ORIGINAL RESEARCH PAPER

Volume-8 | Issue-8 | August - 2019 | PRINT ISSN No. 2277 - 8179

INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

MANAGEMENT OF VASCULAR MALFORMATIONS OF THE HEAD & NECK – A PROSPECTIVE STUDY AND REVIEW OF LITERATURE

Plastic Surgery

Dr. Selvan Ranganathan — Professor & Head, Dept. of Plastic & Reconstructive Surgery, Govt. Chengalpattu Medical College & Hospital, Chengalpattu, Kanchipuram Dist., Tamilnadu

Dr. Surya Rao Rao Venkata Mahipathy* — Professor & Head, Dept. of Plastic & Reconstructive Surgery, Saveetha Medical College & Hospital, Thandalam, Kanchipuram Dist., Tamilnadu *Corresponding Author

ABSTRACT

Background: Vascular anomalies are disorders of the endothelium and surrounding cells that causes localized structural defects of the vasculature. The clinical presentation of vascular anomalies is confusing because all lesions appear in the color spectrum of blue, pink, and red. The most common problem associated with vascular anomalies is psychological distress related to disfigurement as well as functional defects, as many lesions affect the head and neck.

Materials & Methods: This was a prospective study conducted in the Department of Plastic Surgery, Govt. Chengalpattu Medical College & Hospital, Chengalpattu from January 2009 to January 2011. 14 patients with various vascular malformations of the head and neck were included in the study. Patients underwent either conservative management, alcohol ablation, surgical excision or a combined approach. Embolisation was not attempted due to the lack of an interventional radiologist at our institution. The outcomes were later analysed.

Results: A total of 14 patients were included in the study. There were 4 males, 8 females and 2 children. One patient had conservative management, five underwent alcohol ablation (multiple sittings), six had surgical excision and two patients underwent a combined approach. All patients were followed up for a minimum period of 6 months

Conclusion: Management of vascular malformations is a challenge. Haemangiomas were managed conservatively, small low flow lesions were treated by alcohol ablation, localized lesions were managed with excision and primary wound closure and large diffuse lesions were managed by a combined modality.

KEYWORDS

Malformation, conservative, alcohol ablation, surgical excision

INTRODUCTION

Vascular anomalies are a new rapidly developing field involving multiple disciplines, both medical and surgical. The role of a plastic surgeon is an essential part of this program.[1] The estimated prevalence is 4.5%, and the anomalies are usually diagnosed during infancy or childhood [2]. They cause cosmetic disfigurement and functional issues and are therefore a psychological barrier to the patient. In this study, we managed a range of vascular malformations with alcohol ablation, surgery, or a combined approach.

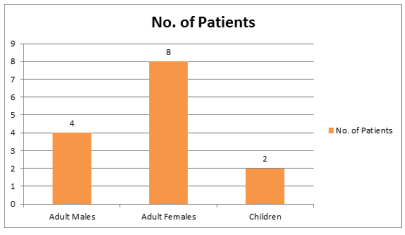
MATERIALS & METHODS

This was prospective study done in the department of Plastic Surgery, Govt. Chengalpattu Medical College & Hospital, Chengalpattu, for a period of 2 years, from January 2009 to January 2011. All patients with vascular anomalies of the head and neck were included in the study. A total of 40 patients were included in the study. The details of the patient's demographic data with history and clinical examination were noted. Patients were subjected to radiological investigations like ultrasonogram (USG) with doppler study, computed tomography (CT) with angiography and magnetic resonance imaging (MRI) accordingly. Lesions were either managed conservatively, ablated using absolute alcohol, surgical excision or by a combined approach. Alcohol ablation was done either independently or prior to surgery to decrease blood loss and for easier handling of the lesion. 2 to 3 ml of absolute alcohol was taken with an equal volume of 2% ligocaine and injected into the lesion. This was repeated at 6 weekly intervals. This was done only for small low flow lesions. For large and diffuse lesions, alcohol was injected 1 week prior to surgery. Surgery was done under general anaesthesia, the lesion was excised in toto and sent for histopathology. The defect was closed either primarily, grafted or covered with a flap. All patients were given post-operative antibiotic therapy, analgesics and anti-edema measures.

