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ORIGINAL ARTICLE

Profile of AV Malformations - An Experience of Tertiary Care Centre

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Abstract

Background: AV malformations are aberrations in the development of vessels with varied presentations that may lead to aesthetic, limb and life-threatening complications. Identification and treatment of the vascular malformations is a challenging task for surgeons. **Aims & Objectives**: To study clinical presentation/ diagnostic modalities/ treatment strategies, complications and recurrence rates of surgical and other interventional strategies. **Material and Methods:** A retrospective study of 150 patients who were treated over a period of 3 years from a tertiary centre with regular follow-ups was done to study the best diagnostic and treatment plans for initial and recurrent lesions. **Results:** The study showed an age group of <1-75 years with a wide range of clinical presentations. Doppler ultrasonography supplemented by CT/MRI angiography was the main diagnostic tool. Surgery followed by hybrid and intralesional interventions were used for primary and recurrent lesions showing varying degrees of recurrence rates (25%). **Conclusion:** A multidisciplinary approach is important in the management of AV malformations. Coloured Doppler with CT/MRI angiography was the best diagnostic tool. Overall surgery alone or in hybrid forms was the mainstay of treatment.

Keywords

Vascular Malformations, Arteriovenous malformations, venous malformations, sclerotherapy

Introduction

AV malformations are a group of conditions that result from congenital aberrations encompassing a wide variety of lesions related to the disorder of vascular development causing a challenging task for surgeons in terms of diagnosis and treatment. The Peripheral vascular malformations encompass a wide spectrum of lesions that can present as incidental findings or symptomatic lesions and can produce potentially life or limb-threatening complications [1]. Vascular malformations are thought to result from developmental errors during Embryogenesis. According to the International Society for the Study of Vascular Anomalies (ISSVA), the malformations are categorized as low or high flow[2]. Low-flow malformations can further be classified depending on their primary cell type: venous, lymphatic or capillary, while high-flow malformations are of the arteriovenous type 2.

The risk of vascular malformations increases with genetic conditions including Blue rubber bleb nevus syndrome, CLOVES (congenital lipomatous overgrowth, vascular malformations, epidermal nevi, and spine deformities), Hereditary hemorrhagic telangiectasia (also called Osler-Weber-Rendu syndrome), Klippel-Trenaunay syndrome (KTS), Parkes Weber syndrome.

Symptoms of vascular malformations are diverse and depend on the type of malformation with incidental diagnosis but they can also lead to pain, thrombosis,

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hypertrophy of limbs, neuritic signs & symptoms, compression of functional structures, and cosmetic complaints[3].

Additionally, recent advancements in genetics have allowed for the development of targeted therapies, which are currently being studied and are under evaluation [4,5]. These therapies might prove beneficial, especially for patients with therapy-resistant, diffuse, multiple, recurrent, inaccessible lesions.

Different surgical techniques can be used to eradicate vascular malformations. Well-defined malformations are suitable for surgical treatment. Complete removal is not desired in cases where the patient is suffering from life or life-threatening complications. These patients may benefit from debulking surgery. Though regrowth can occur even after debulking surgery it remains a better option than causing damage to the vital structures. In some patients, compartmentalization is another option for management. In this technique, the surgeon places big sutures in numerous locations of the malformation to create multiple divisions within the lesion [6]. Results after this surgery especially recurrence are inadequately reported [7,8].

Non-surgical treatment of vascular malformations, such as embolization, has developed progressively over time. After embolization, regression of lesions with a decrease in their vascularity can make complete or subtotal excision of lesions easier. This outcome could be short-term or used in a palliative setting. Sclerotherapy is acknowledged as a good treatment option, especially for slow-flow venous malformation[9]. Repeated courses of sclerotherapy can clinically improve malformations, therefore, this technique can also be used as an adjuvant therapy to surgery [10]. Studies show that preoperative embolization reduces blood loss and makes dissection of malformations easier during surgery.

We reviewed our experience over the last 3 years with vascular malformations with emphasis on their clinical spectrum, best diagnostic tools, and best preferred available treatment for AV malformations in our tertiary center. This study is the first of its kind being conducted by the Department of Cardiovascular and Thoracic Surgery Super-speciality Hospital, GMC Jammu and no such study has been conducted in the past. Such a study will help in making protocols for the best diagnostic and management strategies for our patients with minimum morbidity and recurrences.

