HEAD AND NECK VASCULAR ANOMALIES
OUR CLINICAL EXPERIENCE AND LITERATURE REVIEW.

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Abstract
Background- Head and neck vascular anomalies are a spectrum of lesion ranging from simple asymptomatic malformations to life-threatening airway vascular tumors. Management of these lesion poses a real challenge to even most experienced otolaryngologists. Systematic evaluation and prompt diagnosis and judicial choice of treatment are essential for proper management of these lesions.

Material and methods- A retrospective review of the head and neck vascular anomalies over three years, from 2017 to 2019 in a tertiary referral centre. Clinical features, radiology, treatment and follow-up data were studied. We did a literature review to give a comprehensive analysis regarding the diagnosis and treatment of the vascular anomalies of the head and neck region.

Results- A total of 25 patients were managed in our department in two years. Of these 25 patients, three patients had infantile haemangioma(12%), six patient had congenital haemangioma(24%), six patients had lymphatic malformation(24%), four patient had an arteriovenous malformation (16%), and six patient had venous malformation (24%). Six patients (24%) underwent surgery as treatment, which includes one case of non-involuting congenital hemangioma (NICH) of the temporal bone, one facial AVM, twolateral neck lymphangiomas, one skull base lymphangioma and a venous neck malformation. All the infantile haemangiomas were managed with oral propranolol, three lymphatic malformations underwent sclerotherapy, while the three Arteriovenous malformations (AVM), five congenital haemangiomas and four Venous malformations (VM) were under observation.

Conclusion- Managing a vascular anomaly requires a prompt diagnosis based on the ISSVA classification, patients' symptoms and the location of the lesion. Not all vascular anomaly requires active management. Careful case selection and a multidisciplinary team are essential for adequate management of the vascular anomalies.

Key Words: Vascular Malformation, Hemangioma, Lymphangioma

Introduction
Vascular anomalies are a distinct group of disease entity which are believed to be caused by an abnormality in angiogenesis and vasculogenesis. (1) The head and neck region is the most common site of these abnormalities and is seen in 4.5% of
children. There has been much confusion regarding the correct nomenclature over the years. Virchow and Wegner developed the first histological classification. In 1976, in order to provide a solution to this growing misconception, a multidisciplinary workshop was performed which was followed by several other meetings ultimately leading to the establishment of the "International Society for Study of Vascular Anomalies" (ISSVA) in 1992. The ISSVA came up with their "Binary classification" in 1996 approved at the XI workshop conducted in Rome (Table 1). This classification is based mainly on the works of Mulliken and Glowacky in 1982. All the vascular malformations are broadly classified into vascular tumors and vascular malformations based on their physical findings, clinical behaviour and cellular kinetics. This classification underwent update at the XX workshop held at Melbourne in April 2014. This update further extensively subclassified the vascular anomalies and incorporated the genetic associations. The latest classification was published by ISSVA in April 2018 and has been stated that the classification will further evolve as the understanding of the biology and genetics of these group of disease keep on growing.

Table 1 – ISSVA classification (1996)

<table>
<thead>
<tr>
<th>Vascular malformation</th>
<th>Vascular tumor</th>
</tr>
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<tbody>
<tr>
<td>Single vessel</td>
<td>Vascular tumor</td>
</tr>
<tr>
<td>Venous</td>
<td>Haemangioma</td>
</tr>
<tr>
<td>Lymphatic</td>
<td>Congenital haemangioma</td>
</tr>
<tr>
<td>Arteriovenous</td>
<td>Rapidly involuting Congenital haemangioma (RICH)</td>
</tr>
<tr>
<td>Combined and complex malformation</td>
<td>Non-involuting Congenital haemangioma (NICH)</td>
</tr>
<tr>
<td>lymphaticovenous</td>
<td>Lobular haemangioma</td>
</tr>
<tr>
<td>Capillary Venous</td>
<td>Kaposiform haemangioendothelioma</td>
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<tr>
<td>Capillary Lymphaticovenous</td>
<td>Capillary Lymphaticovenous</td>
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<tr>
<td>Capillary Arteriovenous</td>
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<td>Capillary Arteriovenous</td>
<td>Capillary Arteriovenous</td>
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Table 2 – Cohort with Vascular anomalies.

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Number of patients</th>
<th>Management</th>
</tr>
</thead>
<tbody>
<tr>
<td>Infantile Haemangioma (IH)</td>
<td>3 (12%)</td>
<td>3</td>
</tr>
<tr>
<td>Congenital Haemangioma (CH)</td>
<td>6 (24%)</td>
<td>5</td>
</tr>
<tr>
<td>Lymphatic Malformations (LM)</td>
<td>6 (24%)</td>
<td>3</td>
</tr>
<tr>
<td>Arterio-venous Malformation (AVM)</td>
<td>4 (16%)</td>
<td>2</td>
</tr>
<tr>
<td>Venous Malformation (VM)</td>
<td>6 (24%)</td>
<td>5</td>
</tr>
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</table>

Discussion and Literature review

Infantile haemangioma (IH)

Figure 1 – Infantile haemangioma

This is the most common vascular tumor. These are real benign tumors of infancy (4-10%) (2). The lesion is common in whites, premature infants, low-birth-weight infants, multiple gestations, infants born to mothers of advanced age, and with history of chorionic villi sampling. The male to female ratio is 1:3. The lesions are Typically

Material and method

A retrospective review of all the vascular anomalies treated in the department from January 2017 to December 2019. The patient age at the time of presentation, clinical presentation, duration of symptoms, location of the vascular anomaly, imaging, type of management, and follow up were analyzed and studied. Imaging was performed in the patients in whom interventional management was planned.

