18 Emergencies in Vascular Malformations

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18.1 Introduction

The aim of this chapter is to present an overview of the different emergency presentations of various vascular malformations including identification, clinical features, diagnostic criteria as well as management principles.

Venous malformation (VM) is one of the most common vascular malformations and is a low flow variety. It grows in proportion with the body, exhibiting lifelong growth, and does not regress spontaneously [1]. Other low flow malformations include lymphatic malformations (LM) and capillary malformations (CM). The high-flow ones are termed as Arteriovenous malformations (AVM) which consist of multiple arteriovenous shunting within a nidus which consists of a capillary network [2].

Clinical presentation of vascular malformations is extremely variable and ranges from asymptomatic spots of simple visual concern to lesions with high blood flow or located in critical sites that may be life-threatening [3]. In this chapter, we shall focus on the emergency or critical clinical presentations of various vascular malformations.

18.2 Capillary Malformations

Although these are commonly recognized as port-wine stains, they are sometimes a part of syndromes like Sturge–Weber syndrome and can affect the eyes as well as the brain. The most significant clinical ophthalmic manifestation is glaucoma, which should be recognized and treated early. However, the most common manifestation is increased choroidal vascularization which is recognized by a characteristic image at the back of the eye known as "tomato ketchup." This lesion is usually asymptomatic in childhood but can lead to retinal detachment in adult life [4].

Brain involvement or leptomeningeal angiomatosis usually occurs on the same side as the capillary malformation and is evident clinically as epileptic symptoms with focal tonic-clonic seizures on the contralateral side of the body, with onset during the first year of life. Response to treatment is variable and most seizures usually become resistant to antiepileptic drugs, leading to slow progressive hemiparesis [5].

18.3 Lymphatic Malformations

Traditionally described as lymphangioma or cystic hygroma, these lymphatic malformations (LM) most frequently involve the head and neck region and present as soft, easily compressible masses

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(Figure) with thin overlying skin that may swell in dependent positions or when venous pressures increase (crying or Valsalva) [6]. Bleeding within the cyst or a mixed veno-lymphatic malformation may result in blue discoloration of the overlying skin. When they spread, cervical lesions may compress the pharynx or trachea when the mediastinum is involved and sudden growth of a cervical lymphatic malformation may be an emergency because the airways can be compromised, sometimes leading to respiratory distress. Such sudden growth can be triggered by accidental injury, sclerotherapy, bleeding, or infection [6].

The most common treatment option for LM is the use of sclerosants using various agents like absolute ethanol, Sodium Tetradecyl Sulfate (STS), or bleomycin These agents cause irreversible damage to the endothelium, inducing local inflammation and ultimately leading to fibrosis.

Multiple sessions are often required to achieve regression of the lesion. Swelling, a consistent consequence of sclerotherapy, can cause upper airway obstruction requiring observation in the intensive care unit, intubation, and sometimes even a tracheostomy [7].

The only potentially curative modality for LM is surgical resection. The goals of resection focus on gross debulking of defined anatomic field, limiting blood loss, and minimizing damage to surrounding structures. It is vital to remember that complete or radical resection is typically not without damage to surrounding normal structures especially nerves [8]. Hence, over time relapse may occur, which can be dealt with by sclerotherapy.

18.4 Venous Malformations

Venous malformations (VM) have low blood flow and are categorized as either superficial or deep, and as localized, multicentric, or diffuse [9]. The appearance of most superficial ones is purple color; the subcutaneously located or mucosal ones appear more bluish or greenish whereas the deeper intramuscular ones may appear as ill-defined swelling with normal overlying skin. Mucosal VM especially present with bleeding on minor trauma (Fig. 18.1).

Within the digestive tract, the most common site of the VM is in the small intestine which occasionally may be asymptomatic. Intestinal location of VMs can be associated with severe complications ranging from chronic anemia to acute abdominal conditions due to massive gastrointestinal bleeding (hematemesis or melena), intussusception, or volvulus that ultimately require an emergency surgery [10]. Such gastrointestinal VM may be a part of the Blue Rubber Bleb Naevus Syndrome in which there are associated multiple cutaneous VM which are blue in color and rubbery in consistency.

