Original Research Article

Vascular malformations - Treatment modalities

Balaga Venkata Rao[1], M. Swatantra Bharathi[2*]

[1]Associate Professor, Department of Surgery, ACSR Government Medical College, Nellore, Andhra Pradesh, India
[2]Assistant Professor, Department of Plastic Surgery, Sidhartha Medical College, Vijayawada, Andhra Pradesh, India
[*]Corresponding author email: **bharathisreya@gmail.com**

International Archives of Integrated Medicine, Vol. 6, Issue 9, September, 2019.

Available online at http://iaimjournal.com/
ISSN: 2394-0026 (P) **ISSN: 2394-0034 (O)**
Received on: 27-08-2019 **Accepted on:** 02-09-2019
Source of support: Nil **Conflict of interest:** None declared.
How to cite this article: Balaga Venkata Rao, M. Swatantra Bharathi. Vascular malformations - Treatment modalities. IAIM, 2019; 6(9): 58-65.

Abstract

Background: Vascular malformation is a general term that includes congenital vascular anomalies of veins, lymph vessels, both veins and lymph vessels, or both arteries and veins vascular malformation, is a blood vessel abnormality. These are all present at birth and become apparent at different ages. Most are known to occur during development of the arteries, veins, and/or lymph vessels, but without specific cause. Most are congenital and few appear late in life.
Aim: It was using different treatment modalities to reform, reshape, and reorganize to obtain near normal appearance and function.
Materials and methods: An observational study done in VIMS - Visakha Institute of Medical Sciences, a superspeciality hospital, 20 cases of vascular malformations are taken from period January 2017 –August 2019. Consent was taken from all the patients, and counseling was done regarding the procedures we do, about surgical/ non-surgical methods, stages required and follow up.
Results: Statistical analysis was done, results were tabulated and displayed in pie diagrams. Satisfaction scale was shown by VAS (visual analogue scale).
Conclusion: Vascular malformations are mesenchymal lesions may be congenital/ acquired and may present late in life. Sometimes few congenital hemangiomas spontaneously resolve by growth of child, some if large may require steroids for suppression and to prevent recurrences. They are known for recurrences or residual lesions and surgery may be done in stages to obtain optimal results. Non-surgical methods are done by intralesional sclerosants.

Key words

Vascular malformation, Treatment modalities, Satisfaction score.

Introduction

Vascular malformations are benign (non-cancerous) lesions that are present at birth, but may not become visible for weeks or months after birth. Unlike hemangiomas, vascular malformations do not have a growth cycle and then regress but instead continue to grow slowly throughout life. Most vascular malformations are sporadic (occurring by chance), though some are inherited in a family as an autosomal dominant trait. Autosomal dominant means that one gene is necessary to express the condition, and the gene is passed from parent to child with a 50/50 risk for each pregnancy. Males and females are equally affected and there is great variability in expression of the gene. In other words, a parent may unknowingly have had a hemangioma because it faded, but child is more severely affected.