Material and Methods

A retrospective analysis of 150 patients with vascular malformations who were managed in Govt. Super speciality Hospital, GMC, Jammu in the Department of CTVS treated over 3 years (2020-2023) has been done. Their medical records were analyzed and the vascular malformations were evaluated based on their mode of presentation, patient's age at presentation, symptomatic presentation, diagnostic modalities, and treatment procedures, in-hospital morbidities, complications & recurrences.

Additional diagnostic studies like Ultrasonography and Colour Doppler flow studies were done to study the nature of flow to differentiate between slow-flow and high-flow lesions. MRI & CT angiography were used to localize the vascular malformation in relation to involved tissues and to study the feeder and surrounding vessels.

Inclusion Criteria:

1. Age: <1-75 years
2. Ready for regular follow-ups
3. AV Malformation of extremities and non-visceral areas
4. One or more symptoms like pain swelling bluish marks and throbbing pulsatile lesions
5. Patient choosing multi-modality treatment

Exclusion Criteria

1. Visceral AV Malformations
2. Brain AVM
3. Known sensitivity to intralesional agents
4. Patient lost to follow-ups

Results

The patient's medical records were analyzed retrospectively for a period of 3 years. The age range was less than 1 year to 75 years. A total of 150 cases were included in the present study constituting about 85 male and 65 female patients.

In all the patients, appropriate clinical history & physical examination were done to make a proper diagnosis.

After assessing the patients, it was observed that there were about, 110 slow flow and 40 high flow lession present.

The distribution of patients by age shows that the highest percentage (33.3%) is in the 10-30 years age group, while the lowest percentage (6.7%) is in the 0-1 year and >70 years age groups. This indicates that most patients are in the younger to middle-aged groups.(*Table 1*)

The data shows a male-to-female ratio of approximately 56.7% males to 43.3% females, indicating a higher

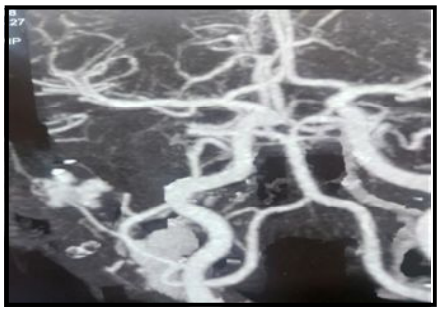
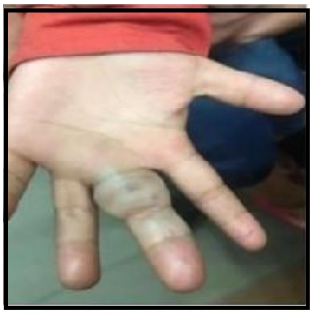


Fig1: CT angiography showing AV malformation



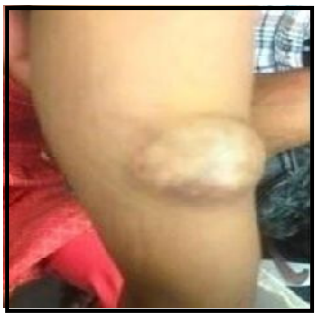


Fig 2,3: Manifestations of venous malformations on finger and knee

Table 1: Age group of the patients

Age	No. Of Patients	Percentage(%)
0-1 Year	10	6.7
1-10 Years	40	26.7
10-30 Years	50	33.3
30-50 Years	20	13.3
50-70 Years	20	13.3
> 70 Years	10	6.7

Table 2: Male Female Ratio

Sex	No. Of Patients	Percentage (%)
Male	85	56.7
Female	65	43.3

Table 3: classification of lesions

Flow Characteristics	No. Of Patients	Percentage(%)
Slow	110	73.3
High	40	26.7
Venous	65 (Out Of All The Lesions)	

Table 4: No. of cases according to their presentation

Clinical Presentation	No. Of Patients	Percentage (%)
Incidental	20	13.3
Pain	70	46.7
Painless Swelling	35	23.3
Pulsatile masses	15	10
Others	10	6.7

Table 5: Proposed plan of treatment

Proposed Treatment	No. of patients	Percentage (%)
Surgery alone	70	46.7
Hybrid Procedure	30	*20*
Intralesional treatment	50	33.3

Table 7: Associated Complications

Complications	No. Of Patients	Percentage
Seroma	6	4
Hematoma	6	4
Skin necrosis	4	2.6
ThrombophlebiQs	4	2.6
DVT	3	2
Neuroparaxia	4	2.6
Ischemic changes	3	2
Recurrence	38	25