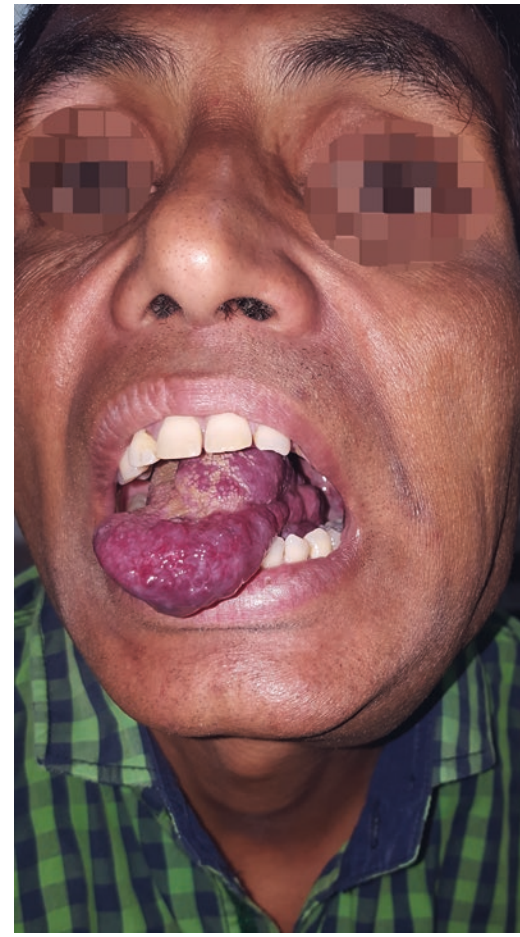

Fig. 18.1 Vascular malformation of tongue causing bleeding

Sudden increase in the size of VM is typical of bleeding within the VM. Trauma or accidental injury to subcutaneous or intramuscular VM can lead to hematoma or neurologic symptoms due to adjacent nerve injury or compression.

Apart from bleeding, the other major complication associated with VM, especially extensive VM or syndromes like Klippel–Trenaunay Syndrome (KTS in Fig. 18.2) is a coagulation disorder called Localized Intravascular Coagulation (LIC) [11]. Localized intravascular coagulopathy occurs due to stasis within the dilated and distorted low flow vessels with the formation of thrombin and subsequent conversion of fibrinogen to fibrin, which is followed by fibrinolysis and evidence of fibrin degradation products like d-dimer.

Newly formed microthrombi in LIC bind to intravascular elementary calcium deposits and form pathognomonic stone-like structures called "phleboliths." These phleboliths can often be palpated in patients with superficial VM, whereas in deep VM, they can be visualized on plain radiography or T-2 weighted MRI. The presence of phleboliths may represent indication for anticoagulation especially when the accompanying lesion is large and extensive [12].

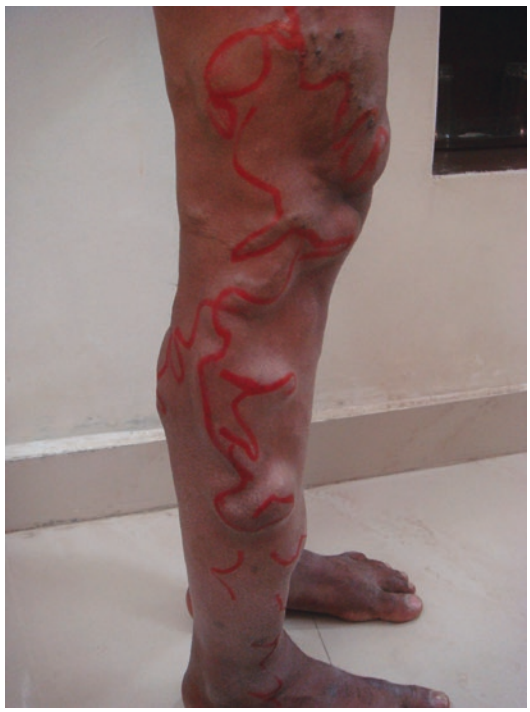

Fig. 18.2 Klippel–Trenaunay syndrome

LIC is of important clinical concern due to the potential for more serious thrombo-embolic events, including superficial thrombosis, deep venous thrombosis or pulmonary embolism as well as thrombo-hemorrhagic Disseminated Intravascular Coagulation (DIC) with life-threatening hemorrhage.

The conversion of LIC to DIC is marked by the consumption of platelets and factors of coagulation. Increase in prothrombin time and decrease in coagulation factor V are the earliest blood test findings. A number of events such as sclerotherapy, surgical resection, bone fracture, prolonged immobilization, and pregnancy or menstruation are known to trigger the conversion of the LIC to DIC, with hemorrhage related to consumption of coagulation factors and multiorgan failure related to disseminated microvascular thrombosis [13].

Additionally, low fibrinogen level and high d-Dimer reflects high consumption due to clotting associated with high fibrinolysis and increased risk for bleeding and requires preventive management by low molecular weight heparin (LMWH) which also helps to treat the pain caused by LIC and to prevent decompensation of severe LIC to DIC. Traditionally pain associated with VMs was considered as an inevitable consequence of the pathogenesis and progression of VM. However, recently the pain in the VM has been found to have a close relationship with underlying coagulopathy to cause LIC in many VM patients [14].