Vascular anomalies represent a spectrum of disorders from a simple "birthmark" [11, 12] to life-threatening entities. Incorrect nomenclature and misdiagnoses are commonly experienced by patients with these anomalies. Accurate diagnosis is crucial for appropriate evaluation and management, often requiring multidisciplinary specialists. Classification schemes provide a consistent terminology and serve as a guide for pathologists, clinicians, and researchers. One of the goals of the International Society for the Study of Vascular Anomalies (ISSVA) is to achieve a uniform classification. The last classification [10, 11] (1997) stratified vascular lesions into vascular malformations and proliferative vascular lesions (tumors). However, additional disease entities have since been identified that are complex and less easily classified by generic headings, such as capillary malformation, venous malformation, lymphatic malformation, etc. We hereby present the updated official ISSVA classification of vascular anomalies. The general biological scheme of the classification is retained. The section on tumors has been expanded and lists the main recognized vascular tumors, classified as benign, locally aggressive or borderline, and malignant. A list of well-defined diseases is included under each generic heading in the "Simple Vascular Malformations" section. A short definition is added for eponyms. Two new sections were created: one dealing with the malformations of individually named vessels (previously referred to as "truncular" malformations); the second groups lesions of uncertain or debated nature (tumor versus malformation). The known genetic defects underlying vascular anomalies are included in an appendix. This classification is meant to be a framework, acknowledging that it will require modification as new scientific information becomes available. Vascular anomalies (vascular tumors and vascular malformations), often named "angioma" or hemangioma, in fact represent a broad spectrum of disorders from a simple "birthmark" to life-threatening entities, which affect mainly infants, children, and young adults. Certain vascular anomalies, such as salmon patch (nevus simplex) or infantile hemangioma, are very common. Others such as port wine stain are uncommon, and still others are quite rare. Despite long-standing efforts to promulgate standard classification, nomenclature terminology of vascular anomalies continues to be confusing [1-4]. The term "hemangioma" is, for example, commonly used to name different types of vascular tumors as well as vascular malformations, despite the different constitution, natural evolution, and treatment of these 2 groups of lesions. Incorrect nomenclature and misdiagnoses are commonly experienced by patients with vascular anomalies [2]. Accurate diagnosis and common terminology are crucial for appropriate evaluation and management, often requiring multidisciplinary specialists.

One of the goals of the International Society for the Study of Vascular Anomalies (ISSVA) is to achieve a uniform classification. The 1996 ISSVA classification stratified vascular anomalies into vascular malformations and proliferative vascular lesions (tumors) [5, 6]. This classification was then "unofficially" updated on the basis of evolving knowledge a decade later [7]. However, since then, knowledge about these

disorders has increased considerably. The genetic basis of many types of vascular malformations has been elucidated and additional disease entities have been identified that need more precise classification rather than generic headings such as capillary malformation (CM), venous malformation (VM), lymphatic malformation (LM), etc., which have been used previously. The ISSVA Classification of Vascular Anomalies was recently updated by the Society's Scientific Committee and Board to incorporate these changes and was adopted at the last workshop in Melbourne, Australia (April 2014). The interactive document is available at www.issva.org. This classification is meant to represent the state-of-the-art in vascular anomalies classification, acknowledging that it will require modification as new scientific information becomes available.

There are several types of vascular malformations:

- **Capillary (port wine stains):** Always present at birth as pink or purple skin patches. Distributed in dermatological fashion.
- **Venous malformation:** Often confused with a hemangioma, these malformations are soft to the touch and the color disappears when compressed. They are most commonly found on the jaw, cheek, tongue and lips, or anywhere in body like intra muscular, intracranial and so on.
- **Lymphatic malformations:** These form when excess fluid accumulates within the lymphatic vessels.
- **Arteriovenous malformations:** Abnormal connections between arteries and veins, resulting in a high flow, pulsating collections of blood vessels. These are congenital and also acquired due to trauma.
- **Mixed:** A combination of any of the other four types (lymphaticovenous).

Materials and methods

An observational study was done in VIMS - Visakha Institute of Medical Sciences, a superspeciality hospital. 20 cases of vascular malformations were seen in the plastic surgery out-patient department from period January 2017 – August 2019 which were included in the study. Twenty cases were randomly selected for which an intervention was done either surgery or intraesional sclerosants. Apart from these cases to outpatient department we noticed also hemangiomas in children for whom masterly inactivity or steroid orally are given and treated. Necessary investigations like imaging which included computed tomogragraphy angiogram. Magnetic resonance imaging techniques for confirmation were done. Patients whom selected were decided for treatment and optimal treatment modality was choosen accordingly either surgery or intralesional sclerosants. Consent was taken from all the patients, counseling was done regarding the procedures we do, stages required and follow up. Initially as per the classification if ISSVA we could come to a conclusion of type of vascular anomaly and accordingly treatment plan was instituted. Either surgery, intralesional scelosants, oral steroids, interventional radiotherapy. In our institute Visakha Institute of Medical Sciences we had opted two modalities of treatment which were feasible and available to us. Intralesional sclerosants and other surgical excision/ debulking/ contouring according to the presentation of case was done. Few cases needed staged surgical excision of 3-4 times to get a desired result and some cases had residual/ recurrences.

