Title: Review of Treatment Options for Arteriovenous Malformations.

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Manuscript Info

Abstract

Despite advances in the knowledge of vascular anomalies, arteriovenousmalformations (AVMs) remain an enigma & its management remains a challenge. Complete eradication of the nidus of an AVM is the only potential “cure.” But this however is often difficult. Single therapeutic modality is however unlikely to lead to a significant long-term reduction and combination therapy is increasingly considered to be the treatment of choice. Current treatment options consist of direct puncture embolization technique as an adjunct, it facilitates surgical resection of the lesion (kademani et al oralsurg oralmed oralpathol oralradiolendod, 2004). Other techniques used are resection, curettage, embolization, sclerotherapy, and even radiotherapy. The purpose of this study is to conduct a retrospective analysis of the clinical results of a contemporary management program for AVM.

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Introduction:

Vascular lesions include haemangiomas and vascular malformations. Among these Arteriovenous malformations (AVMs) are the dangerous group of vascular birthmarks formed by direct connection between an artery and a vein without intervening capillaries.

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AVMs have 31% prevalence in the maxillofacial skeleton. AVMs are usually latent during infancy and childhood and may enter an active phase in adolescence, present with catastrophic hemorrhage either spontaneously or if a biopsy or tooth extraction is done. They have an unpredictable course of causing facial asymmetry, loose teeth, tinnitus, headaches and bruits, and can be complicated by life-threatening conditions such as hemorrhage, high output heart failure, and ischemic necrosis. Patients are often concerned by the presence of intermittent bleeding. Although the mechanisms are poorly understood, infection, trauma, or hormonal changes in puberty and pregnancy are known can trigger the expansion of AVMs.

Management of these lesions is complex, because of their highest risk for excessive bleeding, propensity for rapid enlargement, high morbidity, and mortality which requires an integrated team approach including surgeons, interventional radiologists, and critical care intensivists. Advances in critical care, interventional radiology, and hypotensive anesthesia offer increased success in treating such patients, but morbidity and mortality are still found to be high.

Various techniques of management taken in to the study are
1. Medical Therapy
2. Embolotherapy- direct puncture embolization, superselective intraarterial embolization, trans venous, transfemoral, embolization through mental foramen
4. Lasers
5. Surgical- enbloc resection, curettage, resection and immediate replantation.
6. Conservative methods- buccal window approach ,toothpouch suture technique
7. Cryotherapy
8. Resection and immediate reconstruction
9. Multidisciplinary approach – embolization+ excision, sclerotherapy+ excision etc…,

Medical Therapy:
Steroids have become a mainstay in the treatment of proliferative vascular malformations. High doses of systemic or intra lesional steroids are the first line treatment, and a good response is observed in 30% of patients who used prednisolone at a dose of 20 to 30 mg/d for 2 weeks to 4 months. Intralesional triamcinolone acetonide (4 mg/mL) leads to increase in mast cells and regression of the tumor. The use of interferon alfa-2a has also been documented but some concern about its toxicity especially in children. Johann et al have stated that the disadvantage with intralesional steroids are adrenal suppression, weight gain, atrophy and necrosis.

Embolotherapy:
Embolization means the occlusion of a vessel by the introduction of a foreign material. Various agents used are-Absorbable: gelfoam, oxygel; Non-absorbable : 1. Particulate agents: silastic spheres, steel pellets.2. Injectable fluids: microfibrillar collagen, silicone rubber.3. Non particulate agents: platinum coils, detachable balloons. DSA (digital subtraction angiography) is used to diagnose AVM and also to block the feeding artery to reduce vascularity. Resection should be carried out within 5 to 7 days after embolization because of chances of recanalization of AVM with time and the embolization materials would be absorbed slowly, thus reducing its efficacy. Embolization should be considered as an adjunctive option because it just interrupts the main blood flow. Kademani et al, Liu et al said that for nonresectable lesions embolization alone can be done along with trans osseous direct puncture embolization. Carneiro et al says massive hemorrhage from AVMs can be managed with direct puncture embolization. Zhang et al says for intraosseous lesions near the mental foramen direct trans venous embolization through the mental foramen can be done.