A hypercoagulable status is commonly associated with VMs and may present with chronic intermittent superficial thrombosis, Deep vein Thrombosis (DVT), or Pulmonary Embolism. Therefore, a thrombotic risk profile to evaluate a hypercoagulable state should be performed in all VM patients as a routine part of diagnostic evaluation prior to surgery or sclerotherapy, a detailed coagulation profile is critical in order to identify those patients at increased risk of hemorrhage due to an impaired primary hemostasis.

18.5 AV Malformations

AVM is a high-flow malformation with multiple arteriovenous shunting within a nidus which consists of a capillary network. The commonest sites are intracranial followed by extracranial head and neck, extremity, trunk, and visceral [14]. The clinical presentation ranges from an asymptomatic mass to cardiac failure. Bleeding occurs more frequently with AVM than with other vascular malformations, while other presenting symptoms include pain, ulcer, ischemic steal, or skin changes of venous hypertension (Figs. 18.3, 18.4, 18.5 and 18.6).

The four stages of AVM as proposed by Schobinger are as under [1]:

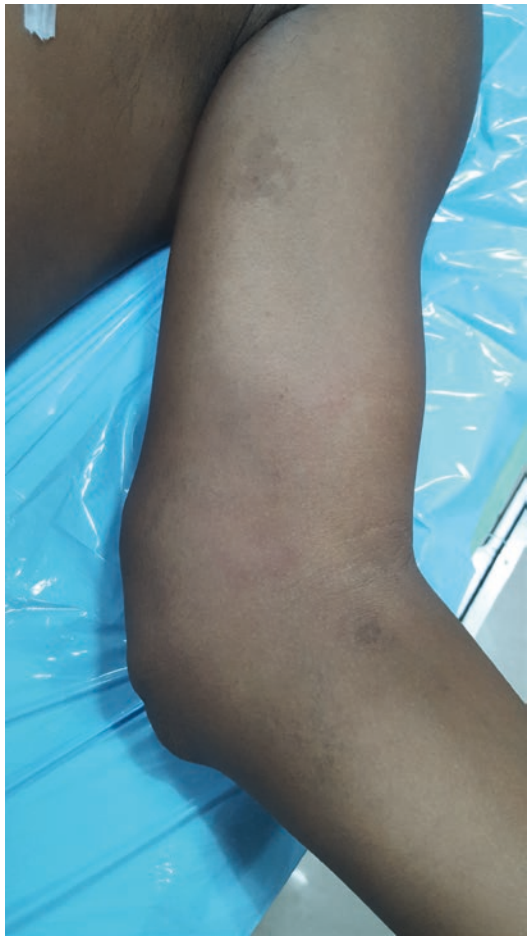

Fig. 18.3 Elbow AVM causing pain

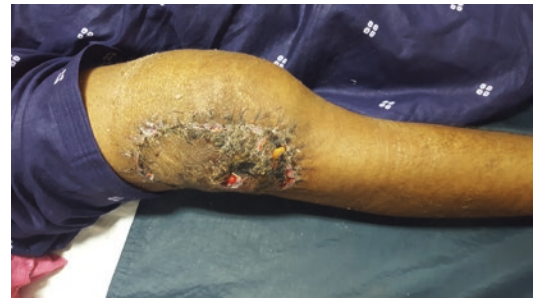

Fig. 18.4 AVM over knee with bleeding. Note incision for attempted but failed surgical treatment

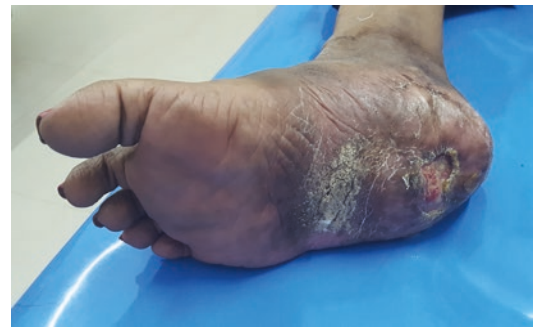

Fig. 18.5 Foot AVM causing ulcer

- Stage 1: *Quiescent*. The malformation is apparently stable and does not grow. This phase is transient in almost all cases, but it can last for decades.
- Stage 2: *Growing*. Spontaneously, or after physical or hormonal changes, the AVM becomes larger and the growth rate is unpredictable.
- Stage 3: *Symptomatic*. Growth brings the AVM close to the skin or mucosa, nerves, and major vessels. This can cause ulceration, bleeding due to rupture, or lead to pain due to irritation of nerves.
- Stage 4: *Decompensating*. High-flow diversion of blood through the AVM can which can lead to the development of dilated cardiomyopathy and, ultimately, heart failure.

