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Single Centre Surgical Experience with Congenital Vascular Malformations in North India

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Abstract

Introduction: Congenital Vascular Malformation (CVM) is a benign vascular tumour that results from arrest in the embryogenesis of vessel formation. Multimodal therapy is advocated in the management of CVM, from radiological embolization to surgical excision. It has been proposed that 'surgically accessible' lesions managed with complete excision may provide optimum results with least recurrence. We present our surgical experience in such patients at a single centre in North India.

Methods: Patients presenting with clinical CVM were evaluated with duplex ultrasound/ Magnetic Resonance Imaging scan to confirm diagnosis and assess extent. They were counselled about natural history of disease and treatment options. Localised lesions suitable for R0 resection with minimum residual disability/cosmetic disfigurement were counselled for excision. Records of patients undergoing surgery from March 2022 to October 2024 were accessed and analysed for demographic details, clinical presentation, any complications, histological diagnosis and recurrence.

Results: Eleven patients underwent excision biopsy of CVM at a new semi-urban setup over two years, out of total of 45 patients who presented in study period. A female preponderance was noted (8/11). The mean age of presentation was 29 years (SD 13.0, range 10-53). Suprapatellar bursa was breached inadvertently during excision of a vastus medialis CVM. One patient developed superficial surgical site infection. There were no recurrences over a median follow-up of 12 months (range 1-24 months).

Conclusion: Individualized decision for CVM is advocated. In the modern era of minimally invasive surgical procedure preference, complete surgical resection of CVM in selected patients can provide recurrence free treatment.

Keywords: Congenital vascular malformation; Peripheral vascular malformation; Arteriovenous malformation; Venous malformation; Hemangioma.

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INTRODUCTION

Congenital vascular malformation describes malformed vessels resulting from arrested development during various stages of embryogenesis.[1] CVMs are distinct from haemangioma in their origin and clinical presentation. A true haemangioma is characterized by rapid growth and slow regression, usually by 12 years; contrarily, CVMs never regress or disappear and need definitive management.[2,3] Hamburg classification divides CVM into arterial / venous / arteriovenous shunt / lymphatic / combined / capillary defects.[4] Mulliken has proposed classifying them into fast-flow lesions and slow-flow lesions based on the blood flow within the lesion.[3] Against the backdrop of treatment options like traditional surgical extirpation and percutaneous options, including sclerotherapy/embolization, the concept of a 'surgically accessible' lesion has emerged.[5] Excisional surgery may provide the best outcome for surgically accessible lesions, alone or combined with endovascular options. The nidus of the lesion needs to be eradicated for the possibility of a 'cure', so it is best suited for limited, localised lesions in current practice.

METHODS

The retrospective case record analysis of the Department of Cardiovascular Surgeries from March 2022 to October 2024 was done. The data was collected from operative records and the Hospital information system.

Patients with CVM on clinical examination confirmed with relevant imaging (duplex/ Computed Tomography / Magnetic Resonance Imaging) who underwent surgery were included in the study. Total of 45 patients presented to our OPD over the study period. Patients and their guardians were educated about the natural history of vascular malformations and various management modalities. They were evaluated with ultrasound/ duplex scan, followed by magnetic resonance imaging if needed, for confirmation of diagnosis, and assessment of extent and resectability. The planning for post-excision tension-free primary wound closure was done clinically. Those who were deemed candidates for R0 resection were counselled for surgery. All excised specimens were sent for histopathological examination. Patients were clinically followed up at 6 month interval post discharge for a period of 1 year, and advised annual follow-up thereafter. Patients with diffuse or multi-site CVM who were not amenable to R0 resection CVM at risk of significant disability after excision (lesion occupying entire flexor/extensor muscle compartment of a limb), and patients who needed extensive reconstruction (facial bones/entire nose) were counselled appropriately and referred for endovascular intervention.

