadikin General Hospital between January 1st, 2019–December 31st, 2023. Data was obtained through the patient's medical record, including gender, history of maternal gestational conditions, age of onset, age of diagnosis, clinical symptoms, location of the lesions, type of vascular anomalies, further examinations, and treatment recommendations. Results: Of the 25 patients included, the proportion was quite similar between vascular malformations (52%) and vascular tumours (48%). The skin lesions appeared mostly at birth, 84.6% for vascular malformations and 66.7% for vascular tumours, yet the age of diagnosis was mainly found at 2–10 years (46.2%) and 6–24 months (33.3%), respectively. All patients had been taken for medical advice, but only 8% obtained the correct specific diagnosis from previous medical records. Both vascular malformations (38.4%) and vascular tumours (41.7%) had major anatomical distribution on the head and neck. The most common diagnosis found in this study was capillary malformation (32%), with the primary clinical symptoms being pain and pruritus. Dermoscopic examination was the most frequent further examination performed (60%). Up to 32% of patients received a treatment plan that incorporated multiple modalities. Conclusion: Early identification of the type of vascular anomaly is important to determine the appropriate management and prognosis of the disease.

Keywords: *pediatric, vascular anomaly, vascular malformation, vascular tumour*

1. INTRODUCTION

Vascular anomalies are a diverse range of conditions with distinctive clinical characteristics.1,2 For an extended period of time, the terminology used in describing vascular anomalies has been ambiguous and inconsistent. A wide variety of vascular anomalies have been referred to as "hemangioma" and "angioma", while vascular malformations itself should not be associated with the suffix "oma", since the term implies tumour and cellular proliferation.3 Mulliken and Glowacki's brought a shift in vascular anomalies terminology in 1982,4,5,6 and categorized it according to clinical, hemodynamic, radiologic, and histologic characteristics. Vascular anomalies further classified into two main groups: (1) vascular tumours (characterized by cellular proliferation); and (2) vascular malformations (blood vessel structural anomalies). This classification was adopted and developed by the International Society for the Study of Vascular Anomalies (ISSVA) in 1996, and it recently updated in 2018.4

Despite the fact that the classification has existed since 1982, in 2011, Hassanein et al.,7 found an alarmingly high percentage (71%) of mistaken terminology and incorrect classifications that were used across many disciplines.

Accurate diagnosis of vascular anomalies is difficult to determine because of the wide variety of classification schemes, the ambiguity of terminology, and the lack of agreement throughout different disciplines about proper diagnostic and therapeutic criteria.4

Vascular anomalies are caused by interrupted development at different phases of vasculogenesis, which in most cases affect the skin, though they can affect any organ system.1,4 Whereas vascular tumours characterized by cellular proliferation and hyperplasia, vascular malformations are not true tumours and characteristically exhibit normal endothelial turnover. It is caused by errors in vascular morphogenesis that become evident at birth, and grow proportionally with less endothelial mitotic activity.1 Due to the diversity in nature and type of lesions, treatment strategies vary among the specific types of vascular anomalies.8

In most cases, for pediatric patients, the parents have endured many opinions, multiple imaging modalities, and even tissue collection procedures before an accurate diagnosis established and appropriate management was obtained.2,4

A more comprehensive understanding of clinical presentation, diagnosis, and treatment strategies will help the clinicians to provide better management for the patients. Study related to vascular anomaly in pediatric patients at a tertiary referral hospital in West Java has never been carried out. Therefore, this retrospective study aims to describe the clinical characteristics, further examinations, and treatment recommendations of vascular anomalies in pediatric patients at the Pediatric Dermatology Outpatient Clinic of Dr. Hasan Sadikin General Hospital, Bandung