Specific emergencies related to the location of AVM include the following:

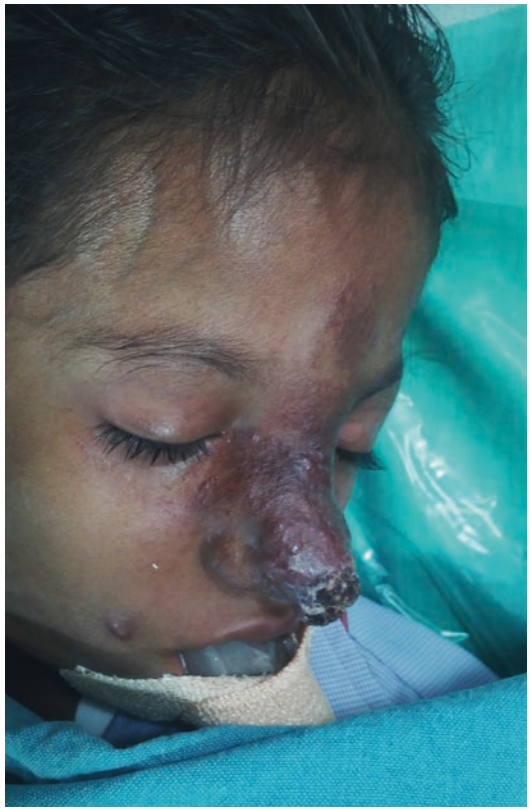

Fig. 18.6 Nose AVM presented with massive bleeding

1. *Brain and Spinal AVM*: Hemorrhage is the commonest presentation of brain AVM, followed by seizures and headaches. The overall risk of hemorrhage for brain AVM is 2–4% per year. Larger nidus, deep infratentorial location, and deep vein drainage are independent predictors of increased risk of hemorrhage. Also, ruptured brain AVMs have higher rates of rebleeding within the first year of presentation [15].

   The current treatment options include conservative management, surgical resection, stereotactic radiosurgery, endovascular embolization, or combinations of these treatments (multimodal therapy) [16].

   Spinal AVMs most frequently occur in the thoracolumbar region and progressive congestive myelopathy is the predominant clinical presentation. However, cervical lesions may present with subarachnoid hemorrhage [17].

2. *Pulmonary AVM*: Pulmonary arteriovenous malformations are abnormal direct communications between pulmonary arteries and pulmonary veins without the interposition of a capillary bed. Approximately 80–90% of patients presenting with PAVMs eventually present as Hereditary Hemorrhagic Telangiectasia (HHT) or Rendu–Osler–Weber Syndrome, whereas the remaining are sporadic cases [18].

   HHT may be clinically diagnosed on the basis of the Curacao criteria [19]. Three criteria are thus needed among the following:

   - Multiple mucocutaneous telangiectases
   - Spontaneous and recurrent epistaxis
   - Visceral involvement
   - A family first-degree history of HHT

   Often, thrombosis develops within the dilated venous sacs. In addition, there is a proliferation of anaerobic bacteria due to the right to left extracardiac shunt. This leads to paradoxical embolism and often leads to thromboembolic stroke or cerebral abscess events. Adams et al. reported that transient ischemic attacks and lacunar brain strokes are usually the first clinical manifestation of pulmonary embolism resulting from PAVM [20]. Yet a brain abscess is the most serious neurological complication of PAVM and occurs in 5–10% of patients with PAVM [21]. The reason is that right to left shunting in the lungs enables bacteria to avoid the filtering effect of pulmonary capillaries and consequently, the brain becomes the first and the most frequently affected target for bacterial emboli [22].

   Hemoptysis or hemothorax due to spontaneous rupture of a fistula sac is rare and usually occurs in pregnancy. This can often lead to a life-threatening situation with hypotension and shock and is diagnosed by X-ray or CT scan chest. Immediate therapeutic options include angiographic embolization. However, in presence of multiple AVMs, a posterolateral thoracotomy with wedge resection or lobectomy is the preferential choice because of low mortality and minimal recurrence rate [23].