RESULTS

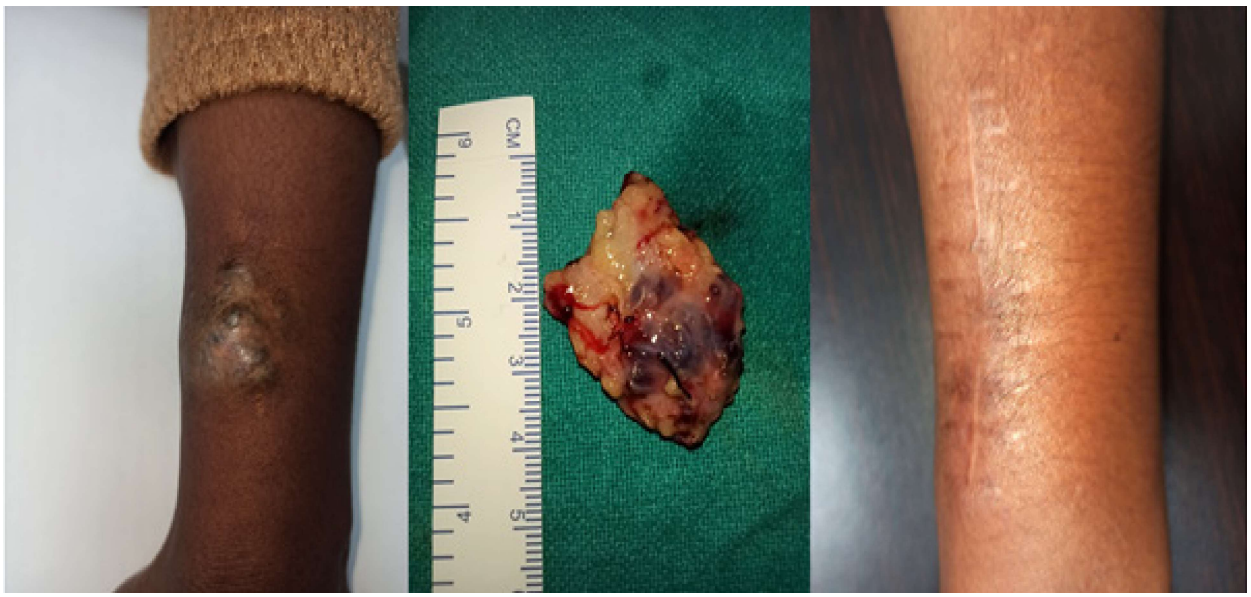

Fig. 1: Clinical photograph showing recurrent venous malformation preoperatively, excised specimen and postoperatively

Over the study period, 45 patients presented with CVM in our OPD. Mean age of presentation was 22.7 years (SD 10.2, range 1-53). 26 of them were females. 26 had lesions localised to extremities. Four out of forty five patients had high flow lesions; three had lymphatic defects. Four extensively involved a muscle compartment of a limb, with risk of significant disability after complete resection. Two cases were of superficial but diffuse CVM which made R0 resection difficult. Four cases extensively involved head and neck, which made resection/reconstruction too challenging for us at present. Unusual presentations included one patient with lesion involving right breast, and another with umbilical lesion. Two presented with recurrent lesions, one on leg, and another on wrist. The patient with wrist CVM had recurrence after 3 years of index surgery, performed elsewhere (Fig. 1).

Out of 45, 11 patients underwent surgery for CVM (Table 1). Three were males, and eight were females. The mean age was 29 years (SD 13.0, range 10-53). The size of the lesion varies from 1.2x1.0 cm to 5.8x4.9cm. All patients had the swelling for multiple years, and the chief reason for seeking medical care was a persistent increase in size and apprehension. One patient sought medical attention when the swelling grew fast during her first pregnancy. 11/45 underwent excision biopsy with no residual lesion clinically as well on pathological examination.

Demographic details, size, pathological diagnosis and complications are listed in Table 1.

Table 1: Clinical details and patient outcomes

Age (yrs)	Gender (M/F)	Chief Complaints	Site	Size (cm)	Pain (+/-)	Treatment	Pathological diagnosis	Comments
42	F	Right leg slow-growing lump since 5 years	Calf (Soleus)	2.2x1.9x2.8	+	Excision biopsy	Venous malformation	Developed superficial surgical site infection postoperatively
26	F	Recurrent lesion on the left wrist for 3 years	Left Wrist	2.4x0.7x2.3	+	Excision biopsy	Glomangioma	-
25	F	Right shoulder progressively enlarges swelling since 6 months	Right Shoulder	1.2x1.0x1.2	-	Excision biopsy	Venous malformation	Presented during pregnancy
10	F	Soft cystic swelling in the medial side of the right distal forearm for the last 6 years	Right forearm	3x2x1	-	Excision biopsy	Venous malformation	-
19	F	Swelling over right thigh for the last 6 years	Thigh (vastus medialis)	2.5x1.6x5.0	-	Excision biopsy	Arteriovenous malformation	-
33	F	Swelling in left forearm for last 8 years	Left forearm	3x2x1.1	-	Excision biopsy	Venous malformation	-
13	M	Swelling present over left lower thigh for 2 years	Left thigh (vastus inter medius)	5.8x4.9x2.5	-	Excision biopsy	Arteriovenous malformation	Suprapatellar bursa entered inadvertently; repaired
29	M	Swelling in front of neck x 4 y	Suprasternal notch	2x.7x1.8	-	Excision biopsy	Arteriovenous malformation	-

table cont.....

53	F	Lump below right knee since childhood	The medial aspect of the right upper leg	4.0x3.0x5.0	-	Excision biopsy	Venous malformation	Meningioma detected post-operatively
43	M	Left axillary swelling x 6 months	Left axilla	9.0x6.5x3.0	-	Excision biopsy	Lymphangioma	-
27	F	Swelling over left upper leg since ten y	The medial aspect of the left upper leg	5.3x4.2x3.0	+	Excision biopsy	Venous malformation	-

No intraoperative complications were noticed except the inadvertent opening of the suprapatellar bursa in Patient 7, a case of CVM in the vastus medialis (Fig. 2).

