

Management Of Arterio Venous Vascular Malformation Masquerading As A Mucocele Using Sclerotherapy-Review Of Literature And A Case Report.

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ABSTRACT :The management of vascular anomalies is an extremely challenging area and is as divergent as the nature of the lesions. Traditionally, embolization with the resection of the lesion has been used. The purpose of this report is to present a case treated successfully using sclerosing solution injections alone.

METHODS: Management of the arterio venous malformation using Intra-lesional injections of sodium tetradecyl sulfate to the lesion is discussed. The procedure was performed two times at two weeks interval. Imaging was performed using Dopplers ultra sonography.

RESULTS: Complete resolution of the lesion was found following sclerotherapy.

CONCLUSIONS: Conservative interventional management using Intra lesional injection of sclerosing solution was successful in treating vascular anomaly.

KEYWORDS - Arterio-venous malformation, Sclerotherapy

I. INTRODUCTION

Vascular malformations remain difficult both diagnostically and therapeutically despite continued efforts over the decades. This is chiefly due to their variety, with a wide range of clinical presentations from a simple birthmark to a life-threatening condition containing embryonic remnants of a developmental defect. Further the condition has been complicated by various factors like an unpredictable clinical course, confusing nomenclature, erratic response to treatment, frequent recurrence, and high morbidity following conventional treatment.[1-4] Vascular lesions of the maxillofacial region are classified by Mulliken and Glowacki (1982) as either: 1) Hemangiomas or 2) vascular malformations.[1] Hemangiomas are the most common cutaneous tumor of infancy and demonstrate rapid growth followed by a slow spontaneous involution or regression within five to seven years. While vascular malformations enlarge with the growth of the child and do not undergo spontaneous involution.[5] Vascular malformations are subdivided based on blood flow rate: “slow flow” (capillary, venous, lymphatic or mixed) versus “fast flow” (arteriole, arteriovenous, fistulae or shunt) subtypes. Diagnostic imaging like Colour Doppler ultrasonography helps in differentiating between these subtypes by flow analysis. Thus one can determine flow rates.[6] Some of the imaging modalities for diagnosing vascular malformations include MRI (with or without intravenous gadolinium enhancement) to evaluate the relation of the lesion to the surrounding tissues.

The other major diagnostic tool includes Magnetic resonance angiography (MRA) which provides detailed information regarding flow characteristics and the extent of local tissue involvement.[7] Though Plain radiographs are of little use for evaluation of vascular malformations; they are reported to show calcified phleboliths or cortical erosion of bone in approximately 6% of cases.[8] The management of vascular lesions depends upon the lesion’s location, blood flow characteristics, symptoms, functional disability and cosmetic deformity.[9] Traditionally, surgical excision is frequently advocated for lesions with pain, functional impairment, progressive growth, compressive neuropathy or mass-related complications.[10] If the lesion is greater than 2 cm in dimension with an arteriovenous shunt and local tissue infiltration of the lesion the risk of recurrence after surgical excision increases.[11] Sclerosing agents are substances that cause a marked tissue irritation or thrombosis with subsequent local inflammation and tissue necrosis resulting in fibrosis and tissue contraction. Some of the sclerosing agents include sodium morrhuate, boiling water, nitrogen

mustard and sodium tetradecyl sulphate .They have been used both to treat symptomatic hemangiomas and for embolization of high flow vascular malformation.[12, 13]The use of sclerosing agents as a definitive treatment of arterio venous malformation on the upper labial mucosa is hitherto unreported. Here in this report we describe the diagnostic imaging feature for a case of vascular malformation and its treatment with Intra lesional injection of sclerosing agent (sodium tetradecyl sulfate).

II. CASE STUDY

A 50-year-old woman attended for the treatment of a pathologic lesion in the lower right back tooth region since 2 years . No abnormality or asymmetry was perceptible during extraoral examination. During intraoral examination of hard tissue , occlusal dental caries was seen with respect to 46 , with grade II furcation involvement and Miller's class II gingival recession. On palpation inspectory findings were confirmed and base of the cavity was soft and yielding. Tooth was tender on vertical and horizontal percussion with Grade II mobility . On routine hard tissue examination all permanent teeth were present except 23 with retained deciduous tooth 63 , Occlusal caries was seen with respect to 46 , 27, 37 and Class I molar relation on both right and left side .Attrition with respect to upper and lower anteriors and Cervical abrasion wrt 63 was seen . On routine examination of soft tissues ,a well defined swelling was seen on the right upper labial mucosa extending medio laterally from the distal aspect of 12 to right retro commissure area and superior-inferiorly from the upper lip to 0.5 cm beneath the labial vestibule , measuring about 1*1.5 cm ,roughly oval in shape, smooth surface, bluish in colour, interspersed with erythema .On palpation inspectory findings were confirmed. Swelling was soft in consistency with no obvious raise in temperature over the swelling , nontender on palpation, non fluctuating ,non reducible and compressible. "Fig. 1" On diascopy lesion did not blanch on pressure "Fig .2" A clinical diagnosis of Mucocele wrt upper right labial mucosa was given for the soft tissue lesion and a differential diagnosis of Hemangioma., Kaposi's Sarcoma and Arterio Venous Malformation was given Routine laboratory examination was established. Hematologic factors were within the normal range. Aspiration was positive for a vascular lesion. Rapid filling of the syringe with blood during aspiration placed vascular malformation high on the list of differential diagnoses. "Fig. 3, 3a" Doppler analysis revealed the high flow nature of the lesion. "Fig. 4, 4a" Management of the lesion was executed with the injection of sodium tetradecyl sulfate. The possibility of recurrence and ineffectiveness of the treatment was explained to the patient and the patient fulfilled the consent form . After an infiltration of local anesthesia , sodium tetradecyl sulfate (Thrombovar; Aventis Pharma France, Laboratoires, Chiesi S.A., Courbevoie, France) was administered twice at two-weeks of interval. Each time, 2 ml of sodium tetradecyl sulfate (STS) was injected using a 26-gauge syringe. "Fig 5" A decrease in the size of lesion was apparent after each session, with complete resolution. Follow up at 5 months showed no recurrence of the symptoms. "Fig 6 "