RESULTS

A total of 14 patients were included in the study. There were 4 males, 8 females and 2 children. (Table 1)

Table 1 – Sex distribution of the patients



One patient had conservative management, five underwent alcohol ablation (multiple sittings), six had surgical excision and two patients underwent a combined approach. (Table 2) All patients were followed up for a minimum period of 6 months

Table 2 – Type of procedures done

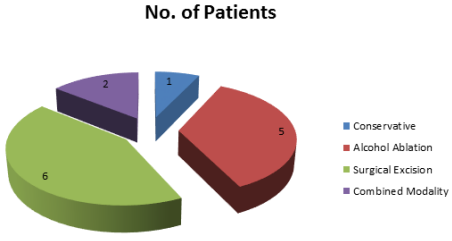


Fig. 1 – Haemangioma of upper eyelid & trunk managed conservatively

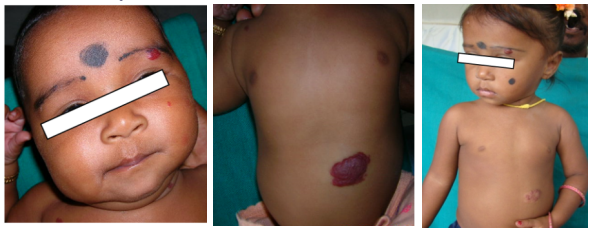


Fig. 2 – Venous malformation Rt. Cheek managed with alcohol ablation (4 months follow up)

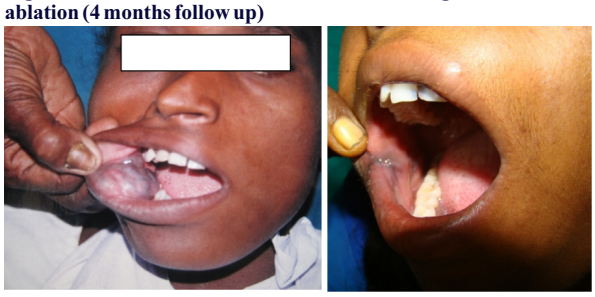


Fig. 3 – Venous malformation lower lip managed with alcohol ablation (3 years follow up)

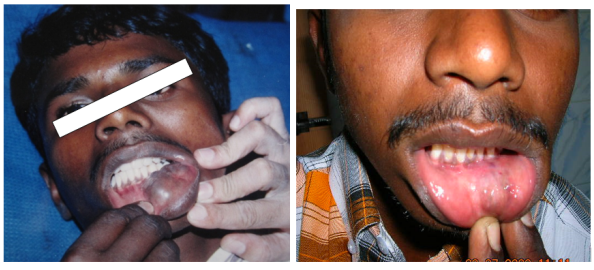


Fig. 4 – Capillary malformation lower lip treated with alcohol ablation

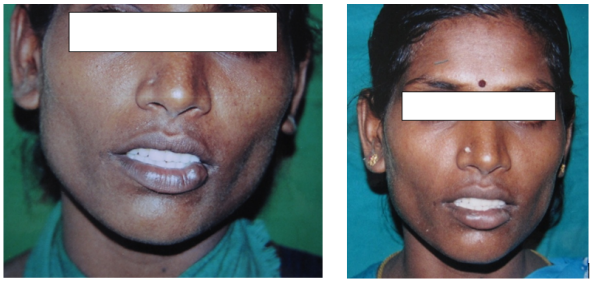


Fig. 5 – Lymphatic malformation tongue treated with alcohol ablation (5 months follow up)

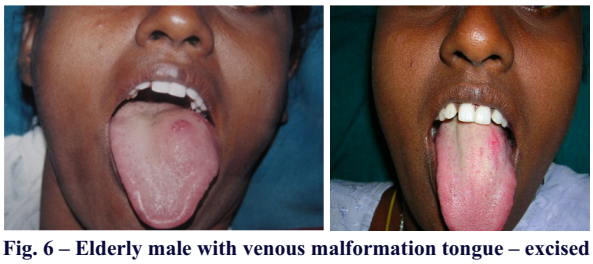


Fig. 6 – Elderly male with venous malformation tongue – excised and closed primarily

