

Vascular Anomalies: The Epidemiological Profile at Srinagarind Hospital

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Background: Vascular anomalies or vascular birthmarks can be divided into 2 major groups: (i) vascular tumors and (ii) vascular malformations. Currently, there are many treatment modalities for these diseases and the treatment plans are varied among sub-specialty physicians.

Objective: To explore the epidemiology of vascular anomalies at Srinagarind Hospital during 2009-2011.

Material and Method: Retrospective chart was reviewed from the out patient clinic's database at Srinagarind Hospital, Faculty of Medicine, Khon Kaen University, Thailand.

Results: There were total of 126 vascular anomalies cases. 89 cases were diagnosed with vascular tumors and 37 cases were vascular malformations. Among 89 cases of vascular tumors, infantile hemangiomas are the most common type (95.5%). The treatment methods for vascular tumors were medical treatments, which were used in majority of the cases (60%), followed by surgical excision, laser treatment, intralesional corticosteroids injection, and the combination of medical, laser and surgical treatment. There were total of 37 cases of vascular malformations. Most of the cases were venous and lymphatic malformations. Treatment methods for these patients were surgical excision, bleomycin injection, and radiation.

Conclusion: Vascular anomalies have various presentations. Treatment is challenging and multidisciplinary teams are involved in taking care the patients with this entity of disease. Setting up vascular anomalies clinic is essential and suggested for the patients with vascular anomalies' problems.

Keywords: Vascular anomalies, Vascular tumors, Vascular malformations

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Vascular anomalies is the term representing vascular lesions which occurred at the time of birth or soon afterward. These can be divided into 2 major groups: (1) vascular tumors and (2) vascular malformations. The classification is based on natural history of growth patterns and differences in cellular kinetics. Most recent classifications from The International Society for the Study of Vascular Anomalies (ISSVA)⁽¹⁾ have recently been published and widely used among sub-specialty physicians.

Vascular tumors

Vascular tumors demonstrate endothelial cell proliferation with typical natural history of rapid growth and spontaneous involution over a period of time. Vascular tumors can be classified as benign, locally

aggressive, and malignant as describe in Table 1.

Among benign vascular tumor, infantile hemangiomas are the most common type. The presentation of infantile hemangiomas usually occurs with a small/tiny preliminary cutaneous lesion followed by the classic growth pattern in 3 phases as follows:

1) Proliferative phase: hemangiomas grow rapidly with maximum growth during 6-9 months of age and usually stop proliferates by the age of 1 year.

2) Involuting phase/plateau phase: hemangiomas have no further growth.

3) Involute phase: hemangiomas start to involute.

The timing of involution is unpredictable. Historical literature⁽²⁾ reports that completed involution occurs at an estimate rate of 10% per year, so that approximately 50% will be totally involuted by 5 years, 70% by 7 years, and 90% by 9 years of age⁽²⁾. According to this classic natural history of growth and involution pattern, without treatment, infantile hemangiomas will resolve themselves spontaneously over time. However, some conditions require early treatment and

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intervention. These include airway hemangiomas, hemangiomas around eyes, nose, mouth, ulcerated hemangiomas, and hemangiomas associated with other anomalies such as PHACES syndrome (Fig. 1). The treatment indications for infantile hemangiomas are listed in Table 2.

The current treatment modalities for infantile hemangiomas are systemic corticosteroids. In various forms, the treatment includes topical, intralesional, and oral formulations with the most common being oral prednisolone which aims to control growth of hemangiomas in proliferative phase.

Other treatment modalities, such as interferon alfa-2a, cyclophosphamide, Vincaalkaloid, and pulsed-dye laser have been used as the second line medication when infantile hemangiomas have no response to those systemic corticosteroids. Most recently, propranolol (a non-selective beta-blocker that is used to treat hypertension, supraventricular tachycardia, congestive heart failure, and thyrotoxicosis) has become a new treatment option in treating infantile hemangiomas. The first initial report which successfully utilized this medication with infantile hemangiomas was published in New England Journal in 2008⁽³⁾. Since 2008, several reports⁽⁴⁻¹²⁾ have demonstrated the effectiveness of propranolol for the treatment of infantile hemangiomas.

However, to determine the safety profile and the true value of propranolol as a first line of therapy for infantile hemangiomas, a randomized controlled trial should be performed to prove its usefulness and warrant a larger



Fig. 1 The patient with PHACES syndrome having large segmental hemangioma on the face.