III. DISCUSSION

Vascular malformations are comprised of abnormally formed channels that are lined by quiescent endothelium. Although vascular malformations are congenital in nature, they may not be seen at birth and may not be evident until additional growth or vascular engorgement is seen as a response to trauma,thrombosis, infection, or endocrine fluctuations.[5] Unlike hemangiomas, which involute , the size of AVMs generally increases in size proportionately as the child grows. The mean age at presentation is 19 years with equal predilection for both males and females . AVMs in the maxillofacial skeleton are common with approximately 31% presenting in the head and neck.[16]Histologically, they present with chromosomal-induced errors in endothelial development but demonstrate normal endothelial turnover and thin-walled, dilated channels with sparse smooth muscle cells and adventitial fibrosis. The clinical presentation of vascular abnormalities vary from an asymptomatic birthmark to life-threatening congestive heart failure or an exsanguinating haemorrhage.[6,12,14] The afflicted, it is observed, often seek help from a number of different physicians and undergo repetitive examination for diagnosis, and frequent failed attempts at "definitive" treatment which results in exacerbation of symptoms, lesion recurrences, and disability. Intraosseous vascular malformations of the maxillofacial region can lead to dental emergencies and may cause disfigurement, morbidity and even death.[12,14]Arteriovenous malformations are categorized according to Schobinger clinical staging to describe the degree of progression as Stage I (quiescence), lesion has a bluish-pink stain and warmth. Doppler ultrasonography reveals arteriovenous shunting at this stage and the lesion remains stable for long periods. Stage II (Expansion) has pain, bleeding, and ulceration, stage III (destruction) has dystrophic skin changes, ulceration, bleeding and pain and stage IV malformation (decompensation) have high output cardiac failure.[26] Classification of vascular malformation histologically includes *capillary or small vessel* which Presents with prominent mitotic figures ,plump endothelial nuclei, and intraluminal projections of endothelial cells that simulated vascular or perineural invasion. They have a prevalence of 30% in head and neck region with a local recurrence rate of 20%.[25]

Cavernous or the Large vessels histologically, shows large vascular spaces. 19% occur in head and neck region. Recurrence rate is 9% . *Mixed vessel*. histologically have perineural infiltration, with lymphoid follicles more than that of cavernous type but comparatively lesser than capillary. 5% occur in head and neck region. With a recurrence rate of (28%). [25] Various sclerosing agents (sodium morrhate, boilingwater, nitrogen mustard, etc.) have been used to treat high-flow lesions, but proves ineffective as the agent is moved from the site of action due to blood flow.[15] The ligation of the external carotid has been listed as an adjunct but most authors strongly advise against it, since many anastomoses (internal carotid, ophthalmic, vertebral, cervical, and contralateral external carotid) promote the rapid appearance of a collateral circulation.[5-21] Embolization, combined with surgical treatment, is still the most common approach.[12,13,16,17,22-24]This procedure controls the acute hemorrhagic phase, but does not eliminate the risk of a recurrence, owing to the appearance of a collateral circulation. [23-24]

IV. CONCLUSION

In this study the use of sclerosing agent as a treatment resulted in complete regression of the lesion without any collateral anastomosis. The benefits of the unconventional, non invasive technique applied in the patients treated for vascular malformation include a more esthetic outcome, decreased likelihood of blood loss and danger of transfusion, and inexpensive – as the patient can be treated on an out patient status. However, further investigations are suggested in order to better determine the appropriate selection of patients for this approach.

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Figures

Figures



Figure 1: Picture of Arterio venous malformation. Superficial compressible lesion with bluish discoloration on the Upper labial mucosa in a 50 year old female



Figure 2: Picture showing lesion not blanching on diascopy.



Figure 3: Picture showing blood on aspiration.



Figure 3 a: Picture showing blood on aspiration.



Figure 4: Grey scale ultrasound image of a 1.5 x 0.8 cm hypochoic submucosal lesion on the upper labial mucosa with high flow representing high flow vascular malformation.



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Figure 5: showing Injection of 2 ml of sodium tetradecyl sulfate (STS) using a 26-gauge syringe.



Picture 6: Post treatment follow up picture of the patient ,clinically showing absence of vascular lesion