Table 6: Investigations

Investigation	No. Of Patients	Percentage
USG with Colour Doppler	150	100
CT angiography	70	46.7
MRI Angiography	80	53.3

proportion of male patients than female patients.(*Table 2*) Most patients (73.3%) have slow-flow lesions, while 26.7% have high-flow lesions.(*Table 3*) AV malformations (with exclusive 65 venous malformations)and 40 high-flow AV malformations. Most of the slow flow lesions presented as painless compressible masses (diffuse or well localized)while the fast flow malformations presented at early ages, initially painless masses later on warm and painful having palpable thrills & bruitis .Pain is the most frequent clinical presentation, affecting 46.7% of patients, followed by painless swelling at 23.3%. Incidental findings are seen in 13.3% of cases, with pulsatile masses and other symptoms less common.(*Table 4*) All patients underwent USG with Color Doppler, while 53.3% also had MRI angiography and 46.7% underwent CT angiography. This indicates a comprehensive use of imaging techniques to evaluate patients.(*Table 5*) Surgery alone was the most frequently proposed treatment, recommended for 46.7% of patients. Intralesional treatment was suggested for 33.3% of patients, and a hybrid procedure was recommended for 20%.

This distribution reflects a preference for surgical approaches, with a substantial number also considered for less invasive or combined methods.(*Table 6*)

The most common complication was recurrence, affecting 25% of patients, while seroma and hematoma each occured in 4% of cases. Other complications, such as skin necrosis, thrombophlebitis, neuroparaxia, DVT, and ischemic changes, were less frequent, each affecting between 2% and 4% of patients.(*Table 7*)

Recurrence to some extent was seen in 20 -25% of cases of these 10 patients were treated with repeat excision and in 7 patients intralesional sclerotherapy was given.

In our series, we had one patient of Blue rubber bleb nevus syndrome who was treated by limited resection and foam sclerotherapy, and three patients of Klippel-Trenaunay syndrome(KTS) treated in a staged manner.

Discussion

Vascular malformations are a complex and heterogeneous group of congenital anomalies that are categorized based on flow characteristics into slow-flow (capillary, venous, lymphatic) and fast-flow (arteriovenous malformations) types. These malformations often present with overlapping features and can include components of multiple subtypes, such as mixed lymphatic-venous malformations or arteriovenous malformations (AVMs) with significant arterial components that contribute to high flow within venous channels (Madani *et al*)[1] The importance of thorough history-taking and clinical examination cannot be overstated, as they are essential for the correct diagnosis in the majority of cases. Our study, like others, observed a higher prevalence of vascular malformations in males, with the peak incidence occurring in the 21-30year age group (Shen *et al*) [11]. The use of advanced imaging modalities such as ultrasound (US) with greyscale, Color Doppler, and spectral Doppler tracing, as well as CT angiography and magnetic resonance imaging (MRI) with digital subtraction, plays a crucial role in diagnosing these lesions, given the diagnostic challenges posed by their overlapping characteristics. In our study, Color Doppler, CT, and MRI were the primary diagnostic tools, which were invaluable for confirming the type of lesion, delineating anatomical boundaries, and planning surgical interventions (Behravesh *et al*) [12] [*Figure 1*]

Our data show that the majority of patients are between 10 and 30 years old, a demographic profile consistent with findings by Upton *et al* [3]. Their review of upper limb vascular malformations reported a similar age distribution, emphasizing that younger patients are commonly affected by these conditions. This supports our observation that vascular malformations predominantly impact younger to middle-aged individuals.Our study also reports pain as the most frequent symptom (46.7%) followed by painless swelling (23.3%). These symptoms align with the findings of Kunimoto *et al* [2] who identified pain and swelling as prevalent in vascular anomalies. This correlation underscores the importance of these symptoms in both diagnosis and management, as highlighted in their review.We utilized Doppler ultrasound (USG), MRI angiography, and CT angiography for imaging. Trop *et al* [5] emphasize the role of Doppler US in diagnosing venous malformations, which corroborates our practice of using Doppler ultrasound as a primary diagnostic tool. Additionally, Flors *et al* [13] support the use of MRI for detailed imaging of soft-tissue vascular malformations, consistent with our findings where 53.3% of patients underwent MRI angiography. This comprehensive imaging approach is critical for accurate diagnosis and treatment planning.In terms of treatment, our study shows that surgery alone is the most common recommendation (46.7%), with intralesional treatments and hybrid procedures following. This preference for surgical intervention aligns with findings by Rajput &Vishwakumar [6], who describe various surgical