Counseling of patients and attendants was done in all cases regarding residual/ recurrences and long follow up. Routine surgical profile and anesthesia fitness was taken. For all the patients consent was taken and in few patients video counseling was taken regarding risk of surgery in case of AV malformations in few of untoward effects like hemorrahagic complications and death. Surgeries were performed either in regional anesthesia or general anesthesia depending on the age of patient, size and nature of lesion whether it is slow flow or fast flow and

finally the competence and experience of the surgeon. A few venous malformations small /which were not emenable to surgery were treated with intralesional sclerosants and each episode a gap of 3-4 months is given so that the tissue reaction and settlement of scar occurs. All the patients after the procedure visual analogue satisfaction score was analyzed. All patients were followed till maximum desirable effect is obtained since the settlement of scar occurs after 3-4 months period and sometimes residual or recurrences need another surgery or intralesional sclerosants.

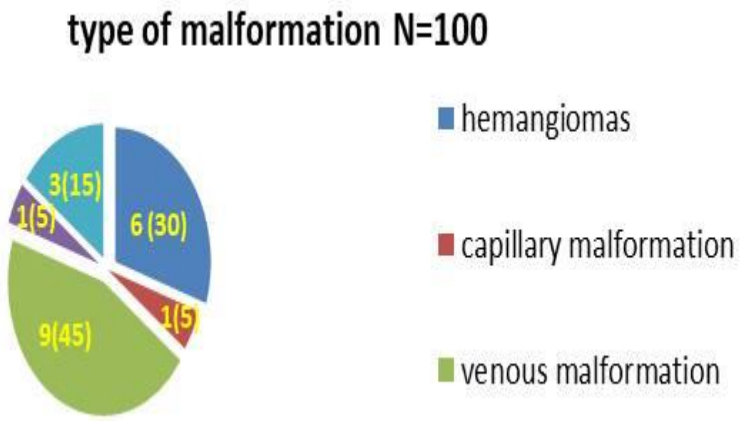
Statistical analysis

Data was entered and analyzed in Microsoft excel spread sheet (version 2016). Categorized data in simple proportions and quantitative data were expressed using means and standard deviation.

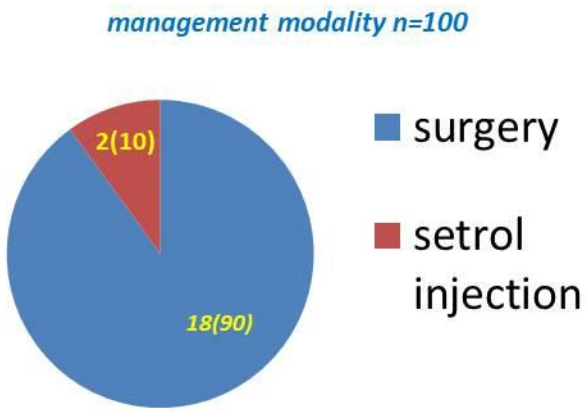
Results

All 20 patients were admitted and required treatment given either surgery or intralesional sclerosant injection. Results were tabulated and few variables were taken into consideration for statistical analysis, like type of lesion, (classification), anesthesia used either regional or general, longest and shortest follow up to out-patient department, management modality opted for each case and finally the satisfaction of patients was evaluated by visual analogue score (VAS). Shortest follow up being 3 months and longest follow up being 32 months (**Graph – 1 to 3 and Figure – 1 to 4**).

Graph – 1: Type of malformation.



Graph – 2: Management modality.



Graph – 3: Anesthesia.