Sclerotherapy:
Sclerosant injection initiates inflammatory reaction in the walls of AVMs and induces necrosis, and vascular block. Sclerotherapy is aimed at obliterating the lumen by fibrosis. several agents can be used as sclerosants: STS (sodium tetradecyl sulfate), ethanol, polidocanol, hypertonic saline, and sodium morrhuate. Johann et al said that sclerotherapy is safe, less invasive, with low risk of hemorrhage and cost effective. common complications are pain, swelling, redness and burning, Lee et al mentioned that these complications are considered acceptable when compared to its effect on quality of life. Most complications are the result of extravasation of the sclerosant. Using ultrasound guidance, the clinician can assess the site of entry in to the lesion, as well as decreasing the risk of perivascular or unintentional intra-arterial injection. Sclerotherapy is especially useful where: Excision is not
possible, There is a need to debulk the tumor before surgery or for Cosmetic reasons. Patient does not want to go for surgery. Kim et al. proposed that sclerotherapy should be considered as first line of treatment because of its effectiveness, fewer complications and low morbidity.

Rehman et al. advocated that foam sclerotherapy is more effective than the original sclerotherapy. Theddeus et al. mentioned that intralesional alcohol injection is an adjunctive treatment option for vascular lesions. Chimaoji proposed a simple technique of managing soft tissue AVMs in economically poor countries with the use of tobacco-pouch suture technique and combination of ligation and injection of sclerosing agents for medium sized lesions. Color Doppler guided ultrasound sclerotherapy (CDU) - Doppler flowmetry is useful in the initial categorization of patients and appear to contribute significantly to the clinical findings. CDU is noninvasive, relatively inexpensive and can be repeated as often as necessary, predicts the response of the lesion to sclerotherapy.

SOFT TISSUE AVM'S CAN BE MANAGED BY

Lasers:-
Xue et al. states that laser can be used for selective photothermolysis rather than nonselective tissue destruction. The following lasers are found to be effective: pulsed dye laser, Nd-YAG laser, argon laser, and carbon dioxide laser. Johann et al. says laser therapy is associated with skin atrophy, scarring, hyperpigmentation, depression of the skin at the site of therapy and high cost. Ales vesnaver et al. used Nd:YAG lasers for managing smaller vascular lesions and is less invasive, simple, less scarring, provides a bloodless field with minimal post-operative discomfort and pain. Miyazaki et al. used KTPlasers it is more effective because of its greater selectivity and specificity for hemoglobin.

Surgical Therapy:-
It was shown that surgical resection is an effective method for the treatment of AVMs. Kim et al. says surgical excision allows the complete removal of nidus with no recurrence but the only disadvantage is massive bleeding. Enblock resection is indicated for AVMs involving the lip, cheek, floor of the mouth and parotid gland compromising the esthetics. Segmental resection is recommended for the treatment of bone lesions. Bone reconstruction should be done after resection of intraosseous lesions. Surgical complications include: Early: Hemorrhage, airway compromise, hematoma, skin-necrosis, coagulopathy. Late: Restricted mouth opening, dysphagia and recurrence. Any delay from embolization to surgery exceeding 48 hrs may lead to greater risk of bleeding due to increased blood flow through collateral vessels feeding the lesion.

Conservative Management:-
It involves the extraction of teeth and removing the underlying lesion through the alveolar process and packing the mandible with oxidized cellulose. The procedure results only in the loss of the involved teeth and preserves the adjacent normal structures. This technique is found to be simple and safe.

Brusati et al. reported a modified technique for treatment of intraosseous AVMs. The technique consists of SIAE and piecemeal removal of the lesion through burred holes made in the cortex and packing the holes with bone wax to control hemorrhage curettage should be carried out 2 months later. This markedly reduced bleeding and favorably completed removal of the AVM. This is best performed in the presence of intact lingual and buccal cortex in order to ensure adequate curettage and a tight packing of the cavity. Hossein behnia et al. says superselective arterial embolization, followed by trans mandibular curettage is effective and less invasive for small intraosseous AVMs with favorable esthetics.