2. MATERIAL AND METHODS

Study design and participants

This descriptive retrospective study was carried out at the Pediatric Dermatology Outpatient Clinic of Dr. Hasan Sadikin General Hospital, Bandung. This study included 25 patients under the age of 18 years old, presented with vascular anomalies. Patient's data were obtained secondarily both from paper-based medical records and electronic medical records (EMR) of vascular anomalies that were admitted from January 1st, 2019 to December 31st, 2023. The inclusion criteria included medical records from the Pediatric Dermatology Outpatient Clinic of Dr. Hasan Sadikin General Hospital, with diagnosis of vascular anomalies, covering conditions falling under the categories of vascular malformation and vascular tumour (International Classification of Disease, 10th revision). The particular details obtained from each patient's comprehensive medical records included gender, history of maternal gestational conditions including gestational age and birth weight, age of onset, age of diagnosis, clinical symptoms, anatomical distribution of the lesions, type of vascular anomalies, further examinations, and management recommendations Diagnosis Categories

Table 1. ISSVA Classification 2018 of vascular anomalies

<table>
<tr><td rowspan="2">Vascular Tumours</td><td colspan="4">Vascular Malformations</td></tr>
<tr><td>Simple</td><td>Combined</td><td>Of Major Named Vessels</td><td>Associated with Other Anomalies</td></tr>
<tr><td>Benign</td><td>Capillary malformations</td><td rowspan="4">defined as two or more vascular malformations found in one lesion</td><td rowspan="4">abnormalities in the origin/course/number of major blood vessels that have anatomical names</td><td rowspan="4">syndrome in which vascular malformations are complicated by symptoms other than vascular anomalies</td></tr>
<tr><td rowspan="3">Locally aggressive or borderline</td><td>Lymphatic malformations</td></tr>
<tr><td>Venous malformations</td></tr>
<tr><td>Arteriovenous malformations</td></tr>
<tr><td>Malignant</td><td>Arteriovenous fistula</td></tr>
<tr><td></td><td></td><td></td><td></td><td></td></tr>
</table>

Adapted from: ISSVA classification 2018[6]

Patients were grouped using the ISSVA classification 2018. Vascular anomalies are categorized into two main groups based

on the clinical and histopathological examination, including "vascular malformation", in which structural vascular abnormalities appear without endothelial cells (ECs) proliferation, and "vascular tumour" which involves proliferative alteration of ECs. Vascular tumours are further subclassified into benign, locally aggressive or borderline, and malignant type. On the other hand, four categories are used to further define vascular malformation, including simple, combined, malformation of major named vessels, and malformation associated with other anomalies. The ISSVA classification 2018 overview table is presented in **Table 1**.

3. RESULTS

Based on the study results, there were 25 cases of vascular anomalies during the study period, consisting of 13 (52%) cases of vascular malformations and 12 (48%) cases of vascular tumours. There was no predominance sex in vascular malformation group, while female predominance was seen in vascular tumour group. Most patients in the vascular malformation group (76.9%) and the vascular tumour group (66.7%) were born at full term. However, one third of vascular tumour patients (33.3%) were born preterm. The majority of patients (58.3% for both groups) have a birth weight 2500–3000 g, and no patient was born with a low birth weight (<2500 g).

In the vascular malformation group, the majority of patients (84.6%) had the onset of skin lesion at birth, followed by ages 1–6 months and 2–10 years (7.7% for both). However, the age of diagnosis in this group was mainly found at 2–10 years (46.2%), followed by >10 years (30.7%), and 6–24 months (23.1%). Similar findings were also observed in the vascular tumour group, where most cases (66.7%) had skin lesion at birth.

The majority of this group's diagnoses were made between the ages of 6–24 months (33.3%), followed by 1–6 months (25%), 2–10 years (16.7%), >10 years (16.7%), and at the time of birth (8.3%).

Table 2. Characteristics of vascular anomalies patients

	Vascular Malformations **No (%) of patients (N=13)**	**Vascular Tumours** **No (%) of patients (N=12)**
Sex		
Male	6 (46.2)	2 (16.7)
Female	7 (53.8)	10 (83.3)
Gestational age		
Preterm	3 (23.1)	4 (33.3)
Full term	10 (76.9)	8 (66.7)
Post term	-	-
Weight of birth		
<2500 g	-	-
2500-3000 g	7 (53.8)	7 (58.3)
>3000-3500 g	3 (23.1)	1 (8.3)
>3500 g	-	-
Unknown/unspecified	3 (23.1)	4 (33.3)
Age of onset		
Birth	11 (84.6)	8 (66.7)
1-6 months	1 (7.7)	2 (16.7)