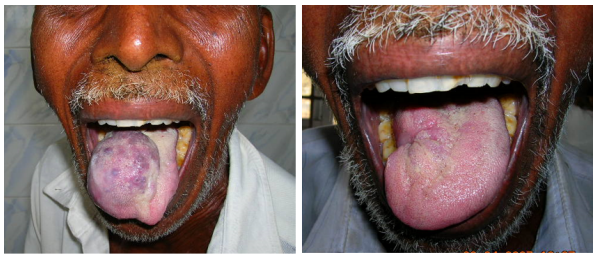


Fig. 7 – Capillary malformation lower lip treated with excision and primary closure

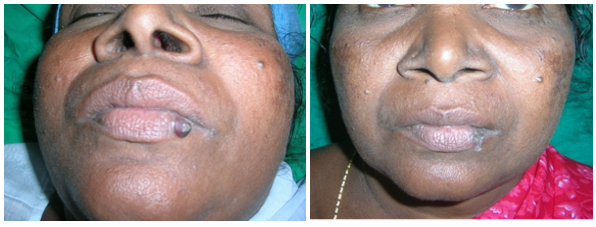


Fig. 8 – Arteriovenous malformation forehead treated by excision

Fig. 9 – Vascular malformation Lt. cheek treated by excision and limberg flap

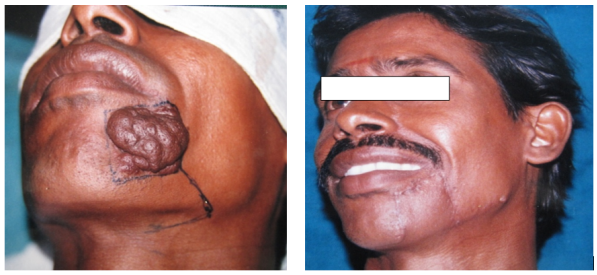
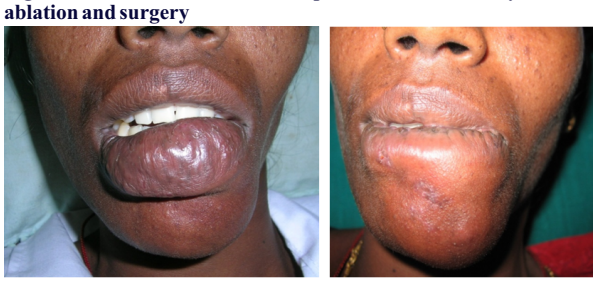


Fig. 10 – Vascular lesion lower lip and chin treated by alcohol ablation and surgery



DISCUSSION

Vascular lesions are localized structural defects of the vasculature. They are common in infancy and childhood, but many a time, we see them in adulthood due to the nature of the lesion, neglect or due to a complication. Vascular anomalies lead to local complications like bleeding, infection, obstruction, pain, thrombosis, ulceration but can also cause general complications such as congestive heart failure, disseminated intravascular coagulation, pulmonary embolism, thrombocytopenia, and sepsis [3]. There has been a confusion in labeling these lesions, so in 1982, Mulliken and Glowacki introduced a simple classification based on the clinical, histochemical, and cellular criteria to distinguish various vascular anomalies which provided framework for the proper identification of these lesions.[4] This was the basis of a new classification system which was adopted by the International Society for the Study of Vascular Anomalies (ISSVA), in which vascular anomalies were divided into tumors and malformations according to the presence or absence of endothelial mitotic activity.[5][Table 3]

Table 3 – Overview of the ISSVA classification of vascular anomalies (approved at the 20th ISSVA Workshop, Melbourne, April 2014, last revision May 2018)

Vascular Anomaly

A) Vascular Tumour

Benign

– Infantile hemangioma
- Congenital hemangioma
– Rapidly involuting (RICH)
- Non-involting (NICH)
- Partially involuting (PICH)
- Tufted angioma
- Spindle-cell hemangioma
- Epithelioid hemangioma
- Pyogenic granuloma
- Others

Locally aggressive or borderline

- Kaposiform hemangioendothelioma

- Retiform hemangioendothelioma
- Papillary intralymphatic angioendothelioma (PILA),Dabska tumor
- Composite hemangioendothelioma
- Pseudomyogenic hemangioendothelioma
- Polymorphous hemangioendothelioma
- Hemangioendothelioma not otherwise specified
- Kaposi sarcoma
- Others