Result

A total of 25 patients were treated in the department in the study period. The minimum age of the study group was 1 month newborn baby, and the maximum age was 37 years, most of our study group belong to the age group less than 18 years (21 patients). Of these 25 patients, three patients had infantile haemangioma (12%), six patient had congenital haemangioma (24%), six patients had lymphatic malformation (24%), four patient had an arteriovenous malformation (16%), and six patient had venous malformation (24%). All the infantile haemangiomas were managed with oral propranolol, three lymphatic malformations underwent sclerotherapy, while the two Arterio-venous malformations (AVM), five congenital haemangiomas and four Venous malformations (VM) were under observation. (Table 2)
absent at birth. Occur in infancy and proliferate until ten months of age. After the first year of life, the lesions typically start to shrink. A hereditary form of IH is associated with chromosome 5 abnormality. The lesion typically occurs in the head, neck and face (60%). They are classified as superficial, deep, segmental focal and mixed. They are classified into six basic growth stages: Nascent, early proliferative, late proliferative, plateau phase, involution and abortive. Based on the site of involvement, the clinical features range from asymptomatic reddish patch to airway obstruction and high-output cardiac failure. Lesion around the eye can cause amblyopia. The lesion in the oral cavity and lip can cause feeding problems. Sometimes IH may be associate with PHACES syndrome (Posterior fossa intracranial abnormality, Haemangioma, Arterial abnormalities, Cardiac defects and coarctation of the aorta, Eye abnormality and Sternal clefting). The most characteristic feature of IH is GLUT-1 positivity on immune-staining of its endothelium. This is the feature that differentiates IH from all other forms of vascular anomalies. The MRI characteristic during the proliferative and plateau phase is focal, lobulated soft tissue mass that is isointense to muscle on T1 and hyperintense on T2, with avid enhancement. Multiple flow voids can be seen on echo-spin sequence. While a CT scan shows a homogenous mass with intense enhancement. Phleboliths and calcification are not seen in IH.

In our study population, we had 3 cases of IH (Fig 1). The first patient had a lesion in the scalp and the posterior triangle, the second patient had a lesion involving the cheek which had stared at 5 months which showed an increase in size over four months, while the third case had the haemangioma of the parotid region and the ear lobe area.

Infantile haemangioma (IH), though are classically described to regress at 9-10 months of age, not all IH can be managed by mere observation. The site of lesion and the symptomatology largely dictates the management plan. The lesions in that cause severe functional and cosmetic deformity should be treated early. The lesions in the subglottis, the eyelid and nasal tip need a special mention as these can cause significant morbidity. In the past, high dose corticosteroids were the mainstay of treatment which mainly act as antiangiogenic agents. Both systemic and intralesional have been used. Oral Prednisolone was given in a dosage of 1 to 5 mg/kg, in a tapering dose over 4 to 6 weeks. Other antiangiogenic chemotherapeutic agents like interferon and vincristine have also been used. The central nervous system toxicity was a particular concern and hence were not very much favoured.

Since the introduction of the non-selective - blocker- propranolol, the treatment of IH has seen a dramatic shift. Since its introduction in 2008 by Léauté-Labrèze and colleagues, propranolol is the first line of treatment for IH. The mechanism of action of the therapeutic effect of propranolol are due to adrenergic vasoconstriction, decreased expression of vascular endothelial growth factor (VEGF) and basic fibroblast growth factor (bFGF) and triggering the apoptosis of the capillary endothelial cells. All these factors reduce the blood vessel density and thereby cause visible involution of the lesion as early as 24 hours from the initiation of treatment. Propranolol needs to be used for six months to 1-year, serious adverse effects include bradycardia, hypotension...
and hypoglycaemia. Pulse-dye laser can be used in superficial HI. Surgery should be considered in cases where there is a functional compromise of vision or breathing and those that are unresponsive to medical therapy. Airway HI is the most challenging of all; they may be asymptomatic, can cause sleep apnoea and acute stridor after an episode of airway infections. The current management protocol of airway HI is to establish airway intubating with a small uncuffed endotracheal tube with empirical use of high dose corticosteroids and propranolol. (1,3,8) All the three cases of IH in our study group were managed with oral propranolol after a cardiology evaluation.