6-24 months	-	1 (8.3)
2-10 years	1 (7.7)	1 (8.3)
Unknown/unspecified	-	-
Age of diagnosis		
Birth	-	1 (8.3)
1-6 months	-	3 (25.0)
6-24 months	3 (23.1)	4 (33.3)
2-10 years	6 (46.2)	2 (16.7)
>10 years	4 (30.7)	2 (16.7)

Table 2 shows the characteristics of vascular anomaly patients including sex, gestational age, weight of birth, age of onset, and age of diagnosis. All patients had previously been taken for medical advice. Most of the patients (68%) received an uncertain specific diagnosis. Twelve percent of the patients had a nonspecific vascular anomaly diagnosis, while another 12% had an incorrect specific diagnosis. Only 8% of patients received the correct specific diagnosis out of all cases

Table 3. Previous diagnosis

Diagnosis before clinic visit	**No (%) of patients (N= 25)**
No diagnosis specified	17 (68)
Nonspecific vascular anomaly	3 (12)
Incorrect specific diagnosis	3 (12)
Correct specific diagnosis	2 (8)

The skin lesions of vascular malformations were evenly distributed between the head and neck as well as the lower extremities (38.4%), while lesions of vascular tumours were mainly located on the head and neck (41.7%)

Table 4. Predominant anatomical distribution of the lesion

Predominant location of lesion	**Vascular Malformation %(N=13)**	**Vascular Tumour %(N=12)**
Head and neck	5 (38.4)	5 (41.7)
Upper extremities	1 (7.7)	-
Lower extremities	5 (38.4)	1 (8.3)
Trunk	-	2 (16.7)
Combination of multiple locations	2 (15.4)	4 (33.3)

Table 5. Type of vascular anomalies

Clinical diagnosis determined	No (%) of patients (N= 25)
Infantile hemangioma	7 (28)
Congenital hemangioma	3 (12)
Tufted angioma	1 (4)
Granuloma pyogenic	1 (4)
Verrucous venous malformation	4 (16)
Port wine stain	8 (32)
Sturge-Weber syndrome	1 (4)

The data in **Table 5** showed a list of precise diagnosis for the type of vascular anomalies in our outpatient clinic. Port wine stain (capillary malformation) (32%) was the most frequent primary diagnosis, followed by infantile hemangioma (28%), verrucose venous malformation (16%), and congenital hemangioma (12%). The diagnosis of tufted angioma, pyogenic granuloma, and Sturge-Weber syndrome were within the same proportion of patients (4%).

Figure 1. Vascular malformations. Capillary malformation caused facial disfigurement (A); Asymptomatic capillary malformation (B); Venous malformation on the right knee (C); Venous malformation on the left ankle (D).

Figure 2. Vascular tumours. Superficial infantile hemangioma on the forehead (A); Infantile hemangioma with ulceration (B); Tufted angioma (C,D).

The **Figure 1** showed vascular malformation patients, and **Figure 2** showed vascular tumor patients.

Table 6. Symptoms associated with the vascular lesions

Symptoms Associated*	Vascular Malformations %(N=13)	Vascular Tumours %(N=12)
Pain	30.7	41.6
Pruritus	30.7	41.6
Swelling	-	8.3
Infection	15.3	8.3
Ulceration	-	16.6
Bleeding	30.7	33.3
Disfigurement	23.0	-
Functional compromise	-	-

*Data are percentage of patients complaining each symptom. Multiple symptoms may have been recorded for each patient.

Table 7. Further examinations

Further examination*	No (%) of patients (N=25)
Dermoscopic examination	15 (60)
Histopathologic examination	11 (44)
Immunohistochemistry examination	2 (8)
Ultrasonography	2 (8)
Computed tomography (CT) angiography	4 (16)
Magnetic resonance imaging (MRI)	1 (8)
Unknown/no specific further examination	10 (40)

*Data are percentage of further examination and multiple examination may have been recorded for each patient.