management approaches for vascular anomalies. The use of intralesional sclerotherapy, as discussed by Lee *et al* [8], reflects our practice of employing less invasive methods in conjunction with or as an alternative to surgery, providing a balanced approach to treatment . Sclerotherapy, particularly with agents such as absolute ethanol, has proven effective for treating large, extensive VMs; however, it requires careful administration due to risks of nerve damage, skin necrosis, and potential systemic toxicity [8]. Commonly used sclerosants include 3% sodium tetradecyl sulfate (STS), polidocanol, and bleomycin [10]. Our study identifies recurrence (25%) as the most common complication, with seroma and hematoma being less frequent. Behravesh *et al* [12] discuss recurrence as a significant issue in venous malformations, aligning with our findings. Linh *et al* [14] also address the high recurrence rates following treatment for arteriovenous malformations, which reinforces the importance of effective management strategies to minimize recurrence. The recurrence rates observed in AVMs, particularly those in the head and neck region, underscore the need for a multidisciplinary approach involving interventional radiologists, plastic surgeons, orthopedic surgeons, and palliative care specialists, as highlighted by (Linh *et al* & Kiran SK *et al*,) [14-15]

Conclusion

In our study, Doppler USG/CT/MRI angiography was the main diagnostic tool. Surgery, intralesional treatment, and hybrid interventions were used for management with less morbidity and low recurrence rates. Their cure is often challenging and when not possible, treatment should aim at symptomatic control and improvement of patient's quality of life.

References

1. Madani H, Farrant J, Chhaya N, Anwar I, Marmery H, Platts A et al. Peripheral limb vascular malformations: an update of appropriate imaging and treatment options of a challenging condition. The Br J Radiol 2015;88(1047):20140406.
2. Kunimoto K, Yamamoto Y, Jinnin M. ISSVA classification of vascular anomalies and molecular biology. Int J Mol Sci 2022;23(4):2358.
3. Upton J, Coombs CJ, Mulliken JB, Burrows PE, Pap S. Vascular malformations of the upper limb: a review of 270 patients. J Hand Surg 1999;24(5):1019-35.
4. Queisser A, Seront E, Boon LM, Vikkula M. Genetic basis and therapies for vascular anomalies. Circ Res 2021;129(1):155-73.
5. Trop I, Dubois J, Guibaud L, Grignon A, Patriquin H, McCuaig C, et al. Softtissue venous malformations in pediatric and young adult patients: diagnosis with Doppler US. Radiology 1999;212(3):841-5.
6. Rajput DU, Vishwakumar CS. Compartmentalisation: A method of managing a large AVM of the scalp. JPRAS open 2018;15:56-60.
7. Jackson IT, Keskin M, Yavuzer R, Kelly CP. Compartmentalization of massive vascular malformations. Plastic Reconst surg 2005;115(1):10-21.
8. Lee BB, Kim DI, Huh S, Kim HH, Choo IW, Byun HS et al. . New experiences with absolute ethanol sclerotherapy in the management of a complex form of congenital venous malformation. J Vasc surg 2001;33(4):764-72.
9. Berenguer B, Burrows PE, Zurakowski D, Mulliken JB. Sclerotherapy of craniofacial venous malformations: complications and results. Plastic Reconst Surg 1999;104(1):1-1.
10. Cabrera J, Cabrera Jr J, Garcia-Olmedo MA. Sclerosants in microfoam: a new approach in angiology. Int Angiology 2001;20(4):322-9.
11. Shen Y, Wang Z, Yang X, Zheng L, Wen M, Han, et al. . Novel classification for simple peripheral arteriovenous malformations based on anatomic localization: Prevalence data from the tertiary referral center in China. Frontiers in Cardiovas Med 2022;9:935313-19.
12. Behravesh S, Yakes W, Gupta N, Naidu S, Chong BW, Khademhosseini A, et al. Venous malformations: clinical diagnosis and treatment. Cardiovasc Diagnosis Therapy 2016;6(6):557-69.
13. Flors L, Leiva-Salinas C, Maged IM, Norton PT, Matsumoto AH, Angle JF, et al. MR imaging of soft-tissue vascular malformations: diagnosis, classification, and therapy follow-up. Radiographics 2011(5):1321-40.
14. Linh DT, Khanh L, Ha NH, Son TT, Duc NM. Recurrence after treatment of arteriovenous malformation of the head and neck. AIMS Med Sci 2022;9(1):9-17.
15. Kiran SK, Shaji PC, Rajendran S, Gopinath TN, Jagdish J. Lower limb vascular anomalies and malformations with contrast-enhanced magnetic resonance angiography: A case series. Ind J Vascul Endovasr Surg 2021;8(Suppl 1):S72-4.