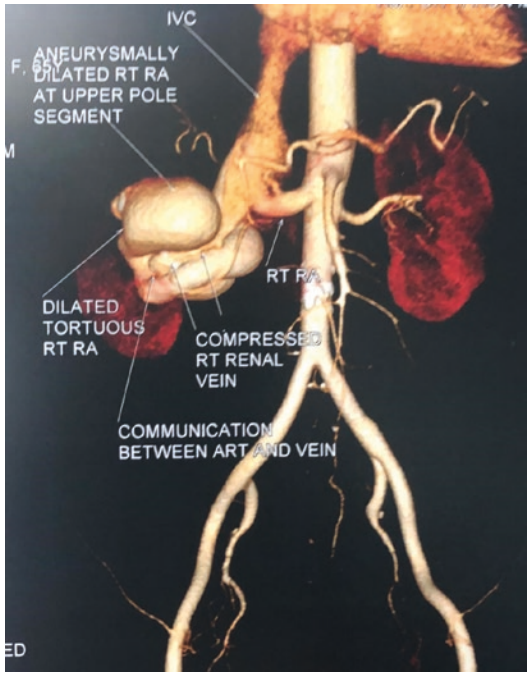

Fig. 18.7 CT angiography shows right Renal AVM with aneurysm

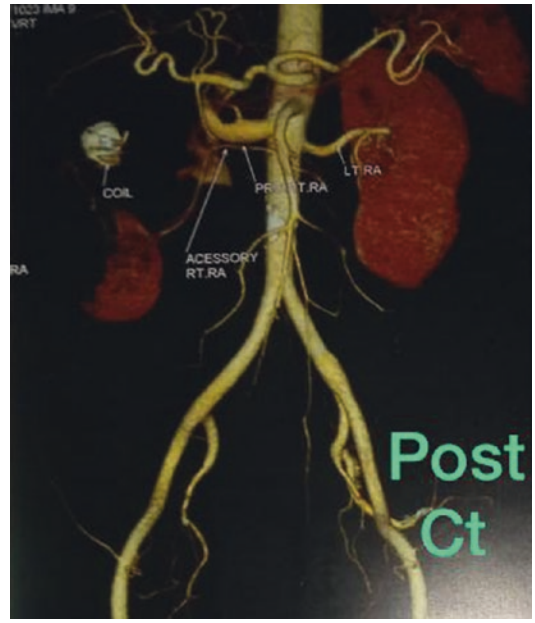

Fig. 18.8 Post-AVM embolisation CT angiography confirms total occlusion of renal AVM

The aim of transcatheter embolization is to occlude all the PAVM feeding arteries by a selective catheterization of pulmonary arteries by using a coaxial system, via a percutaneous femoral approach. Embolization materials used for this purpose include fibered coils, detachable coils, or the Amplatzer plug device. The anchor technique consists of coil blockage within a small collateral branch of the main feeding artery immediately upstream of the PAVM. This allows anchoring of the coil and thus creates a scaffold of the first coil, which permits the blockage of other subsequent devices and also prevents further accidental device mobilization and distal migration to the left circulation [24].

3. *Renal AVM*: Congenital renal AVM are uncommon and sometimes associated with HHT; Some of them present as congenital AV fistula which are high-flow shunts associated with venous aneurysm (Figs. 18.7 and 18.8). The majority of patients present with hematuria, flank pain, or perinephric hematoma. If asymptomatic, they may present later with arterial hypertension and the occurrence of high-output cardiac failure ranges from 5 to 30% in these patients [25].
4. *Pelvic AVMs* may produce severe pain, pelvic congestion, sexual dysfunction, and occasionally, high-output cardiac failure and hemorrhage. Although the primary blood supply of pelvic AVMs is from hypogastric arteries, there may be multiple feeding branches from the inferior mesenteric artery, middle sacral artery, lumbar artery, and femoral arteries. Because of the complexity of feeding branches, complete surgical treatment is often not possible, and recurrences after successful surgical intervention are common. Hence, endovascular embolization is the accepted therapeutic option for these patients [26].
5. *Uterine AVM*: Congenital AVM is rare, although acquired AVM is more commonly encountered due to uterine surgery, curetting, or infection (Fig. 18.9). Uterine AVMs frequently cause sudden massive bleeding in

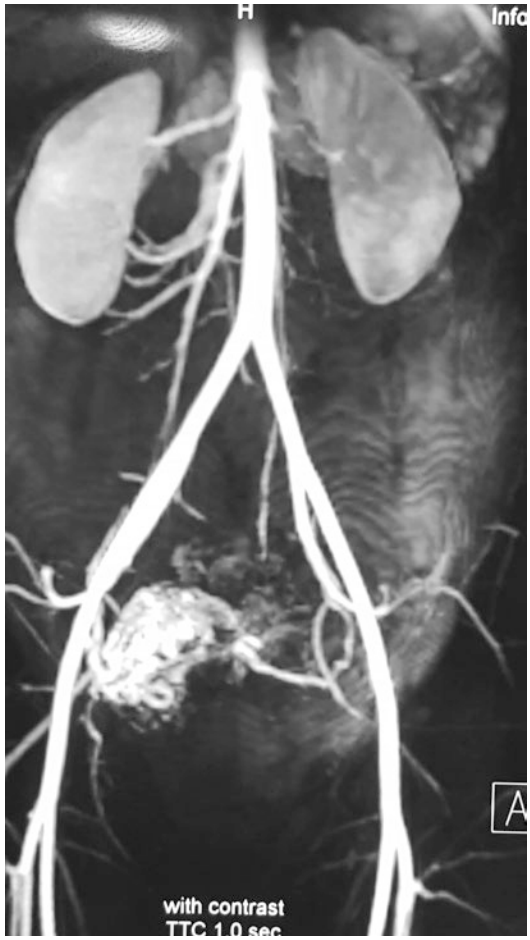

Fig. 18.9 MRI angiography shows uterine AVM

young patients, and in an unstable patient, appropriate diagnosis is important because emergent treatment with D&C can worsen the underlying condition, leading to profuse uterine hemorrhage, shock, and potentially hysterectomy [27]. Emergency angioembolization is the preferred treatment of choice and has excellent results, often avoiding the need for hysterectomy (Figs. 18.10 and 18.11).