The majority of patients (76%) had clinical symptoms, while the rest of the patients presented with asymptomatic lesions. Pain and pruritus were the primary symptoms found in both groups (**Table 6**). In the vascular malformation group, disfigurement occurred in 23% of patients, and this was not observed in the vascular tumour group. In contrast to the vascular malformation group, swelling and ulceration were only observed in the vascular tumour group. The incidence of bleeding in the vascular tumour group was higher than in the vascular malformation group.

Table 8. Dermoscopic features

Dermoscopic features*	Vascular Malformations %(N=13)	Vascular Tumours %(N=12)
Red to reddish-purple globules, and lacunae	15.3	41.6
Red to reddish-white structureless area	-	16.6
White to bluish-white structureless area	23.0	8.3
Bluish to bluish-red lacunae and structureless area	46.0	8.3
Skin-colored to light pink structureless area	-	8.3
Linear irregular vessels	23.0	41.6
Pink structureless area	7.6	8.3
Red dots	-	8.3
Blood spots	-	8.3
Ulceration	-	8.3

*Data are percentage of each dermoscopic features in patients. Multiple dermoscopic features may have been recorded for each patient.

Dermoscopic examination (60%) was the most frequent further examination performed on the patients in this study population (**Table 7**).

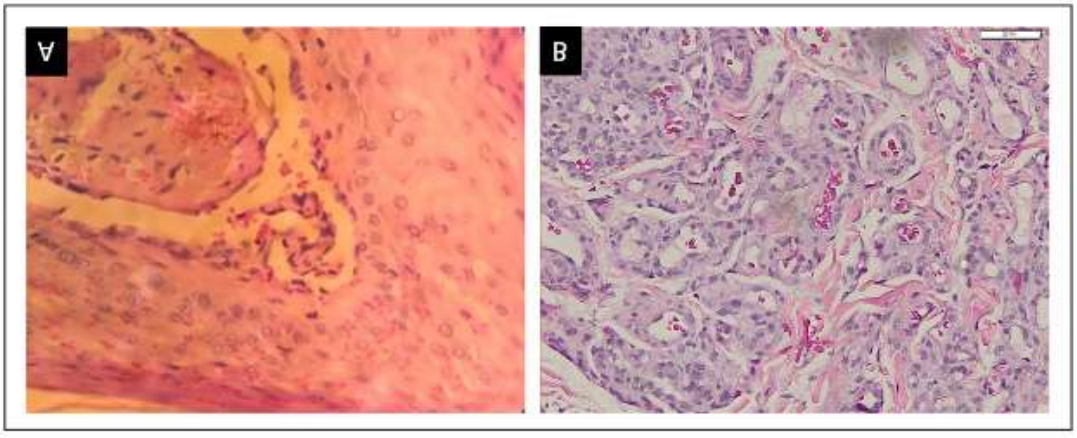

Figure 3. Histopathological examination. Capillary dilatation and lined by endothelial cell in vascular malformation (A, red arrow); Infiltration of lymphocytes (B,yellow arrow), dilated vessels, endothelial cells proliferations forming small capillary vessels in vascular tumour (B, red arrow)

Histopathological examination was carried out in 44% of patients (**Figure 3**), and two of them also had an immunohistochemical examination.

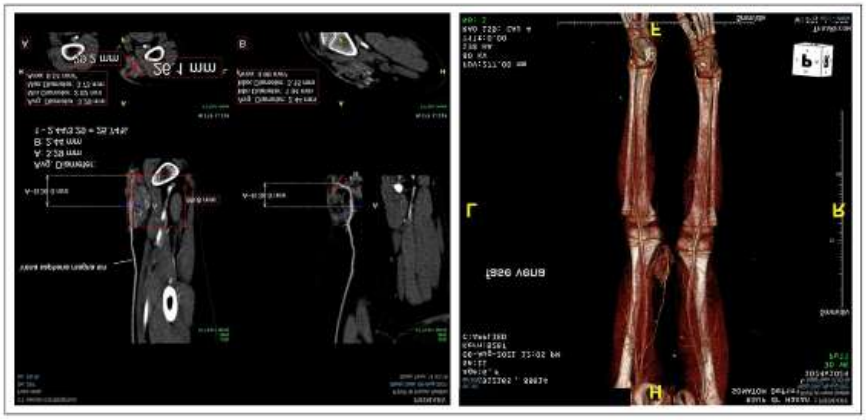

Figure 4. CT Angiography examination. CT angiography revealed the appearance of mild stenosis and arterial collateralization in venous malformation case.

