

Case Report

Diagnostic dilemma in vascular mal-formation of the upper lip: a case report and review of literature



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ABSTRACT

Background: Vascular lesions are broad term used for a wide range of conditions that results in abnormal number, structure, or position of blood vessels now widely divided in two groups; vascular tumors and vascular malformations. These lesions could be clinically diagnosed with 90% accuracy, however radiographic evaluation such as CT and MRI angiograms may be helpful in determining the extent, exact location and flow dynamics of some lesions. This is important because appropriate treatment and prognosis depend largely on the type of lesion. While vascular tumor is known to respond to steroid therapy, vascular malformations do not. **Aim:** To present a case of vascular lesion on the upper lip and its management. **Methods:** We present a case of arterio-venous malformation on the upper lip in a 20year old female and its surgical management. **Findings:** Our case posed a diagnostic and management challenge initially diagnosed as haemangioma, but was eventually diagnosed as vascular malformation with immuno-histochemistry supplemented by CT and MRI angiograms and managed. Definitive treatments undertaken were bilateral upper labial arterial ligation, excision of the lesion and cheilorrhaphy. **Conclusion:** The patient now has an aesthetically acceptable, functioning upper lip. This demonstrates the advantage of using advanced diagnostic aids in diagnosis and surgical management.

Key words: Diagnostic dilemma, vascular malformation, vascular tumors, haemangioma, upper lip.

INTRODUCTION

Vascular anomalies are a wide range of conditions that result in an abnormal number,

structure, or position of blood vessels.^[1] Many classifications have been proposed, but James Wardrop^[2], first recognized the differences between true haemangiomas and the less



common vascular malformations in 1818.^[3] However, despite Wardrop's work,^[2] descriptive identifiers such as Strawberry haemangiomas, salmon patch, stork bite," "angioma simplex," or the indiscriminately applied "haemangioma" continued to be used until the 1980s,^[4,5] when Mulliken and Glowacki developed a system that considers histology, biological behavior, and clinical presentation of these entities.^[6] It is, however, now widely accepted that they can be subdivided into two groups; vascular tumors and vascular malformations.^[1-3]

Vascular tumors are endothelial neoplasms characterized by increased cellular proliferation; vascular malformations, on the other hand, are the result of abnormal development of vascular elements during embryogenesis and fetal life.^[6] Haemangioma is the most common of vascular tumor, with about 4%-10% incidence and 3-5times commoner in females in most studies.^[1,2,7,8] It occurs most commonly in the craniofacial region (60%), followed by the trunk (25%) and extremities (15%).^[9]

The three stages in the lifecycle of a haemangioma that has been described are (1) proliferating phase (0-1 year of age), (2) the involuting phase (1-5 years of age), and (3) the involuted phase (>5 years of age). These stages are typically clinically apparent and can be distinguished microscopically and immunohistochemically.^[1,2,6,10]

The classification of malformation is based on the clinical, radiological and histological appearance of the abnormal channels, which may be either haematic or lymphatic in nature.^[4] A vascular malformation can be slow-flow (that is capillary, lymphatic, or venous) or fast-flow (that is, arterial). If there are combinations of these elements, the malformation is called an arteriovenous malformation (AVM), lymphatico-venous malformation (LVM), or capillary-lymphatico-venous malformation (CLVM).^[1,2,4,11,12] In contrast to vascular tumors, vascular malformations do not have a growth phase, or an involution phase. Vascular malformations tend to grow proportionately with the child, never regresses, but persist throughout life.^[13]

Vascular malformations occur in 1.5% of the population.^[15] Reports suggest that predominantly venous malformations are the most common with incidence of about 40%, followed by lymphatic malformation 28%, arteriovenous malformation, 14% and capillary malformation, 11%.^[12] Clinical presentation of vascular malformations is extremely variable depending on the size and location of the lesion, and it ranges from asymptomatic spots of mere aesthetic concern to lesions with high blood flow or located in critical sites that may be life-threatening.^[13]

The commonest complaint is usually cosmetic. Other complaints are ulceration, with or without superimposed infection, bleeding, swelling and pain.^[11] Clinical diagnosis of vascular lesions have been said to have as high as 90% accuracy,^[3,8,10] however CT and MRI angiograms evaluation may be helpful in determining the exact extent, location and flow dynamics of some lesions.^[4]

Almost all vascular malformations and nearly 40% of haemangiomas eventually require intervention.^[4] Due to their complexity, a multidisciplinary approach is frequently necessary in managing these lesions.^[4] Confusing terminology has been said to be the greatest impediment to the management of vascular lesions.^[1,4,9] Thus, In order to develop a rational plan for management of oral and maxillofacial vascular anomalies, clinicians must have a basic understanding of the biology of these lesions and the nomenclature and classification system used to characterize them.^[11]

CASE REPORT

We present a 20year old female student referred to our Oral and Maxillofacial Surgery Clinic by a general dentist on account of swelling of upper lip of 16 years duration (figure 1) which posed a diagnostic and management challenge. Patient claimed, she had been treated by a spiritualist prior to presenting to the general dentist that referred her to our center.