Malignant
- Angiosarcoma
- Epithelioid hemangioendothelioma
- Others

B) Vascular malformation
Simple
- Capillary malformations (CM)
- Lymphatic malformations (LM)
- Venous malformations (VM)
- Arteriovenous malformations (AVM)
- Arteriovenous fistula (AVF)

Combined
- CVM (CM+VM), CLM (CM+LM)
- LVM (LM+VM), CLVM (CM+LM+VM)
- CAVM (CM+AVM)
- CLAVM (CM+LM+AVM)
- Others

VASCULAR TUMOURS

Vascular tumors are usually benign tumours of children, the common types being infantile hemangioma (IH), congenital hemangioma (CH), kaposiform hemangioendothelioma, pyogenic granuloma, and others [6].

Hemangioma

Hemangioma is a benign hamartomatous lesion of capillaries with an increased proliferation index. Hemangiomas are classified into infantile hemangioma (IH) and congenital hemangioma (CH). IH is subcategorized depending on the site of its occurrence as focal, segmental, and indeterminate, and also depending on the depth of the lesion from the skin surface as superficial (previously called capillary hemangioma), deep (previously called cavernous hemangioma), and mixed (capillary cavernous hemangioma). CH is further subcategorized into rapidly involuting CH (RICH), noninvoluting CH (NICH) and partially involuting CH (PICH).[7]

Infantile Hemangioma

IH is the most common benign tumor in infancy [4]. IH affects approximately 4%−5% of Caucasian infants but is rare in other populations [8]. It is common in females (4:1)[9]. It usually occurs singly (80%) and involves the head and neck (60%), followed by the trunk (25%) and extremities (15%) [10]. 30%−50% are found at birth as a telangiectasis or an ecchymotic patch with the mean age at presentation at 2 weeks [11]. During the first 9 months after birth, IH grows rapidly (proliferating phase), faster than the growth of the child. IH involves the superficial dermis, whereas the overlying skin appears bluish. Between 9 to 12 months after birth, the growth of IH plateaus. After 12 months of age, IH begins to shrink, its color fades, and the lesion flattens (involuting phase). By 5 to 7 years of age, involution stops in most patients (involuted phase). The local complications include residual telangiectasias, loss of elastic fibers, scarring, redundant skin, or destroyed anatomic structures [12,13]. The immunostaining for an erythrocyte-type glucose transporter (GLUT1), which is specifically expressed in IH, can differentiate IH from other tumors and malformations [14]. Propranolol, a nonselective beta adrenergic blocker, is the first-line treatment of complicated IH. Most IH are managed conservatively because 90% are small, localized, and involute without sequelae. But some patients may require operative treatment because of residual fibrofatty tissue, redundant skin, or damaged structures after involution [10].

Congenital hemangioma

Chs are biologically distinct from the IH. IHs appear more common (70%) than CH (30%). CH clinically presents as fully developed lesions at birth which either rapidly involutes during the 1st year of life or may never show involution. These lesions do not exhibit a proliferative phase and usually do not grow after birth. RICHs are present at birth either as red-purple color plaques with coarse telangiectasia, as flat violaceous lesions, or as a grayish tumor surrounded by a pale halo with multiple tiny telangiectasias. RICH undergoes a rapid regression phase and completely disappears by 12–18 months of age. NICHs are present at birth as pink or purple-colored plaque-like lesions with prominent overlying coarse telangiectasia and peripheral blanching. NICH does not show a regression phase, may grow proportionately with the growth of the child, and can be mistaken for vascular malformations.[15,16] NICH and IH have similar histologic appearances. Tissue-specific immunohistochemical markers such as glucose transporter-1 (GLUT-1), merosin, Fc-gamma-RII, and Lewis Y antigens are positive for IH and thus aid in differentiating IHs from other vascular tumors or malformations.[15,17]

Kaposiform hemangioendothelioma

Kaposiform hemangioendothelioma (KHE) is a distinctive vascular neoplasm of childhood associated with severe thrombocytopenia, petichae, and bleeding, known as Kasabach–Merritt syndrome. The tumor is generally present at birth, although it can also appear postnatally. Males and females are equally affected. They are unifocal and commonly involve the trunk, shoulder girdle, thigh, perineum or retroperitoneum, and less commonly, the head and neck region.[18,19]