Computed tomography (CT) angiography was performed in 16% of patients (**Figure 4**), ultrasonography (USG) in 8% of patients, and only one patient (8%) underwent a magnetic resonance imaging (MRI) examination. A large number of patients (40%) did not undergo specific further examinations.

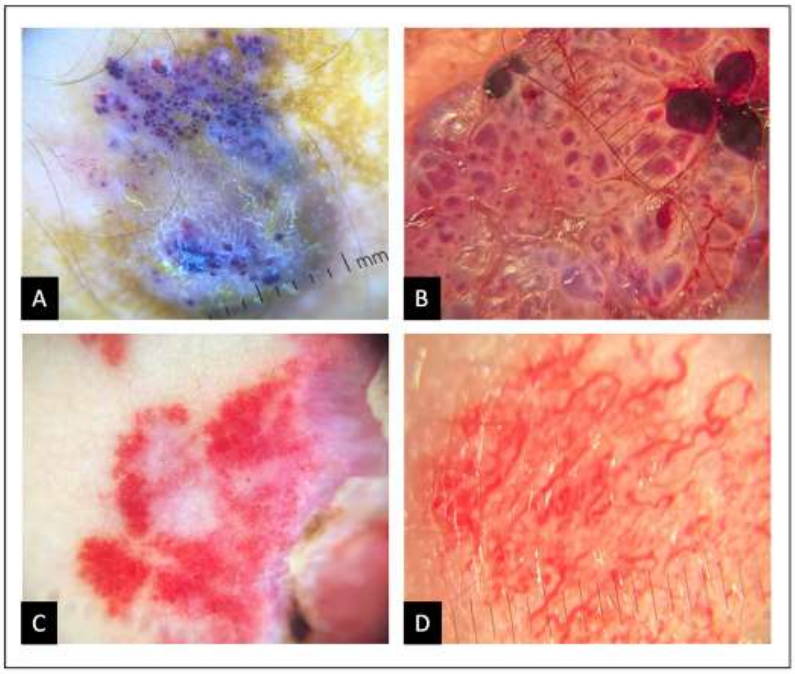

Figure 5. Dermoscopic examination. Vascular malformations: bluish lacune, with dark blue lacune on perifer (A) and red to reddish-purple globules, and lacunae (B). Vascular tumours: red to reddish globules, pink structureless area, red dots (C) and linear irregular vessels (D).

Some dermoscopic findings among the patients was showed in **Figure 5**. Among patients who underwent dermoscopic examination, bluish to bluish-red lacunae (46%) and white structureless areas (23%) were the most common appearances of vascular malformations.

Table 9. Management recommendations

Management	**Vascular Malformations No %(N=13)**	**Vascular Tumours No %(N=12)**
Further diagnostic workup	2 (15.4)	1 (8.3)
Treatment		
Surgery	-	1 (8.3)
Laser	5 (38.5)	2 (16.7)
Cryotherapy	-	1 (8.3)
Topical beta-blocker	-	1 (8.3)
Oral beta-blocker	1 (7.7)	3 (25.0)
Multiple treatment modalities	5 (38.5)	3 (25.0)

The most common features of vascular tumours were red to reddish-purple globules and lacunae as well as linear irregular vessels (41.6% for both), followed by red to reddish-white structureless area (16.6%) (**Table 8**).

Table 10. Specific single management recommendations

Clinical diagnosis determined	Further diagnostic workup No %	Surgery No %	Laser No %	Cryotherapy No %	Topical beta-blocker No %	Oral beta-blocker No %
Infantile hemangioma	1 (4)	1 (4)	1 (4)	-	1 (4)	-
Congenital hemangioma						3 (12)
Tufted angioma	-	-	1 (4)	-	-	-
Granuloma pyogenic				1 (4)		

Verrucous venous malformation	-	-	-	-	-	1 (4)
Port wine stain	2 (8)	-	4 (16)	-	-	-
Sturge-Weber syndrome	-	-	1 (4)	-	-	-

The variation of management recommendations is shown in **Table 9**. In the vascular malformation group, the majority of patients received laser and multiple treatment modalities (38.5%), 15.4% were recommended to have further diagnostic workup, and 7.7% received oral beta-blockers.