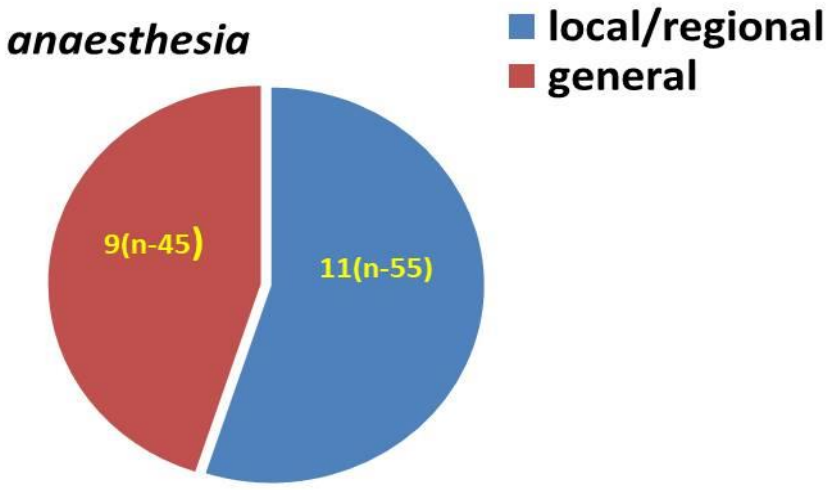


Figure – 1: Visual analogue scale: Assessment of satisfaction - VAS. (Fantastic -18(90), Really good-2(10).)

Figure – 2: Capillary malformation – face (trigeminal distribution).

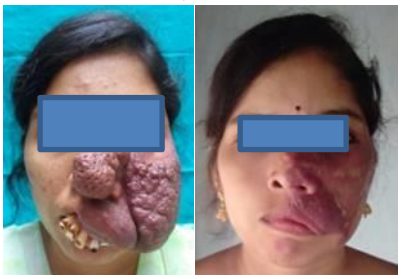
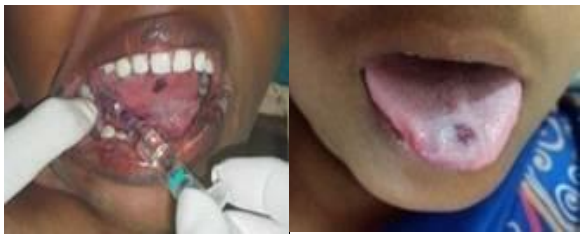


Figure – 3: Venous malformation- intra temporalis – malformation.

Figure – 4: Sclerosant (Setrol) intralesional injection).



Discussion

Dr. Mulliken is pioneer in vascular malformations concept. He is a Professor of Surgery at Harvard Medical School and Senior Associate in Surgery at Children's Hospital Boston. He is co-director of the Vascular Anomalies Center and Director of the Craniofacial Centre. He is the co-founder and past president of the International Society for the Study of Vascular Anomalies. His most recent textbook is Vascular Anomalies: Hemangiomas and Malformations, co-edited with P.E. Burrows and S.J. Fishman. Dr. Mulliken's major clinical interests are cleft lip/ palate, vascular anomalies and craniofacial anomalies. His basic research focuses on molecular causes of craniofacial and vascular anomalies. Dr. Mulliken developed the internationally accepted classification system for vascular anomalies and he is considered one of the world's foremost experts in vascular anomalies.

Vascular malformations are a type of birthmark often present at birth or may appear late in life incidentally with trivial incidences. They may or may not have any symptoms or may cause some causmetic disturbances to individuals which make them social stigma to move in society like playing and moving with peers, school going and others. Some times they are acquired like av malformations which appear late in life. These malformations involve or arise from arteries, veins, capillaries, lymphatics and combinations of these. There are different type of vascular malformations and they are named according to which type of vessel they arise. In combination of different origins one will be predominant like lymphatico venous ,arterivenous and so on. The most common blood vessel abnormality thet impact chidren in the first year of life are hemangiomas, the majority of which gets spontaneous resoption or fade away in few years. 90% by first year of life, few with in 7 years of life and at later stage they appear as faint scaly pale birth marks as combstones of prevoius lesions.