Cryosurgery:-
This technique has fallen into disfavor because it is associated with scarring and hyperpigmentation. Johann et al. , Hartmann et al reported minimal scar contracture, good hemostasis, and little discomfort for large vasoformative tumors.
Discussion:

Resection And Immediate Reconstruction:

Surgical treatment involves resection and replacement of the resected portion of the mandible after having removed its pathological content, this technique avoids the need for bone harvesting as well as maintaining the normal anatomical integrity. Schneider et al.\(^\text{19}\) and Yoshiga et al.\(^\text{20}\) say temporary segmental osteotomy and immediate replantation for large AVMs maintains the normal physiological matrix for favorable growth, development and maintains the anatomical integrity of bone.
**Multidisciplinary Approach:**

Weiliang Chen et al.² said that combination of treatment modalities show more success rate when compared to individual management techniques. Lee et al.²³,²⁴ said embolo/sclerotherapy should always be followed by surgical excision for complete removal of nidus of AVM.

Chen et al.²³ says compared to individual treatment modalities for managing AVMs in children, multidisciplinary approach is found to be more effective with less chances of recurrence.

Vascular abnormalities were originally classified in 1982 by Mulliken & Glowacki based on the lesions’ biological and pathological differences in to hemangiomas and vascular malformations.²⁶ Jackson et al. classified vascular anomalies as low-flow lesions-venous malformations (LFL), and high-flow lesions (HFL)-arteriovenous malformations. Angiography has traditionally been the gold standard for the precise evaluation and their characterization into HFL and LFL.¹⁴

AVMs are rare and they are commonly misdiagnosed as hemangiomas, and patients are often told that they will eventually involute, inspire AVMs do not regress and continue to expand with time. AVMs are thought to arise from errors in vascular morphogenesis. Histologically, AVMs are composed of numerous aberrant arteriovenous shunts associated with dilated capillary beds that are thought to be devoid of autoregulation. Vascular recruitment and collateralization contribute to progressive expansion of AVMs with the acquisition of feeding arteries and draining veins. The subsequent network creates a “nidus” of vascular ectasia with an inherent growth potential.¹

AVMs are present at birth, enlarge with growth, and do not undergo spontaneous regression. Trauma, infection, and hormonal changes are found to be the predisposing factors.²⁷ They display an unpredictable clinical course. It can present with asymptomatic birth mark, a palpable mass, warm pulsatile soft tissue mass with teeth loosening, the symptoms can be secondary to compression of the adjacent vital structures and they can be complicated by life threatening conditions such as hemorrhage, high output heart failure and ischemic necrosis.²² AVMs result in significant physical and psychological morbidity, soft tissue destruction, pain, and bleeding, with a dramatic reduction in quality of life.

Surgery, embolisation are the cornerstones of therapy which in turn requires accurate characterization of lesions including the location, size, extent, feeding and draining vessels along with detailed radiographic studies. Digital subtraction angiography (DSA) examination display the nidus of the AVM, feeding artery as well as investigate the site of bleeding.²⁸ Magnetic resonance imaging (MRI), CT have all been used to diagnose and characterize AVMs. CTA (computed tomographic angiography) has super ceded DSA as the primary imaging technique for diagnosis and characterization of vascular lesions, since CTA is noninvasive, provides sufficient anatomical information regarding the AVM and its relationship to adjacent soft tissues and bone and enables 3D reconstruction of the lesion. The lesions became more conspicuous and border definition improved significantly with CBCT. VR (volume reconstruction) techniques combined with subtraction, cutting and revolving were particularly useful for showing the organization of the nidus as well as feeding vessels, including small feeding arteries that were not appreciated using other techniques.²⁸

The philosophy underpinning our management is based on “prescribing a treatment that is not worse than the original disease” and considering the patient’s wishes being of prime importance. Underlined by our ultimate goal of complete removal of the nidus followed by immediate reconstruction to restore form and function. When this is not achieved, recurrence is almost guaranteed and may lead to gradual worsening.¹⁸ Various modalities of treating AVM are proposed which include: Medical therapy, Embolo-therapy, Superselective intra-arterial embolization (SIAE), Sclerotherapy, Lasers, radiotherapy, Cryosurgery, bone wax packing (BWP) and curettage. Surgical therapy, preoperative embolization followed by surgical resection, multidisciplinary approach etc... The decision making for choosing treatment option and prognosis depend on the anatomic location, as well as the angioarchitecture.