Table 11. Specific multiple management recommendations

Clinical diagnosis determined	**Oral and topical beta-blocker No %**	**Surgery and oral beta-blocker No %**	**Laser and oral beta-blocker No %**	**Surgery and cryotherapy No %**
Infantile hemangioma	3 (12)	-	-	-
Congenital hemangioma		-	-	-
Tufted angioma	-	-	-	-
Granuloma pyogenic	-	-	-	-
Verrucous venous malformation	-	1 (4)	1 (4)	1 (4)
Port wine stain	-	1 (4)	-	1 (4)
Sturge-Weber syndrome	-	-	-	-

In the vascular tumour group, as much as 25% of patients were given oral beta-blocker and multiple treatment modalities, followed by laser treatment (16.7%). **Table 10** and **Table 11** showed specific single and multiple management recommendations for each clinical diagnosis determined.

4. DISCUSSION

The term of vascular anomalies encompasses a wide spectrum of pathologies that are best understood when classified as either vascular tumours or vascular malformations.[3,9] The ISSVA 2018 published the guidelines which break down the groups into significantly more detailed categories, although several pathologies have considerable overlapping clinical and imaging characteristics.[3,4] Despite the fact that vascular anomalies are prevalent, the correct diagnosis are often poorly understood.[3]

It appears vascular anomalies were more common in female than male in this study, with a proportion of 68% and 32%, respectively. This is consistent with research findings conducted by Karmacharya et al.[10] which showed vascular anomalies

are twice higher in female. Vascular tumours, particularly hemangioma, have 2–3 fold higher incidence in female than male.[11] The pathogenesis of higher incidence in female remains unclear, but estrogen is thought to be responsible for promotes endothelial cell proliferations.[11] However, sex distribution in the vascular malformation group was equal, in accordance with the literature which stated that there was no sex preponderance in vascular malformations.[5,12]

Most patients (72%) were born at full term, with the birth weight group dominated between 2500–3000 g. No patient was born with a low birth weight in both groups. Gestational age and birth weight can be associated with the occurrence of several vascular anomalies, particularly in infantile hemangioma.[13] A study conducted by Andersen et al.,[13] showed decreased gestational age at birth and decreased birth weight is contributable as risk factors for development of infantile hemangioma. Increasing evidence supports the role of hypoxia in IH development and hypoxia is postulated to trigger neovasculogenesis in infants. Low birth weight, the most significant risk factor for IH, is known to be associated with hypoxia. Very low birth weight (<1500 g), associated with placental insufficiency and hypoxia, was found to be substantially correlated with infantile hemangioma.[14] Some investigators have noted that infantile hemangioma can first appear as an area of vasoconstrictive pallor. Munden et al.,[14] found in his study that the development of infantile hemangioma was significantly associated with a history of placental hypoxia, such as pre-eclampsia and placenta previa.

In both groups, the onset of skin lesions appeared mostly at birth, in accordance with several literatures indicating that vascular anomalies are primarily present at birth.[5,12,15] Vascular malformations can be identified later in life, but they are primarily congenital.[5,12] Although the majority of cases had the onset at birth, the study's findings indicate that the vascular malformation group is mostly diagnosed between the ages of 2–10 years (46.2%), while vascular tumour group is primarily diagnosed between the ages of 6–24 months (33.3%).