Pyogenic granuloma

Pyogenic granuloma is a common vascular tumor that appears either spontaneously or after trauma. They are commonly seen on the gingiva, where they are presumably caused by calculus or foreign material within the gingival crevice. Hormonal changes of puberty and pregnancy may modify the gingival reparative response to injury, producing what was once called a “pregnancy tumor.” The lesions appear as red, pedunculated nodules that frequently bleed spontaneously. [18,19]

Angiofibroma

Angiofibromas are rare and constitute <1% of all head and neck tumors. They occur exclusively in young males between 10 and 25 years of age. The site of origin of this tumor is thought to arise from nasopharynx or take origin in the pterygopalatine fossa in the recess behind the sphenopalatine ganglion, at the exit aperture of pterygoid canal. The predilection of juvenile angiofibroma (JA) for young adolescent males led to a suggested interrelationship between hormones and JA.[18,20] Clinically, it usually presents with unilateral nasal obstruction, epistaxis, and nasopharyngeal mass.[18,20] Histologically, it is characterized by the presence of a collagenized vascular stroma containing numerous, irregularly shaped blood vessels lacking elastic fibers in their wall.[17,18]

VASCULAR MALFORMATIONS

Vascular malformations, as congenital abnormalities, result from abnormal vessel development and morphogenesis. In general, they are present at birth (but may be hidden in a deep location) and grow in proportion to the child's growth, persisting throughout the lifetime.

Capillary malformations

Capillary malformations (Cms), also known as port-wine stains or nevus flammeus, are the most common type of congenital vascular malformations [21]. These lesions are initially flat and bright pink, red, or violaceous and typically affect the face (90%), followed by the neck, trunk, leg, arm, and hand [22-24]. They often seem to lighten significantly over the first few months of life. This is not indicative of spontaneous resolution but is probably due to a decrease in circulating blood hemoglobin concentration [25]. The incidence is 0.3% in newborns, with an equal sex distribution [8]. CMs occur as a sporadic unifocal lesion and are not associated with any underlying abnormalities. CMs are sometimes associated with other underlying syndromes such as Sturge - Weber syndrome, macrocephaly - capillary malformation syndrome, capillary malformation - arteriovenous malformation syndrome, and overgrowth syndromes such as Klippel - Trenaunay syndrome [26]. Facial CMs initially appear as a faint pink macule, but some patients may develop soft tissue hypertrophy, bony hypertrophy, and/or nodule formation during adulthood. Depending on the size and location, they can cause functional deficits in vision, speech, or eating, and significant psychological distress. The gold standard therapy for facial or aesthetically sensitive CM is still the pulsed dye laser treatment. In patients with associated soft tissue or bony hypertrophy, surgical management can be helpful in restoring the normal anatomy and in re-establishing a symmetric contour [23,24,27,28].

Lymphatic malformations

Lymphatic malformation (LM) results from errors in the development of the lymphatic system; lymphatic tissue may form in an abnormal location [28]. LM is divided into three types according to the size of the malformed channels, namely, microcystic, macrocystic, or combined (microcystic/macrocystic) [4,10]. LM is a soft and compressible lesion that usually appears at birth; however, a small or deep lesion may not become evident until the lesion has grown large enough to cause deformity or symptoms. LM is most commonly located on the head and neck, causing a deformity and psychosocial morbidity. The overlying skin may be normal, have a bluish tinge or have pink vesicles similar to CM. LM is problematic because of its slow expansion over time, and its recurrence. The common complications are bleeding and infection. Intralesional bleeding occurs in up to 35% of LMs, causing pain or swelling [29]. LM is vulnerable to infection because the malformed lymphatics contribute less to antibody production and protein-rich fluid provides a good environment for bacterial growth. Small or asymptomatic lesions may be observed. Symptomatic lesions causing pain, deformity, or threatening vital structures necessitate operative treatment [30].