Based on x-ray and angiographic features the central AVMs in jaws (intraosseous) were divided into 5 types.

Type I AVMs appeared as honeycombed radiolucency on radiography and as diffusely interwoven vessels on angiography.

Type II AVM could be defined as an arteriovenous fistula that consisted of multiple arterial shunts in the wall of a single huge vein.

Type III AVMs comprise compartments of type I and type II fistulas.
Type IV and type V AVMs, have complex angioarchitecture with extensive involvement of the jaw\(^{29}\).

For type I AVMs superselective arterial embolization, followed by complete curettage. For type II AVMs, Filling of the veins with occlusive agents will close all shunts on the wall. Type III AVMs complete obliteration of the complex nidus is mandatory. It may need, multiple procedures of intranidus/intravenous embolization. Type IV and type VAVMs can rarely be controlled by embolization alone surgical resection is commonly inevitable\(^{32}\). Absolute indications for Treatment-Hemorrhage, ischemic complication, arterial insufficiency, chronic venous insufficiency, high output cardiac failure, lesions involving vital structures effecting vision, hearing, breathing, swallowing. Relative indications for Treatment-Various signs and symptoms affecting quality of life, pain, functional impairment, severe cosmetic deformity.

various treatment options are studied in detail and the appropriate treatment options are given in a flow chart

**Soft Tissue Avms Can Be Managed By:-**

Embollization-it is indicated to reduce the flow rate in high flow lesions. For soft tissues AVMs compartmentalization and injection of sclerozing agents should be done\(^{30}\). Embolization alone can be performed in non resectable cases\(^1\). It is done through trans arterial, transvenous, direct puncture, direct puncture through the mental foramen. complete removal of the lesion is not possible it only acts as an adjunct to other treatments and improves the quality of life in cases of life threatening hemorrhage\(^{31}\). First inject platinum coils to reduce the blood flow then perform the ethanol embolization either through the arteries or direct puncture. it does not eliminate the risk of recurrence because of appearance of collateral circulation. Preoperative embolization should be performed 24-48 hrs prior to surgery it facilitates the surgical resection of the lesion with less bleeding within 48hrs to 2weeks\(^{19}\).

SIAE (super selective intra-arterial embolization)-The seldinger intubation technique was used through the femoral artery. The catheter was passed into the common carotid artery; angiograph was taken through the external carotid artery and internal carotid artery. AVM in the oral and maxillofacial region the origins of each feeding artery (eg, inferior alveolar artery, lingual artery, internal maxillary artery, and maxillary artery) were embolized individually. The embolization materials used included polyvinyl alcohol particles, acrylic glue, silky thread (10 to 15 cm), iodine oil, and metal loop.

Tobacco pouch suture technique-alone it can be used for  ligating  small lesions with in the soft tissues and a combination of injection of 10% lukewarm saline into large tumors can be effectice\(^{30}\).A curved needle and chromic catgut 1/0 are used. After penetrating the overlying tissue or mucosa at the border of the tumor, the needle was advanced in intervals of about 1 cm so as to encircle the tumor. One creates a loop, which encircles the vascular structure. The two ends of the thread, which are about 1 cm apart, are tied tightly together. As a result, the afferent vessels are closed and the blood supply interrupted. In large tumors, we can tie ligatures across the opposite sides, which help to complete the interruption of the vascular channels and stop the blood circulation in the tumor by tightening the nose that encircled them and injecting a sclerosant (10% lukewarm saline 2–3 ml).

**Sclerotherapy:-**

It can be used in deep seated inaccessible lesions where they envelop vital structures, such as the facial nerve, where a less-invasive treatment is required. When injected preoperatively, it helps to reduce surgical blood loss and to delineate the surgical extent of resection\(^{20,21,22}\). Sclerotherapy is most preferred because of its advantages of no external scarring, little morbidity and few complications\(^{18}\). a 24-gauge needle was used in a z-track to prevent ooze from the site of injection. sclerosant should be injected after drawing up blood to confirm the tip in the lumen of the lesion, procedure was stopped if patient complains of undue pain or excessive discomfort. Injection of sclerosants into the AVM should be done and the dose is based on the volume of AVMs and patient age; the minimal dose was 4 mL; the maximal dosage was 10 mL. Multiple injections were given on large AVMs. External compress was maintained after each injection. The injection was repeated every 2 weeks, 4 to 6 times. Sclerotherapy is excluded for those involving the vital structures. Incidence of soft tissue injury and neuropathy is more but despite of its disadvantages it is widely used.
Laser therapy:-
The successful application of several types of lasers has been reported for smaller hemangiomas and vascular malformations. due to the limited tissue penetration of laser energy makes radical regression of a voluminous vascular lesion difficult using conventional noncontact irradiation 19,25,28.