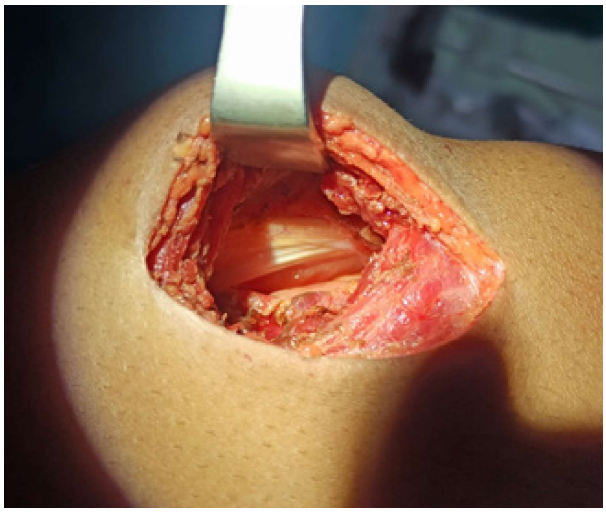

Fig. 2: Intraoperative photograph showing opened-up suprapatellar bursa during excision of CVM from vastus intermedius.

The bursa was closed in the same sitting with help of orthopaedics colleague. The patient was managed conservatively, and after limb physiotherapy, the patient had full range of knee joint movement.

Postoperative complications occurred in few patients. Patient 1 developed superficial surgical site infection following excision of soleal AVM. That patient was managed with debridement, dressing and secondary suturing. Patient 9 developed a headache with nausea after surgery and, in a few hours, altered consciousness (GCS: E2V1M5). On a detailed systematic workup, a right parieto-occipital meningioma with no signs of raised intra-cranial pressure was noticed. Her sensorium improved spontaneously over 24 hours. The patient was advised excision of meningioma. The authors did not find any syndromic association in this patient.

The patient with excision of soleal CVM and SSI developed mild swelling of the foot. One patient, after resection of AVM over the medial aspect of the upper leg, complained of a small area of anaesthesia caudal to the incision. This decreased over time, but a residual area remains with reduced sensation.

No other patient-reported any complications, and no syndromic association was noted. There have been no recurrences over median follow-up of 12 months (1-24 months).

DISCUSSION

CVM often presents as long-standing swelling, sometimes associated with pain. Due to their rarity and vascular nature, diagnosis is delayed and patients have various misbeliefs. However, if patients are appropriately selected and counselled, the surgery rewards both the surgeon and the patient.

We, in this case series analysis, present our experience with the CVM in our institute. Patient selection is important considering that we currently lack comprehensive endovascular services as well as reconstructive surgical backup. We put emphasis on patient education about CVM, suggesting surgery only when it seems to be the best course of action as it is our belief that small CVM do not always need treatment. Many young patients are suggested deferred surgery, allowing them to mature and have a say in management protocol. Our results largely conform along with the published literature.

Arasakumar et al.[6] describe that peripheral AVMs were treated mostly by open surgery in their study of records from NHS Hospitals in England from 2012 to 2018. However, there was a gradual increase in the number of percutaneous procedures.

Another case series analysis from 2014 to 2018 in a tertiary centre in South India reported 13 patients managed with various modalities such as embolization and medical treatment.[7] 7/13 lesions were extra-truncal, 7/13 patients had associated pain. The female-to-male ratio was 2:11, and the mean age of presentation was 23 years. Their report included large, diffuse malformations also, and they reported high recurrence rates. Another study from New Zealand by Visser et al. described their surgical experience in 53 patients (22 males, 31 females, mean age of 29 years) with a recurrence rate of 8.7% over an average follow-up period of 54 months.[8]

Liu et al. have reported a lower recurrence rate and a longer time to recurrence concerning embolization alone in a study of 272 patients with arteriovenous malformations.[9]

This study's limitation is that it is a single institutional study with a small sample size. Moreover, selection bias exists as we could not offer surgical treatment to all consecutive patients presenting to us. To evaluate true recurrence rates in our sample, a longer follow-up period would be needed.

CVM present in all shapes and forms. Successful treatment of CVM involves knowledge about natural history of disease and tailoring treatment to each patient for least recurrence and better patient satisfaction. We offer excision in patients with localized, surgically accessible lesions, to ensure lower recurrence rates.

CONCLUSION

Endovascular treatment is the preferred modality of treatment at present for most vascular diseases. CVM may have diffuse or multifocal presentation. They may also extensively involve body compartments making excision non-feasible. Complications like extensive growth or ulceration may mandate debulking surgery, with possible need for reconstruction. Among all these situations, excision of localized CVM in selected patients remains a safe procedure with minimal complications that offers the possibility of a 'cure'.

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