Table 1. The classification of vascular tumors

Vascular tumors	
Benign vascular tumors	
	Infantile hemangioma/Hemangioma of infancy
	Congenital hemangioma
	Rapidly involuting (RICH)
	Non-involuting (NICH)
	Partially involuting (PICH)
	Tufted angioma
	Spindle-cell hemangioma
	Epithelioidhemangioma
	Pyogenic granuloma (aka lobular capillary hemangioma)
	Others
Locally aggressive vascular tumors	
	Kaposiformhemangioendothelioma
	Retiformhemangioendothelioma
	Papillary intralymphaticangioendothelioma (PILA), Dabska tumor
	Composite hemangioendothelioma
	Kaposi sarcoma
	Others
Malignant vascular tumors	
	Angiosarcoma
	Epithelioidhemangioendothelioma
	Others

powered study involving multiple institutions to address the many remaining questions regarding treatment of infantile hemangiomas with propranolol.

Vascular malformations

Vascular malformations are abnormal clusters of blood vessels that occur during fetal development; therefore, the presentations of these entities occur since birth. Moreover, they show lacking of endothelial cells proliferation with flattened pattern of growth and do not involute spontaneously over a period. The treatment modalities for vascular malformation are mainly the surgical intervention. Medical treatment has no role in this group. Recently, the use of imaging and radio-intervention^(13,14) have become the treatment of choice to correct abnormalities of the vessels. However, this intervention can be done only in the well setting hospital and by the well-trained radiologist sub-specialty.

The epidemiology of vascular anomalies patients, vascular tumors, and vascular malformations seen in Srinagarind Hospital has not been previously characterized. The purpose of this review aimed to gain a better understanding of the spectrum of the vascular anomalies to identify trends in the management of these patients.

Objective

To determine the epidemiology of vascular anomalies at Srinagarind Hospital, Faculty of Medicine, Khon Kaen University during the period of 2009 to 2011. Data including demographic, referring, diagnostic, and treatment information were obtained from the out patient clinic’s database.

Material and Method

The medical records of the patients whom diagnosed vascular tumors and vascular malformations at Srinagarind Hospital, Faculty of Medicine, Khon Kaen University, Thailand between 2009 and 2011 were retrospectively reviewed. The study was approved by the Institutional Review Board No. HE581177.

Statistical analysis was performed using STATA software, version 10. Descriptive statistical methods (mean, standard deviation, median, and frequency) were applied to analyze the demographic data.

Results

Vascular tumors

There were total of 89 cases diagnosed with vascular tumors. Among these, Infantile hemangiomas were the most common type (85 cases, 95.5%), followed by congenital hemangiomas, tufted hemangiomas, and hemangioendothelioma. Female was slightly more common than male with ratio 1.1:1.

Nine percent of the patient had recorded the presentation since birth while the rest of 91% had their presentation within the first month of life. Majority of the cases had the manifestation on the skin located at head and neck region (62%). Lower extremities, upper extremities, and trunk were the following regions of the presentation which were found 19%, 10% and 9%, respectively. Most vascular tumor cases were initially treated by pediatrician (43%), followed by pediatric surgeon (20%), plastic surgeon (15%), and others including orthopedist, otolaryngologist, and ophthalmologist, which were recorded 13%, 5%, and 4%, respectively.

Table 2. Treatment indication for infantile hemangiomas

Treatment indication for infantile hemangiomas
Life-threatening condition; airways hemangioma, high-output heart failure
Impairment of function; vision, hearing, voiding, stooling
Complication; ulceration, bleeding
Significant poor cosmetic outcome
Associated anomalies; PHACES* syndrome, PELVIS# syndrome, LUMBAR+ syndrome

* PHACES; Posterior fossa malformations, Hemangiomas, Arterial anomalies, Coarctation of the aorta and other cardiac defects, Eye abnormalities, Sternal clefting and/or a supraumbilical raphe.

PELVIS: Perineal haemangioma, External genitalia malformations, Lipomyelomeningocele, Vesicorenal abnormalities, Imperforate anus, Skin tag.

+ LUMBAR: Lower body hemangioma and other cutaneous defects, Urogenital anomalies, ulceration, Myelopathy, Bony deformities, Anorectal malformations, arterial anomalies, Renal anomalies

The treatment methods included medical treatment, such as systemic corticosteroids, propranolol, interferon alpha, and vincristine were used in majority of the cases (60%) followed by surgical excision (17%), laser treatment (8%), intralesional corticosteroid injection (4%), and the combination of medical/laser/surgical treatment (3%). The remaining 8% of the patients recieved non-intervention therapy (Fig. 2).