No previous history of trauma or use of medications known to cause lip swelling. There

is history of spontaneous bleeding episodes from the mass, though family history did not reveal any bleeding diathesis.



Figure 1: Patient at presentation

Examination revealed an upper lip swelling, reddish, ulcerated, bleeding spontaneously, tender and mildly purulent measuring approximately 4.0 by 6.0 cm in dimension. There was associated swelling of the nose, the nasal bridge and glabella region.

No palpable lymph nodes were felt and intra-oral tissues appeared clinically normal. A provisional diagnosis of haemangioma was consequently made.

Hematocrit check was 26%, but white cell count was within normal range. Subsequently, the upper lip was cleansed using hibitane in water and sufratulle dressings applied. Patient was placed on amoxicillin 500mg 8hourly for 5 days and acetaminophen 1g 8hrly for 3days. Alternate day dressing was continued for 2weeks, till infection resolved.

Intra-lesional injection of 80 mg of triamcilonone acetone, slowly given after local anaesthesia was then commenced on an alternate day basis for over 4weeks, to which there was a minimal reduction in the size of lesion. This was later reduced to 40 mg every alternate day and was discontinued after 2 weeks when patient started developing maculo-papular rash and trunk obesity. However, response to steroid was not significantly different.

The investigations that were done included CT and MRI angiograms. CT angiogram showed that the lesion was located to the upper lip and

midline of the face without bone involvement. MRI angiogram on the other hand, revealed numerous hypo-dense thin and thick voids, with areas suggestive of hemorrhage. It also showed dilation of both superior labial arteries (figures 2A and 2B).



Figure 2A: MRI, sagittal view showing the lesion

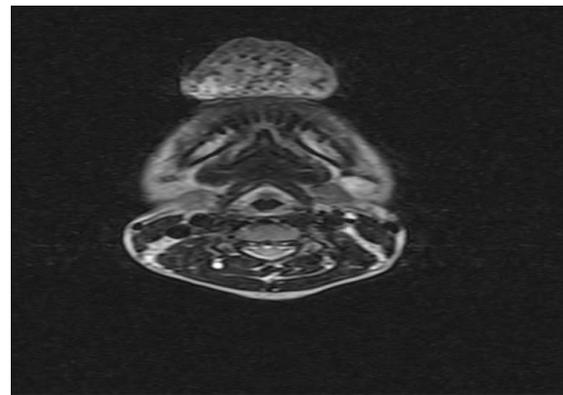


Figure 2B: MRI, axial view showing the lesion

Following transfusion with 1 pint of whole blood, incisional biopsy was carried out under general anesthesia. At surgery, bilateral ligations of the superior labial arteries (figure 3) were done before the sample was taken. Histopathological findings include atrophic and ulcerated parakeratotic stratified squamous epithelium overlying fibrous connective tissue infiltrated by neutrophils, lymphocytes, plasma cells, histiocytes. Deep down the connective tissue are vascular channels of varying sizes, thick and thin walled, some empty and interspaced with areas of haemorrhage

consistent with diagnosis of benign vascular lesion. Immunohistochemical studies confirmed a diagnosis of arteriovenous malformation.



Figure 3: The lesion after upper labial arterial ligation and incisional biopsy

There was a marked reduction in size of the lesion following ligation of the superior labial arteries. After definitive diagnosis of the lesion, a wedge-shaped partial excision of the lesion was done, and reconstruction of the lip (cheilorrhaphy) was accomplished with the remaining tissue under a second anaesthetic session 4 weeks after the initial one. The surgical procedure was well tolerated and the patient was subsequently discharged home after 4 days. Patient now has an aesthetically acceptable and functional lip and she is very happy with the outcome, and 12 months after, the outcome (figure 4) is still stable and there is no evidence of recurrence.



Figure 4: Appearance after surgery (6 months post-operative)

Patient's consent and the institution ethical committee approval were obtained.

DISCUSSION

Vascular lesions can be difficult to diagnose and classify.^[9] Accurate classification is important because treatments and prognosis vary based on the type of lesion.^[2,6,9,11] Diagnosis is based on a combination of clinical features with a variety of imaging techniques, including Doppler US, CT/CTA, MRI/MRA and conventional angiography.^[9] However, differentiation between the types of lesions is usually possible on clinical grounds without the need for sophisticated radiologic or histological analysis.^[6,8,12] On palpation, haemangioma has a firm, rubbery consistency in contrast to vascular malformations, which usually feel soft and are easily compressible,^[14] and may be pulsatile as in our patient. Operation is unnecessary for small haemangioma, but may be necessary for larger lesions, particularly when the tumor interferes with functions such as eating.^[14] In the rare situation where the diagnosis is equivocal, additional studies can be obtained.^[11]

The onset of lesion at the age of 4 years old in our patient supported a variable age of presentation of vascular malformation even though the lesion is said to be present at birth.^[1,2,4-6] Vascular lesions generally have been found to be commoner in females with predilection for head and neck region,^[4,7] this is in consonant with our finding. Macrochelia, ulceration with intermittent bleeding presented by our patient have been documented by other workers.^[14] On vascular lesions generally, though bleeding is said to be more in arteriovenous malformation.^[14] Our patient had an average of 4 episodes of bleeding per month from the lip mass and this started about 6 years prior to presentation, mostly triggered by trauma during mastication and sleep. Bleeding however became spontaneous a few months prior to presentation. Forty percent of haemangioma and more than 90% of AVM have been said to present with bleeding episodes that will necessitate intervention.^[9,15]

Though, clinical diagnosis of vascular lesions is about 90% accurate,^[5,6,8] vascular lesions

can be difficult to diagnose and classify.^[13] A clinical impression of haemangioma was made at the initial presentation of our patient.