All patients had previously been taken for medical advice before visiting our outpatient clinic. Among all patients, only 8% had a specific diagnosis correctly. Most patients (68%) did not obtain a specific diagnosis, and 12% received an incorrect diagnosis. Study conducted by Mathes et al.,[16] on clinical characteristics and management of vascular anomalies showed that only 22% of the study population had a correct diagnosis. In his study, large number of referral letters were received without prior diagnosis, thus these numbers may be low estimates. This implies that a significant percentage of patients with vascular anomalies may not be receiving the ideal management needed or may not even be aware of the fact they have the specific type of vascular anomaly. It is crucial to understand that patients are being sent to the Pediatric Dermatology Clinic due to the difficulty in diagnosing or treating these conditions. Multidisciplinary approaches are particularly required for these difficult situations.[16]

Vascular anomalies are currently considered to be the most complex and confusing vascular disease.[17,18] With the introduction of biological classification, the majority of the misunderstandings related to classification of vascular anomalies have been resolved. It is classified into vascular malformation which caused by localized maldevelopment of vascular morphogenesis, and vascular tumours which associated with hyperplasia that caused by increased endothelial cell proliferation. Whereas vascular tumours may regress as the patient gets older, vascular malformations increase in size and do not fade spontaneously.[18,19] This taxonomy has removed the confusing terminologies, which led to improper diagnosis and treatment earlier.[18,19]

The number of patients of vascular tumours and vascular malformations was quite similar in this study, with the proportion of 48% and 52%, respectively. Based on vascular malformation's flow characteristics, they can be subdivided into slow-flow (capillary, lymphatic, venous, and combined) and fast-flow (arterial. Arteriovenous, and combined).[5,20] In this study, capillary malformation (32%) was the most frequent primary diagnosis, followed by venous malformation and Sturge-Weber syndrome. Vascular malformations affect about 0.3% of the population, and capillary malformations (CMs) account for the majority of them.[5] Capillary malformations, commonly called *port-wine stain* (PWS), is a slow-flow vascular malformation that mainly occur sporadically.[5,21] Typically, PWS are unilateral and segmental, but it may also be bilateral or multisegmental.[1] Approximately 90% of PWS are located on the face, followed by neck, trunk, and extremities.[22] According to studies from several specialized centres, venous malformation is the most common vascular malformation reported, with an overall incidence of 1 in 10.000 in the population.[16] Venous malformation is a congenital lesion made up of venous-type vessels of the skin or mucosa but can involve any structure. The cervicofacial region is affected in 50% cases of venous malformation.[5] This is in accordance with the findings of this study which showed that the head and neck were the most common locations for vascular malformations.

The most common diagnosis in the vascular tumour group was infantile hemangioma, followed by congenital hemangioma, tufted angioma, and granuloma pyogenic. This is in line with numerous published data showing that infantile hemangiomas are the most common benign tumour of childhood, with a reported incidence of 4–5% in children <1 year of age.[6,12,15,23] The skin lesions mostly appeared weeks to months after birth, but 30% to 50% can present with a precursor lesion at birth.[24] Infantile hemangioma can manifest on any part of the body, particularly of the head and neck (60%), trunk (25%), and extremities (15%). Contrary to infantile hemangioma, congenital hemangiomas are fully developed at birth. Congenital hemangioma exhibit proportional growth, are usually solitary, and more commonly present on the extremities.[24] In this study,

the predominant distribution of the vascular tumour group was on the head and neck, lower extremities, or a combination of multiple locations.

The symptoms reported in this study are consistent with other reports. The majority of our patients (76%) have symptoms related to their vascular lesions. Pain and pruritus were the most common symptoms complained about by the patients. These findings were similar to the result of a study conducted by Mathes et al.,[16] that found more than 50% of vascular anomaly patients complained of pain. Some vascular malformations located of the facial area can cause deformities and interfere with normal function, leading to facial disfigurement, as seen in 23% of patients in this study. Ulceration is a common complication of vascular tumours, especially infantile hemangioma. This condition also contributes to significant morbidities, including pain, bleeding, and risk for infection.[25]

In addition to establishing a correct diagnosis, some further examinations were recommended. Dermoscopic examination was the most frequent (60%) further examination performed on the patients in the study population. The heterogeneous clinical morphology of the vascular malformations and tumours poses a significant problem in clinical diagnosis and requires pathological examination to confirm the diagnosis. Dermoscopy is being increasingly used to aid in the diagnosis of various dermatoses by displaying pigmented, non-pigmented, and vascular features. It has significantly increased the prebiopsy diagnostic accuracy of cutaneous lesions by demonstrating correlates of the lesions' pathological features.[26,28] Clinical approach supported by dermascopic examinations can enhance the diagnosis establishment in patients with vascular anomaly, compared to clinical assessment alone (risk ratio: 1.36; 95% confidence interval: 1.10-1.67).[26]