Vascular malformations

Thirty-seven cases of vascular malformation were identified for the study. Most of the cases are venous and lymphatic malformations. All patients had their presentations since birth. The lesions grew proportionately with the patients' growth. Female was slightly more common than male with a ratio of 1.1:1. Majority of the cases manifested on the skin, located on the head and neck region (46%), followed by lower extremities (32%), trunk (13%), and upper extremities (9%).

Most of vascular malformation cases were initially treated by plastic surgeon (35%) followed by pediatric surgeon (30%), and radiologist by radio-intervention unit (30%). For other non-intervention treatment groups, the patients were followed by pediatrician (5%). The treatment options for vascular malformations were surgical excision (36%), bleomycin injection (30%), laser (27%), tapping (4%), and radiation (3%).

Discussion

The data of vascular anomalies at Srinagarind Hospital, Faculty of Medicine, Khon Kaen University showed various presentations. Only 9% of vascular tumors patients had their presentation since birth. Most of them (91%) showed their presentations later within 1 month of life, in contrast, the vascular malformations patients showed totally 100% of their presentations at the time of birth. These findings correlated to the fact that vascular malformations are the result of abnormal morphogenesis during embryo development; therefore, the manifestation had presented since then. Previous literature⁽¹⁵⁾ showed the data of females predominant in vascular tumors, while there was no sex difference in vascular malformations. Our data showed slightly more common cases in girls than boys with the ratio of 1.1:1 in both groups. This can be explained from the small number of the patients from the study, which may have contributed to this result. Both vascular tumors and vascular malformations were most prevalent and

manifested in the head and neck regions. The other areas of presentation are shown in Fig. 3.

The distributions of the patients based on their first visit were varied. Pediatricians played the major role in the patients with vascular tumors while plastic surgeons and pediatric surgeons were involved mainly in the patients with vascular malformations. Fig. 4 showed the distributions of the patients having

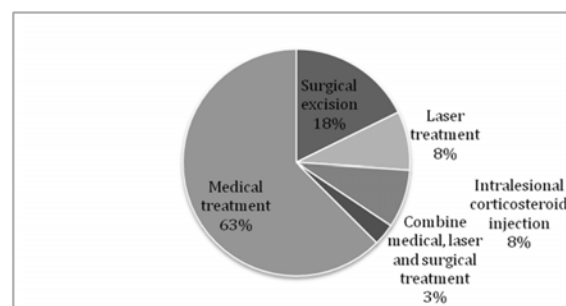


Fig. 2 Treatment options for vascular tumors during 2009-2011.

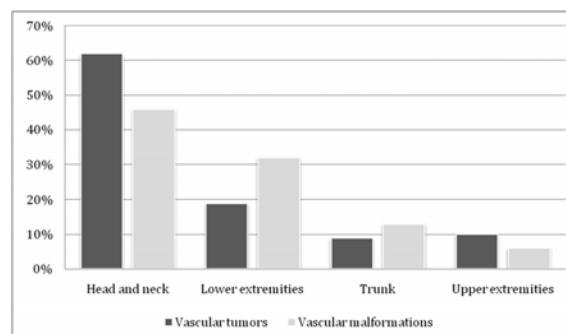


Fig. 3 Comparison of the presenting area between vascular tumors and vascular malformations.

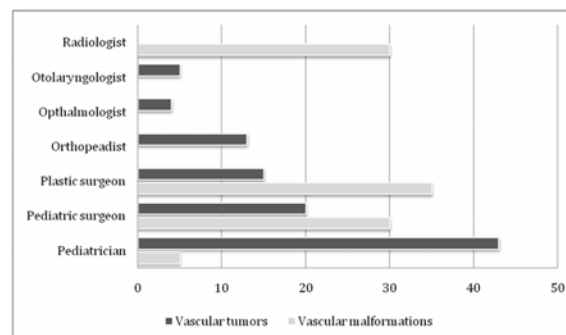


Fig. 4 Distributions of the patients having vascular tumors and vascular malformations in each sub-specialty clinic based on their first visit.

vascular tumors and vascular malformations in each sub-specialty clinic based on their first visit.