Various treatments have been used in the management of vascular lesions, including oral corticosteroids like triamcinolone acetonide or dexamethasone intralesional injection, sclerosing agents like bleomycin, interferon-2b, laser treatment, embolisation and surgery.^[1,2,4,7] Due to the clinical impression of haemangioma, the first line of therapy in our patient was steroid in form of triamcinolone acetonide injection. Various mechanisms of action of corticosteroid have been propounded,^[15] but it is generally agreed that steroids cause a reduction in the levels of various angiogenic and vasculogenic cytokines.^[15] In addition, corticosteroid injection has been said to stabilize the growth of the lesion in at least 95% of patients, and 75% of tumors decrease in size^[16]. Different dosages have been used,^[17] but our patient had a maximum of 80mg intra-lesional triamcinolone acetonide per injection, which has been documented by other workers.^[18]

Our patient response to the steroid therapy was noticeable but not dramatic and therapy was stopped following systemic reactions. The poor response was probably due to the wrong diagnosis of haemangioma, which was later confirmed with histopathology and immunohistochemistry.

Imaging has been documented to aid in the diagnosis of vascular lesion where this is difficult clinically. CT scan, ultrasound, Doppler ultrasound and MRI have been used particularly to diagnose AVM.^[1,8,11] Doppler sonography may show high vessel density and high peak arterial doppler shift in haemangiomas, but not in vascular malformations.^[14] Our inability to make an accurate diagnosis even after CT/MRI angiogram might be due to our poor experience with this lesion.

Many studies agreed that clinical diagnosis of vascular lesions can have an accuracy as high as 90%, but in contrast, immunohistochemistry had to be used in this case to arrive at the final diagnosis.^[3,8,10] Immunohistochemical

techniques have been used as a diagnostic tool particularly to differentiate between vascular tumors and malformations, and also to differentiate between various entities in vascular malformations.^[1,6,11] PCNA, bFGF, VEGF, Wilm 1, collagenase IV, and GLUT-1 markers have been used to differentiate tumors from malformations, with sensitivity and specificity approaching 100% respectively.^[19] During the proliferation phase of haemangiomas, high concentrations of type IV collagenase, vascular endothelial growth factor (VEGF), basic fibroblast growth factor (bFGF) and urokinase may be present.^[3] By contrast, these markers are not present in vascular malformations. Other markers include E-selectin and transforming growth factor-alpha.^[4]

The case presented required surgical intervention as it was aesthetically displeasing to the patient. This is in agreement with the study of Gresham and Adva^[4] that almost all vascular malformations and nearly 40% of haemangiomas eventually require intervention. Moreover, our patient is a female of marriageable age.

The aim of the surgical treatment of a vascular malformation is to perform a complete resection of the malformation in order to prevent its recurrence. However, since vascular malformations often have an infiltrative growth, frequently only subtotal resections can be performed to avoid unacceptable functional or cosmetic disfigurement of the body.^[17] Following the bilateral ligation of superior labial arteries, the partial resection of the lip mass in our patient produced an upper lip that is not only aesthetic, but also functional. Nair *et al.*^[3] reviewed 115 cases of vascular malformation treated surgically and found out that 111 cases gained an acceptable aesthetics with a single procedure.^[3]

Complete surgical excision remains the gold-standard treatment and immediate reconstruction is an integral part of definitive surgery for AVM.^[20] The heterogeneous nature of AVM requires treatment to be tailored for individual patients and the complex excision defects necessitate expertise in a variety of

reconstructive techniques.^[21]

CONCLUSION

We have presented a 20-year-old female with a large arterio-venous vascular lesion on the upper lip with massive ulceration, intermittent bleeding of grave aesthetic concern and loss of function. This was initially diagnosed as haemangioma, but was immunohistochemically confirmed to be a vascular arterio-venous malformation. This was supplemented by MRI angiography findings. Treatment was not achieved through (sclerotherapy) intra-lesional steroid therapy, but by surgical ligation of superior labial arteries, excision of the lesion and cheilorrhaphy. This demonstrates the advantage of the role of modern imaging modalities such as CT and MRI angiograms, and immunohistochemical studies which provide not only diagnosis, but also the extent, location and flow dynamics of the lesion. We advocate suspected vascular lesions of the head and neck, be investigated with similar modalities, for adequate diagnosis and treatment.

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