Venous malformations

Venous malformation (VM) results from errors in vascular morphogenesis. Thin-walled veins with abnormal smooth muscle are dilated, and then the VM expands and the flow stagnates with clotting. VM is present at birth but may not become evident until it has grown large enough to cause a deformity or symptoms. VMs are blue, soft, and compressible; sometimes phleboliths may be palpable. VMs may appear from localized skin lesions to diffuse malformations involving multiple tissue and structures [31]. Larger VMs usually and involve the skin, mucosa, or subcutaneous tissue; VMs also involve muscle, bone, and viscera. They may appear singly (99%); and are located on the head/neck (47%), followed by the extremities (40%) and trunk (13%) [32]. Common complications are ulceration, bleeding, compression of adjacent structures, and chronic low grade consumptive coagulopathy in large and extensive lesions. Pain and swelling are due to thrombosis and phlebolith formation. In the head and neck, VMs may severely affect compression of adjacent structures. Asymptomatic lesions can be managed conservatively, but symptomatic lesions causing pain, deformity, or threatening vital structures may require sclerotherapy or operative treatment [31].

Arteriovenous malformations

Arteriovenous malformation (AVM) results from errors in vascular development during embryogenesis; absent capillary beds lead to shunting directly from the arterial to venous circulation through a fistula or nidus (abnormal channels between feeding arteries and draining veins) [33]. The most common site of extracranial AVM is the head and neck, followed by the limbs, trunk, and viscera [11]. AVM is present at birth but may not become evident until childhood. AVM has a pink-red cutaneous stain with a palpable thrill or bruit, and it is important to distinguish AVM from a CM or hemangioma. Arteriovenous shunting reduces capillary oxygen delivery, causing ischemia. Common complications are pain, ulceration, bleeding, and congestive heart failure. AVM is not a static malformation, progresses over time, and recurs. Capillary malformation - arteriovenous malformation (CM-AVM) results from a mutation in *RASA1* [34]. The goal of treatment usually is to control AVM. For superficial AVMs, patients should prevent desiccation and subsequent ulceration, and compression garments for extremity lesions may reduce pain and swelling. Intervention including embolization, resection, or a combination is focused on reducing symptoms, preserving vital functions, and improving deformities [4,35].

CONCLUSION

Vascular lesions are disorders of aberrant angiogenesis, vasculogenesis, or lymphangiogenesis. It is important that academicians and clinicians should have knowledge regarding the current classification and the different treatment modalities.