Blood vessel malformations may accompany a variety of genetic or inherited syndromes. Patients that present with vascular malformation need mutidisciplinary care to ensure coordinated treatment for all of their symptoms. A wide variety of combinations and syndromes commonly associated with vascular malformations are treated through the divisions of plastic surgery and interventional radiologist in stages to obtain optimal desirable results. Divisions of plastic surgery and interventional radiology, with additional colloboration from orthopaedic surgery, hematology and otolaryngology when indicated. These include:

- Venolymphatic malformation, including angiokeratomas
- Klippel-trenaunay syndromes (capilary, venolymphatic malformations)
- Parkes weber syndromes (capillary, aretrivenous)
- CLOVES (congnital lipomatous overgrowth, vascular malformation, epidermal nevus, spinal abnormalities)
- Proteus syndrome
- Herditary hemorrhagic telangiectasia (osler –weber- rendu syndrome)
- Blue rubber bleb nevus syndome (BRBNS)

Vascular malformations are usually present at birth, grow proportionally with the child, do not expand rapidly during infancy, and do not disappear. Sometimes lesions in older people, blood flow increase through abnormal channels between arteries and veins. Vascular malformations can cause cosmetic deformities of the head and neck which can intervene or interfere with normal function in these areas. This cause increase blood flow to that area will make the nearby tissues to get hypertrophy and hyperplasia and hyper functioning sometimes of certain glands nearby causing various physiological disturbances in the individual. The bone nearby also increase in size bizarre and produce cosmetic deformity and appearance. In certain areas they cause pain, swelling, bleeding, infection and vicious cycle may produce irreducible fibrosis.

Vascular malformations can be affected by hormonal changes during puberty and pregnancy and can result from blood or fluid accumulating in poorly formed veins or lymphatic channels. Most vascular malformations require [6, 7, 8] treatment\ intervention for various reasons like to improve appearance (cosmetic purposes), alleviate pain, swelling, infection and fibrotic hard lesions.

Many vascular malformations are very complex are known for residual or recurrent lesions [11, 12, 13]. So we take a collaborative approach for diagnosis and treatment by different modalities. So treatment of vascular malformations needs multimodality and multidisciplinary means. We need imaging modalities for diagnosis and confirmation initially either by ultrasound, color

Doppler [4, 5], computed tomography or magnetic resonance imaging depending on the availability, patient affordability, surgeon choice and confirmatory possibility percentage. After confirmation different modes of treatment also depends on certain factors like: patient age, site of lesion, type of lesion, percentage of recurrences/ residually of the lesion, surgeon choice, availability of therapy, patient compliance and follow up [7, 8, 9]. Of all patient compliance and follow up is prime most important of all. The most common techniques are:-

- Steroid administration (orally)
- Embolisation
- Laser treatment
- Sclerotherapy
- Surgery

Hemangiomas and capillary malformations or in any malformation initially in childhood steroids are given in tapering doses to reduce the size of the lesions or make the lesions approachable to surgery. Counseling to parents and individuals is very essential for a long follow up and multimodality of treatment. Role of steroids is to reduce the size and inflammation by its anti-inflammatory effect. The side effects of steroids are to be concerned by administration of antacids orally.