6. *Gastrointestinal AVM*: AVM of the pancreas or common bile duct (CBD) is a vascular anomaly in which blood flows from the arterial system directly into the portal venous system without passing through the capillaries. This can lead to hemobilia or massive upper

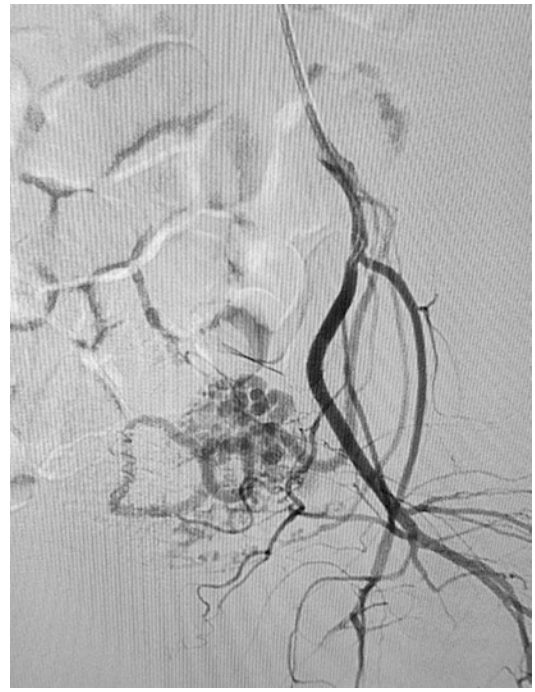

Fig. 18.10 Uterine AVM (before embolisation)

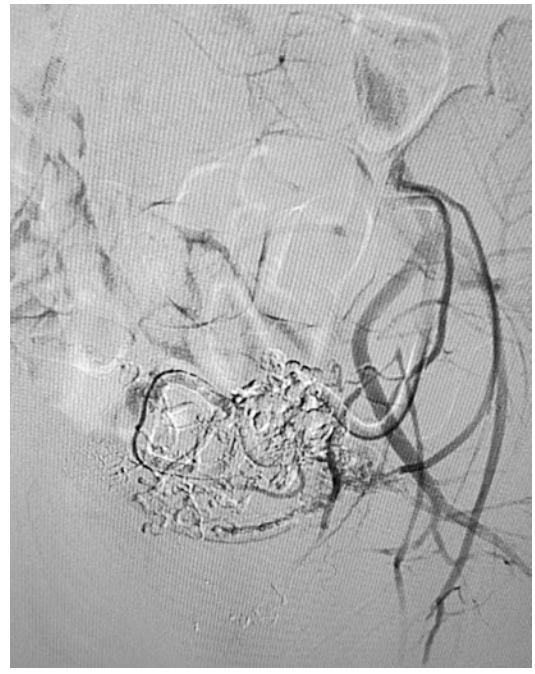

Fig. 18.11 Uterine AVM (after embolisation)

gastrointestinal (GI) bleeding and may be fatal. The treatment can be angioembolization or emergency laparotomy and pancreatoduodenectomy [28].

18.6 Treatment Principles of AV Malformations

These high-flow AV malformations are relatively rare and are far more difficult to treat. Surgical ligation of the feeding arteries of a high-flow AVM is ineffective and can be counterproductive as it often results in rapid recruitment of collaterals, which may render subsequent interventions ineffective.

Complete elimination of the nidus of an AVM is the only possibility for a total cure. However, this is often difficult to accomplish surgically. Debulking, incomplete resection, or ligation of arterial branches lead to recurrence of the malformation. Additionally, difficult surgical exposure and extensive blood loss are common during surgical resection of deep AVMs.

Successful endovascular embolization (embolotherapy) of the nidus of an AVM often requires super-selective catheterization of numerous arterial feeding branches. This is facilitated by using coaxial microcatheter systems. A 2-F to 3-F microcatheter is coaxially introduced through a 4-F to 5-F selective catheter and can be manipulated into the terminal feeding artery. Embolic agents are then delivered via the microcatheter, which is ideal for delivering liquid agents, particles, and small coils. The aim of embolization is to obliterate the nidus while simultaneously minimizing non-target embolization. For adequate embolization, it is recommended to use a combination of any of the three delivery routes: transarterial, transvenous, and direct puncture of the skin [29].