In the majority of treatment modalities for vascular tumors, especially infantile hemangiomas, the most common type among vascular tumors from the study were medical treatments, which included systemic corticosteroids, propranolol, interferon alpha, and vincristine. According to recent review of novel propranolol use for infantile hemangiomas, we currently have no data on the treatment. However, our institute is in the process of data gathering for further study of the medical treatment for infantile hemangiomas with focus on the use of propranolol.

Vascular malformation treatments were mostly surgical excision. The radio-intervention such as bleomycin injection and coagulation of the abnormal vessels by radiologists were becoming more frequent in the treatment of vascular malformations in our institute.

Conclusion

Vascular anomalies have various presentations. Treatment is challenging and multidisciplinary teams are involved in taking care of the patients with this entity of diseases. Setting up vascular anomaly clinics is essential and suggested for the patients with vascular anomalies' problems.

What is already known on this topic ?

The manifestations and treatments of vascular anomalies are varies and depend on sub-specialty care team. Although the International Society for the Study of Vascular Anomalies (ISSVA) has recently published the new classification and widely used among sub-specialty physicians, there were still variety of treatments among these patients.

What this study adds ?

This is the first data collected in Srinagarind Hospital, Faculty of Medicine, Khon Kaen University. The data represented incidence and manifestation of vascular anomalies in a tertiary care setting. These also support the idea of setting up vascular anomaly clinics to provide an holistic approach and multi-disciplinary care for patients with vascular anomaly problems.

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Potential conflicts of interest

None.

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ความผิดปกติของหลอดเลือด: ระบาดวิทยาที่โรงพยาบาลศรีนครินทร์

ลีลาวดี เตหาเสถียร, พัชรินทร์ ตันมิ่ง, สุนีย์ พนมบัวเลิศ, รัฐพล อุปลา, จรูญ เจตศรีสุภาพ

ความผิดปกติของหลอดเลือดสามารถแบ่งได้เป็น 2 กลุ่ม กล่าวคือ (1) vascular tumors และ (2) vascular malformations การศึกษาความผิดปกติของหลอดเลือดในปัจจุบัน มีแนวทางการรักษาที่หลากหลาย ทั้งขึ้นกับแพทย์เฉพาะทางในแต่ละสาขาที่ได้รับการดูแลรักษา

วัตถุประสงค์: เพื่อศึกษาระบาดของภาวะหลอดเลือดผิดปกติที่รับการรักษาในโรงพยาบาลศรีนครินทร์ คณะแพทยศาสตร์ มหาวิทยาลัยขอนแก่น ระหว่างปี พ.ศ. 2552-2554

วัสดุและวิธีการ: เป็นการศึกษาย้อนหลังเก็บข้อมูลผู้ป่วยจากเวชระเบียน ที่รับการรักษาในโรงพยาบาลศรีนครินทร์ คณะแพทยศาสตร์ มหาวิทยาลัยขอนแก่น ระหว่างปี พ.ศ. 2552-2554

ผลการศึกษา: มีผู้ป่วยที่มีภาวะความผิดปกติของหลอดเลือด 126 ราย ได้รับการวินิจฉัยเป็นกลุ่ม vascular tumors 89 ราย และ vascular malformations 37 ราย ในกลุ่ม vascular tumors พบว่า infantile hemangiomas เป็นชนิดที่พบบ่อยที่สุด (95.5%) การรักษาโรคในกลุ่ม vascular tumors ส่วนใหญ่เป็นการรักษาด้วยยา (60%) ยาที่ใช้ในการรักษาได้แก่ systemic corticosteroids, propranolol, vincristine, interferon alpha เป็นต้น การรักษาอื่นๆ ได้แก่ surgical excision, laser treatment และ intralesional corticosteroids ผู้ป่วยกลุ่ม vascular malformations มีจำนวน 37 ราย ส่วนใหญ่เป็นความผิดปกติของหลอดเลือดดำและหลอดเลือดน้ำเหลือง แนวทางการรักษาผู้ป่วยกลุ่ม vascular malformations ได้แก่ surgical excision การฉีด bleomycin และการฉายรังสี

สรุป: ความผิดปกติของหลอดเลือดมีอาการแสดงที่หลากหลาย การรักษาจำเป็นต้องอาศัยการดูแลรักษาาร่วมกันของผู้เชี่ยวชาญหลายกลุ่ม การจัดตั้งคลินิกบูรณาการในการดูแลรักษาผู้ป่วยร่วมกันแบบสหสาขาวิชาจึงมีความจำเป็นในการวางแผนการรักษาผู้ป่วยในกลุ่ม vascular anomalies