Cryotherapy:-
it is also indicated for treating malformations around salivary gland orifices, to prevent the risk of posttreatment stenosis27. Nerves may be damaged irreversibly by photocoagulation. Therefore authors advise the employment of cryosurgery as an alternative in areas adjacent to the nerves not to cause irreversible damage, the paresthesia observed in some cases is only transient.

Surgical Management:-
Surgical management is indicated as a definitive treatment of any type of AVM and complete excision of the nidus of AVMs is possible with least chances of recurrence 9,12,13,19. complete resection was used in the treatment of AVM in the soft tissue, such as the lip and cheek.

Preoperative Embolization and Radical Resection:-
this is the modern and conventional approach mostly used used for unrespectable and recurrent AVMs 17,20. This combination is considered to be the best possible approach with least possible complications and less recurrence 33.

Intraosseous AVM’s Can Be Managed By:-

Embolization:-
Intral esional embolising materials are directly injected in to the lesion to encourage cessation of flow, following angiography. it delineates the extent of the lesion. Trans osseous direct puncture of the lesion renders the lesion hemostatic by introducing thrombogenic or sclerosing agents 23.

Conservative Management:-
Preoperative embolization with bone wax packing and curettage. it is indicated in cases of small to medium sized AVMs.

For intraosseous AVM highly selective embolization performed no more than 24–48 h before the surgical intervention. The feeding Vessels-External carotid, lingual and facial arteries are identified and prepared for clamping. the buccal cortex of the mandible is exposed and a burr of 4 mm in diameter is used to make burr holes. Rapid curettage with a small spoon followed by oxidated cellulose packing in to the holes arrests the bleeding. In this way, a series of holes 1-2cm apart allows one to treat the whole malformation, while limiting blood loss and respecting both mandibular continuity and tooth integrity. The procedure leaves the mandible looking like a piece of Swiss cheese 35.

Surgical management–surgery is not indicated in children due to proximity to vital structures and risk of future growth potential 23. Surgery is the toughest treatment option because of vascularity of the lesion, lack of distinct margins, recurrence and cosmetic deformity. it is only indicated for localized lesions. AVMs involving the jaws can be removed completely.

Trans Mandibular Curettage:-
Trans mandibular curettage via proximal osteotomy without removal of the segment is effective and less invasive to treat small AVMs of mandible with minimal blood loss, preservation of bone and periosteum, more favorable postoperative esthetics and better function.

Resection and Immediate Replantation:-
After embolization the anomalous vessels are explored and ligated followed by sequential mandibulectomy. The osteotomized segment is removed temporarily and is hollowed out with rotating hand piece and bur. Numerous holes were made in the buccal and lingual aspects to aid in revascularization. Bone grafting material is placed inside and the segment kept back with miniplates 15. it can be used in cases of bilateral and collaterally communicating malformation with numerous feeding arteries 16 and for AVMs characteristically displaying intense high-flow vascular blush with early venous shunting. Hypertrophy of feeding vessels 17.
Multidisciplinary Approach:--
this approach is indicated for extensively large arteriovenous malformations. Any combination of treatment options based on the lesion can be chosen. Ulrike ernemann et al 15 mentioned that complete radical resection is achieved by preoperative embolization, intraoperative intervention and resection followed by early postoperative angiographic assessment and resection of residual pathological tissue in a second stage.

Conclusion:--
Individualized treatment protocol should be made based on the condition of the patient and the technical availability to prevent risk of debilitating hemorrhage, decrease frequency of progressive neurological deficit, reduce recurrence. Most often multidisciplinary approach will achieve the best results when well planned and implemented. Advances in the treatment make multidisciplinary approach safer and more effective compared to individual treatment modality.

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