Funding: None
Conflicts of interest: Nil

REFERENCES

1. Jennifer J Marler, John B Mulliken. Current management of hemangiomas and vascular malformations. Cin Plast Surg 32 (2005); 99-116
2. Greene AK, Kim S, Rogers GF, Fishman SJ, Olsen BR, Mulliken JB. Risk of vascular anomalies with Down syndrome. Pediatrics. 2008;121:e135.
3. Greene AK. Vascular anomalies: current overview of the field. Clin Plast Surg. 2011;38:1–5.
4. Mulliken JB, Glowacki J. Hemangiomas and vascular malformations in infants and children: a classification based on endothelial characteristics. Plast Reconstr Surg. 1982;69:412–22.
5. ISSVA. Amsterdam: International Society for the Study of Vascular Anomalies; ISSVA classification for vascular anomalies [Internet] [cited 2018 Dec 20]. Available from: http://www.issva.org/classification.
6. Greene AK. Management of hemangiomas and other vascular tumors. Clin Plast Surg. 2011;38:45–63
7. Ethunandan M, Mellor TK. Haemangiomas and vascular malformations of the maxillofacial region – A review. Br J Oral Maxillofac Surg 2006;44:263-72.
8. Jacobs AH, Walton RG. The incidence of birthmarks in the neonate. Pediatrics. 1976;58:218–22.
9. Drolet BA, Swanson EA, Frieden IJ, Hemangioma Investigator Group Infantile hemangiomas: an emerging health issue linked to an increased rate of low birth weight infants. J Pediatr. 2008;153:712–5. e1.
10. Finn MC, Glowacki J, Mulliken JB. Congenital vascular lesions: clinical application of a new classification. J Pediatr Surg. 1983;18:894–900.
11. Mulliken JB, Fishman SJ, Burrows PE. Vascular anomalies. Curr Probl Surg. 2000;37:517–84.
12. Chang LC, Haggstrom AN, Drolet BA, Baselga E, Chamlin SL, Garzon MC, et al. Growth characteristics of infantile hemangiomas: implications for management. Pediatrics. 2008;122:360–7.
13. Bowers RE, Graham EA, Tomlinson KM. The natural history of the strawberry nevus. Arch Dermatol. 1960;82:667–80.
14. North PE, Waner M, Mizeracki A, Mihm MC., Jr GLUT1: a newly discovered immunohistochemical marker for juvenile hemangiomas. Hum Pathol. 2000;31:11–22.
15. Berenguer B, Mulliken JB, Enjolras O, Boon LM, Wassef M, Josset P, et al. Rapidly involuting congenital hemangioma: Clinical and histopathologic features. Pediatr Dev Pathol 2003;6:495-510.
16. Restrepo R, Palani R, Cervantes L, Duarte A, Amjad I, Altman N. Hemangiomas revisited: The useful, the unusual and the new. Pediatr Radiol 2011;41:895-915.
17. Gontijo B, Silva CMR, Pereira LB. Hemangioma of infancy. An Bras Dermatol Rio J 2003;78:651-73.
18. Gnepp Dr. Diagnostic Surgical Pathology of Head and Neck. 2nd edition. Philidelphia: Saunders; 2009.
19. O'Hara CD, Nascimento AG. Endothelial lesions of soft tissues: A review of reactive and neoplastic entities with emphasis on low-grade malignant ("borderline") vascular tumors. Adv Anat Pathol 2003;10:69-87.
20. Antoniades K, Antoniades DZ, Antoniades V. Juvenile angiofibroma: Report of a case with intraoral presentation. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2002;94:228-32.
21. Johnson WC. Pathology of cutaneous vascular tumors. Int J Dermatol. 1976;15:239–70.
22. Mills CM, Lanigan SW, Hughes J, Anstey AV. Demographic study of port wine stain patients attending a laser clinic: family history, prevalence of naevus anaemicus and results of prior treatment. Clin Exp Dermatol. 1997;22:166–8.
23. Renfro L, Geronemus RG. Anatomical differences of portwine stains in response to treatment with the pulsed dye laser. Arch Dermatol. 1993;129:182–8.
24. Orten SS, Waner M, Flock S, Roberson PK, Kincannon J. Portwine stains: an assessment of 5 years of treatment. Arch Otolaryngol Head Neck Surg. 1996;122:1174–9.
25. Cordoro KM, Speetzen LS, Koerper MA, Frieden IJ. Physiologic changes in vascular birthmarks during early infancy: mechanisms and clinical implications. J Am Acad Dermatol. 2009;60:669–75.
26. Gonzalez ME, Burk CJ, Barbouth DS, Connelly EA. Macrocephaly-capillary malformation: a report of three cases and review of the literature. Pediatr Dermatol. 2009;26:342–6.
27. Lee JW, Chung HY. Capillary malformations (portwine stains) of the head and neck: natural history, investigations, laser, and surgical management. Otolaryngol Clin North Am. 2018;51:197–211.
28. Young AE. Pathogenesis of vascular malformations. In: Mulliken JB, Young AE, editors. Vascular birthmarks: hemangiomas and malformations. Philadelphia: Saunders; 1988. pp. 107–13.
29. Padwa BL, Hayward PG, Ferraro NF, Mulliken JB. Cervicofacial lymphatic malformation: clinical course, surgical intervention, and pathogenesis of skeletal hypertrophy. Plast Reconstr Surg. 1995;95:951–60.
30. Greene AK, Perlyn CA, Alomari AI. Management of lymphatic malformations. Clin Plast Surg. 2011;38:75–82.
31. Greene AK, Alomari AI. Management of venous malformations. Clin Plast Surg. 2011;38:83–93.
32. Boon LM, Mulliken JB, Enjolras O, Vikkula M. Glomuvenous malformation (glomangioma) and venous malformation: distinct clinicopathologic and genetic entities. Arch Dermatol. 2004;140:971–6.
33. Young AE, Mulliken JB. Arteriovenous malformations. In: Mulliken JB, Young AE, editors. Vascular birthmarks: hemangiomas and malformations. Philadelphia: Saunders; 1988. pp. 228–45.
34. Eerola I, Boon LM, Mulliken JB, Burrows PE, Dompmartin A, Watanabe S, et al. Capillary malformation-arteriovenous malformation, a new clinical and genetic disorder caused by RASA1 mutations. Am J Hum Genet. 2003;73:1240–9.
35. Greene AK, Orbach DB. Management of arteriovenous malformations. Clin Plast Surg. 2011;38:95–106.