In this study, the appearance of bluish to bluish-red lacunae and structureless areas were the most common findings of vascular malformations. In vascular tumours, the main appearances were red to reddish-purple globules, and lacunae as well as linear irregular vessels. Viswan et al.,[26] describe that it is possible for the dermoscopic characteristics of different vascular lesions to overlap. On the other hand, the colour and predominant dermoscopic feature might indicate the diagnosis. Hemoglobin is the primary chromophores for different vascular features. Under dermoscopy, oxygenated, deoxygenated, and oxidized hemoglobin appear red, blue, and black, respectively. The factors that determine the variation of dermoscopic structures in vascular malformation and vascular tumour are the nature of the vascular lesions, the diameter of vessels, arrangement of vessels, and the contents in the lumen.[26,28] Red globule and lacunae represent grossly dilated and congested solitary capillaries in the upper dermis, while blue-gray globule represent solitary dilated blood vessels in the upper dermis. In this study, dermoscopic images of red structureless area are only found in vascular tumour group, given that the red structureless area corresponds to diffuse capillary or endothelial proliferation.[26,28]

A total of 44% of the study population had undergone histopathological examination. According to Menoghini et al.,[27] the goal of histopathological examination in vascular anomaly cases is to define the exact nature of the lesion, evaluate the tissue involved, and verify the presence of proliferative foci in the lesion. Most vascular anomalies can also be recognized with various imaging modalities. Few proportion of this study population had further examination in the form of USG, CT Angiography, and MRI. These imaging modalities are usually performed to confirm the suspected diagnosis, to determine the extent of the vascular anomaly, and to search for associated abnormalities.[29]

Treatment of vascular anomalies is warranted when lesions are symptomatic or for cosmetic purposes.[5,12] The most common indications for treatment are pain and functional impairment.[5] In this study population, the majority of patients were received laser therapy, particularly for port wine stains in vascular malformation group. Multidisciplinary treatment modalities include combination of surgery and laser, laser and oral beta-blocker, as well as surgery and cryotherapy. According to this study, the most prevalent treatment for vascular malformation was pulsed dye laser (PDL). PDL treatment is well known to be effective for vascular malformations and has been accepted as the gold standard and the first treatment of choice.[30] PDL with its specific wavelength (585 or 595 nm) and a short pulse duration (400–1500 ms) currently gives the best results in infants and children, particularly for capillary malformation.[5,12]

For vascular tumours, the decision about when to begin treatment is influenced by various considerations, including size and location, psychosocial implications, risks, and benefits of the proposed therapy.[9] The treatment of vascular tumour group in this study was evenly similar between oral beta-blocker and multiple treatment modalities. The assortment of therapies includes a combination of oral beta-blocker and topical beta-blocker; surgery and laser; and moreover, surgery and cryotherapy. Since 2008, beta-blockers have become first-line treatment for infantile hemangiomas, the most common tumour of infancy.[31] Oral propranolol, a nonselective beta-blocker, has become the most widely used treatment for infantile hemangiomas needing systemic therapy. Topical beta-blockers such as timolol appear *to be effective in treating small, superficial infantile hemangiomas.*[31] For the 12% of patients for whom no treatment recommendation was made, it was advised to do additional diagnostic testing and use a multidisciplinary approach in order to determine the definite diagnosis or involvement of extracutaneous manifestations. The knowledge transfer and extensive counseling is as important as the treatment recommendations for many patients who may have been misdiagnosed, or not well informed about the vascular condition.[16]

5. CONCLUSION

It is crucial for clinicians to be able to identify, categorize, and treat vascular anomalies appropriately in pediatric patients. A proper diagnosis will enable the clinicians to give appropriate treatment and evaluate the prognosis.

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Disclosure

The authors report no conflict of interest in this study

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