Selection of the appropriate agent for embolotherapy is as essential as correct patient selection. Various embolic materials available include: ethanol, N-butyl cyanoacrylate (NBCA), polyvinyl-alcohol (PVA) particles, ethylene vinyl alcohol copolymer (Onyx), and endovascular coils and vascular plugs [30]. Figures 18.12 and 18.13 show the use of glue or NBCA in an AVM of the lip, with complete obliteration of nidus. For another complex lesion in post-auricular region, embolisation using Onyx is demonstrated in Figs. 18.14 and 18.15.

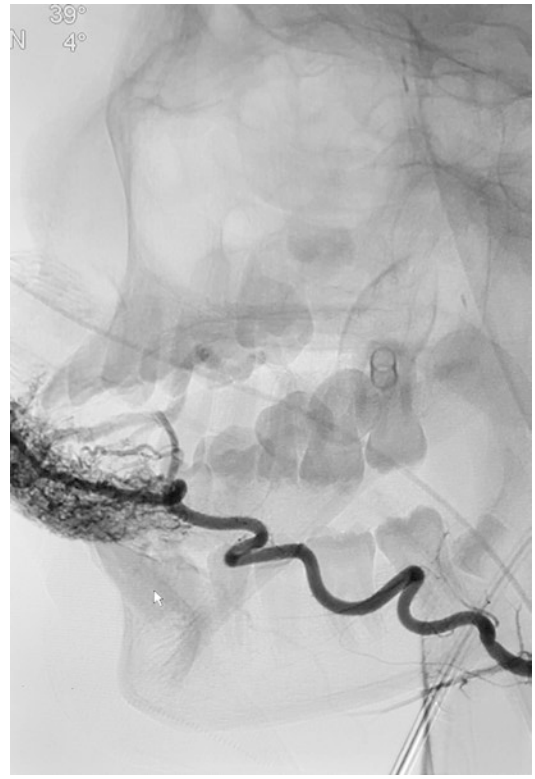

Fig. 18.12 Angiography shows nidus of lip AVM (before embolization)

Gorham-Stout syndrome (disappearing bone disease, phantom bone disease, diffuse skeletal hemangiomatosis) is a very rare syndrome characterized by multiple intraosseous vascular malformations inducing massive osteolysis. The truncal bones and upper extremities are most commonly affected. The vascular malformation may be localized or diffuse, and the degree of bone resorption is variable. Patients with this disorder often present during childhood with an antecedent history of minor trauma resulting in a pathologic fracture.

Localized intravascular coagulopathy (LIC) occurs due to stasis within these vessels with the formation of thrombin and subsequent conversion of fibrinogen to fibrin, which is followed by

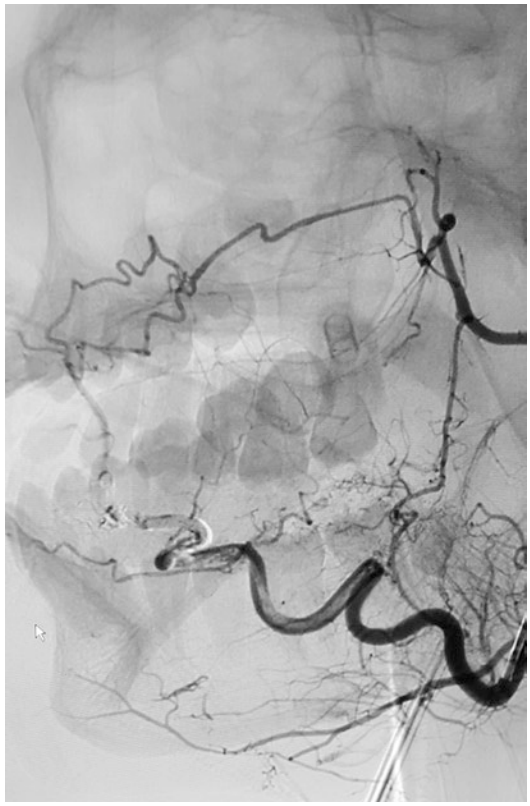

Fig. 18.13 Completion angiography shows obliteration of nidus of lip AVM (after glue embolisation)