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**Congenital haemangioma (CH)**
As the name suggests, the haemangiomas are present at birth. These are high flow vascular tumors. CH is subdivided into rapidly involuting congenital hemangioma (RICH) and non-involuting congenital hemangioma (NICH). The CH is similar to IH on histology; the only difference is that CH is GLUT-1 negative on immunostaining. (1,8) On MRI and CT, the CH is similar to IH except that CH shows more arteriovenous shunting and fistulae more often. (8)
Figure 2- NICH of the temporal bone. a) HRCT temporal bone showing opacity of the right EAC, middle ear and mastoid area and b) haemangioma in the mastoid cavity (arrow).

The study group had six patients with congenital haemangioma, which included a NICH temporal bone (fig 2), two case of CH oral cavity and two cases of cheek haemangioma and a RICH of the tongue.(fig 3)

Figure 3 a) Haemangioma involving the oropharynx b) reduction of haemangioma after six months of oral propranolol therapy, c) cheek haemangioma d) oral cavity haemangioma and e) tongue haemangioma.

_Congenital haemangioma (CH)_ treatment mostly involves observation for RICH and in cases of NICH laser therapy, or surgery may be required based on the location and rate of progression of the lesion. In our study group, we had one NICH of the temporal bone with a history of bleeding from the ear and hearing loss. Serial contrast MRI showed an increase in size; the patient underwent excision of the haemangioma by a transmastoid approach (fig-2).

_Lymphatic malformation (LM)_ management involves three modalities- observation, sclerotherapy and surgical excision. A unilocular, macro cystic posterolateral LM can be observed over time as they regress spontaneously. Sclerotherapy is reportedly effective in case of a macrocystic lesion. This procedure involves multiple needle aspiration of the cyst content with subsequent injection of the cavity with sclerosing agents like Picibanil (OK-432). (12) Surgery is needed in suprahypoid LM, causing airway and pharyngeal narrowing, lesion compressing cranial nerves and lesions involving skull base regions. (1) In our cohort, three patients underwent percutaneous sclerotherapy, while three patients underwent surgical excision, two of the patients had a suprahypoid stage 2 lesion (fig4) and the third patient had orbital and skull base involvement (fig-5). A few congenital LM diagnosed by prenatal ultrasound may require ex utero intrapartum treatment (EXIT) procedures. (1,2)

<table>
<thead>
<tr>
<th>Stage</th>
<th>Description</th>
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<tbody>
<tr>
<td>Stage I</td>
<td>Unilateral Infrahyoid</td>
</tr>
<tr>
<td>Stage II</td>
<td>Unilateral suprahyoid</td>
</tr>
<tr>
<td>Stage III</td>
<td>Unilateral Infrahyoid and suprahyoid</td>
</tr>
<tr>
<td>Stage IV</td>
<td>Bilateral suprahyoid</td>
</tr>
<tr>
<td>Stage V</td>
<td>Bilateral Infrahyoid and suprahyoid</td>
</tr>
</tbody>
</table>
Malformations (VM)

Venous malformations are abnormal dilated or tangled blood vessels with low flow blood. They are compressible non-pulsatile and enlarge on Valsalva or dependency. The overlying skin usually shows a bluish hue. They are found to be associated with dysfunctions of tyrosine kinase Tie2 receptor. The VM is usually asymptomatic but may be associated with pain due to trauma or puberty. Pain may also be due to irritation of the adjacent nerves or due to intralesional consumption coagulopathy. (1,2) Doppler USG shows a low flow; MRI reveals a bright signal on T2WI while a CT demonstrates phleboliths. (13,8)

In our study, we had a young patient with a VM over the suprasternal notch, initially was under observation. The lesion was enlarging in size and was seeming to grow into the superior mediastinum. The lesion was excised under general anaesthesia. (fig-6)

Arteriovenous malformation (AVM)

AVM are high flow vascular malformations, classically are pulsatile mass that can be present
anywhere in the head and neck region, most commonly occurring in the cheek or the auricle. (1,2) These lesions have an abnormal precapillary communication and should not be confused with an arteriovenous fistula, which is acquired and is an abnormal communication between the arteries and veins. The AVM has four clinical-stage- dormancy, expansion, destruction and heart failure. In some patients, the lesion can grow during adolescent age. (1) Embolization and surgical excision are the mainstays of management of Arteriovenous malformations (AVM). Surgical excision can be challenging in many situations, as there can be significant intra-operative bleeding, and there can be significant tissue loss-making reconstruction a real challenge. (2) In our series, we operate a nasolabial AVM after trans arterial angiography and embolization. (fig-7)

Figure 7- Trans arterial angiography and embolization of left nasolabial fold AVM. a) anteroposterior view of AVM showing nidus (*) filled with dye, b) lateral view, c) and d) anteroposterior and lateral view of after embolization of AVM respectively, showing complete absence of nidus. (white arrow- Internal maxillary artery, major feeding vessel)

Conclusion
Vascular malformations represent a spectrum of a complex and highly variable group of diseases. Managing a vascular anomaly requires a prompt diagnosis based on the ISSVA classification. A meticulous history, patient’s symptoms, expectations and the location of the lesion are the most important in decision making. Careful case selection and a multidisciplinary team are essential for adequate management of the vascular anomalies.

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References

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