In our institute the opted the modalities of management are surgery and sclrotherapy. Few lesions are directly approached by surgery and few managed by sclerotherapy. Sclerotherapy is done for venous and lymphatic malformations. By using sclerotherapy the intralesional injection of setrol what we used in our institute causes the endothelial damage of lesions like chemical cautery and the opposing surfaces get adhere with each causing the lesion shrink in size and also the vascularity decreases so that the feeding vascularity and growth potential surface if the lesion are reduced. This sclertherapy also done has another advantage apart from decreasing the size and vascularity it creates a plane of demarcation from surrounding normal tissues thus enhancing the efficacy of the surgical procedure and lessens the blood loss or blood requirements during and after the surgical procedure. The sclerotherapy also enhances fibrosis and recurrences chances are reduced. Prior inoperable lesions are made operable by sclerotherapy. Vital structures are saved or protected particularly on face thus protecting function of sense organs on face. One of the disadvantage or dangerous side effect of sclerotherapy is if drug enters into general circulation it sometimes causes catastrophic hemodynamic disturbances and collapse of patient, that to be explained prior to procedure to all patients. Written and informed consent to be taken and mandatory to explain the risks to all patients undergoing sclerotherapy. A period of 3-4 months interval is given in between two injections for the time to develop fibrosis and sclerotherapy produces an inflammation which should get settled before next injection.

Surgery is done in stages to debulk the disfigured lesions on face or anywhere on body. Surgical procedure is also done in excess bone growth or cartilage accordingly apart from excision of soft tissue and blood vessels. Contouring of a structure is done by excision and repeated till desirable effect is obtained. Surgery creates a layer of fibrosis which cuts off the blood supply and helps in reducing recurrences. It forms definitive procedure for treatment of the malformations among all the management modalities and effective means of procedure when compared to others.

Conclusion

Vascular malformations may present clinically at different ages starting from birth and also in late life. Most of present mainly with cosmetic problems apart from functional disturbances. The approach towards these lesions may vary depending on many factors regarding patient, surgeon, availability of modality and important is patient follow up.

References

1. Hand JL, Frieden IJ. Vascular birthmarks of infancy: resolving nosologic confusion. Am J Med Genet., 2002; 108(4): 257–264.
2. Hassanein AH, Mulliken JB, Fishman SJ , Greene AK. Evaluation of terminology for vascular anomalies in current literature. Plast Reconstr Surg., 2011; 127(1): 347–351.
3. Kollipara R, Dinneen L, Rentas KE, et al. Current classification and terminology of pediatric vascular anomalies. AJR Am J Roentgenol., 2013; 201(5): 1124–1135.
4. Mulligan PR, Prajapati HJ, Martin LG, Patel TH. Vascular anomalies: classification, imaging characteristics and implications for interventional radiology treatment approaches. Br J Radiol., 2014; 87(1035): 20130392.
5. Enjolras O, Mulliken JB. Vascular tumors and vascular malformations (new issues). Adv Dermatol., 1997; 13: 375–423.
6. Enjolras O. Classification and management of the various superficial vascular anomalies: hemangiomas and vascular malformations. J Dermatol., 1997; 24(11): 701–710.
7. Enjolras O, Wassef M, Chapot R. Color Atlas of Vascular Tumors and Vascular Malformations. Cambridge, United Kingdom: Cambridge University Press; 2007.
8. Virchow R. Angioma in die Krankhaften Geschwülste. Berlin, Germany: Hirshwald; 1863, p. 306–425.
9. Wegener G. Ueber Lynmphangiome. Arch Klin Chir., 1877; 20: 641–707.
10. Mulliken JB, Young AE. Vascular Birthmarks: Hemangiomas and Malformations. Philadelphia, PA: Saunders; 1988.
11. Wohlgemuth WA, Wölfle K, Schuster T, Schlimok G, Bohndorf K. Hereditary vascular malformations: classification, symptoms, diagnostics and prognosis. Zentralbl Chir., 2012 Oct; 137(5): 440-5.
12. Moure C, Reynaert G, Lehmman P, Testelin S, Devauchelle B. Classification of vascular tumors and malformations: basis for classification and clinical purpose. Rev Stomatol Chir Maxillofac., 2007 Jun; 108(3): 201-9.
13. Philandrianos C, Degardin N, Casanova D, Petit P, Bartoli JM, Bardot J, Magalon G. Diagnosis and management of vascular anomalies. Ann Chir Plast Esthet., 2011 Jun; 56(3): 241-53.