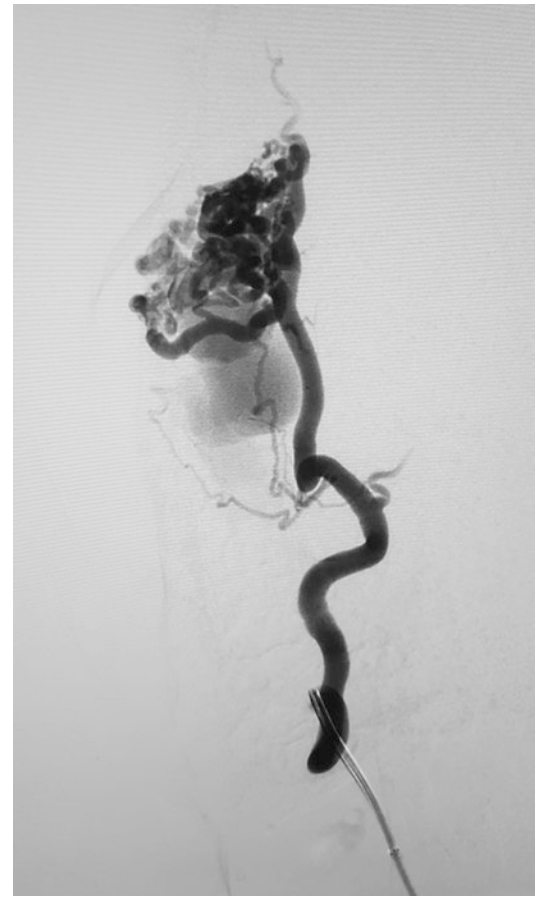

Fig. 18.14 Digital Subtraction Angiography shows nidus of ear AVM

fibrinolysis and evidence of fibrin degradation products like d-dimer LIC is of important clinical concern due to the potential for leading to more serious thrombo-embolic events, including STP, DVT, PE and the associated pulmonary hypertension, and thrombo-hemorrhagic DIC with life-threatening hemorrhage which can occur during or following surgical resection or sclerotherapy. Extensive CVM with large surface area, muscle involvement, and/or palpable phleboliths are strong predicting criteria for coagulation disorders associated with CVM. Assessment of the coagulation profile and D-dimer levels is indicated in patients with extensive CVMs. Coagulation profile and D-dimer levels are indicated in patients with extensive CVMs.

LIC as characterized by elevated D-dimer levels has been observed in approximately 40% of patients with CVMs. Patients with severe LIC would present with highly elevated D-dimer levels associated with low fibrinogen levels. Anticoagulation with LMWH can be used to treat the pain caused by LIC and to prevent decompensation of severe LIC to DIC.

AVM patients can present with pain, functional impairment, concerns about cosmetics, cutaneous and/or muscular ischemia, infection, and even ulceration(s) due to "steal phenomena" and subsequent diminished distal arterial blood flow.

If extensive, AVMs can lead to severe episodes of life-threatening hemorrhage, limb-threatening ischemia, and high-output cardiac failure due to large volume arteriovenous shunting. It may occasionally involve vital or critical structures like airways, eyes, or manifest by extension into the brain, perineum, genitals or intra-articular location (Hauert disease).

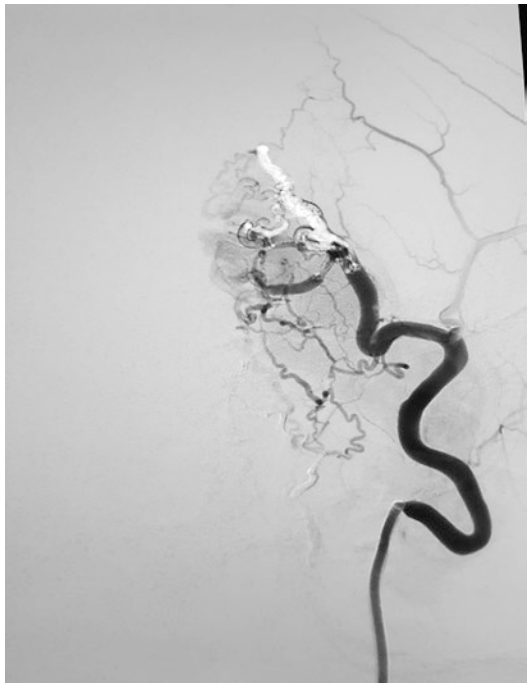

Fig. 18.15 Post-onyx embolization of ear AVM

In cases of trauma, secondary infection, abrupt hemorrhage of the lesions, or changes in hormonal levels can lead to pain, swelling, and even bleeding [13]. Venous malformations in parapharyngeal space, tongue, and soft palate may be accompanied by swallowing, speech, and airway problems.

18.7 Conclusion

Vascular malformations remain one of the most complex and ill understood diseases treated by vascular surgeons and interventionalists. A proper knowledge of emergency presentations of these infrequent disorders helps in deciding and delivering appropriate treatment.

Treatment of these vascular anomalies is challenging and often involves various therapeutic options. Multidisciplinary approach with full integration of open surgical and endovascular therapy has become the mainstay of treatment in the contemporary management of VMs.

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