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Hemangiomas of head and neck: A review

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Abstract

Hemangiomas which are tumors of vascular origin constitute 7% of all benign tumours. Hemangiomas of the head and neck region comprise about 60-70%. Hemangiomas can be infantile or congenital. Hemangiomas are tumors that occur fully grown at birth and do not manifest the postnatal course and lifecycle of common IH. They can be sub classified as rapidly involuting congenital Hemangiomas (RICH) and Non Involuting congenital Hemangiomas (NICH). The Purpose of this article is to give a comprehensive review of hemangiomas and the diverse treatment option available for this complex endothelial tumor and to stress the fact of individualizing a treatment protocol.

Keywords: hemangioma; congenital hemangioma; infantile hemangioma

Introduction

Hemangiomas which are tumors of vascular origin constitute 7% of all benign tumours [1]. Hemangiomas of the head and neck region comprise about 60-70% [2]. According to Enzinger and Weiss, Hemangiomas are broadly classified into Capillary, Cavernous and miscellaneous forms like Verrucous, Venous, Arterio venous Hemangiomas and so forth [3]. Hemangiomas can be infantile or congenital Hemangiomas. Infantile Hemangiomas (IH) are more common in pre-mature infants with a reported incidence of 23% in neonates weighing less than 1200 grams [4]. It is also the most common tumors of infancy and childhood occurring in 4-10% of Caucasian infants [5]. In contrast congenital Hemangiomas are tumors that occur fully grown at birth and do not manifest the postnatal course and lifecycle of common IH. They can be sub-classified as rapidly involuting congenital Hemangiomas (RICH) and Non Involuting congenital Hemangiomas (NICH).

Infantile Hemangiomas

Infantile Hemangiomas grow rapidly, regress slowly and never recur. The three stages in the lifecycle of a Hemangioma are proliferating phase (0-1 yrs of age), involuting phase (1-5 yrs of age) and involuted phase (>5 yrs of age). The three stages can be distinguished clinically, microscopically and immunohistochemically [6]. Histologically in the proliferating phase, there are plump rapidly dividing endothelial cells forming tightly packed sinusoidal channels [7], whereas in the involuting phase the endothelial proliferation is decreased, apoptosis is increased and fibro fatty replacement of Hemangiomas has just begun [8]. During the involuted phase all that remains are few tiny capillary like feeding vessel and draining veins surrounded by islands of fibro fatty tissue admixed with dense collagen and reticular fibres. Immunohistochemically angiogenic proteins such as fibroblast growth factor [bFGF] and vascular endothelial growth factor [VEGF] and enzymes involved in remodeling of extra cellular matrix are prominent in proliferating hemangiomas and diminishes to normal level during regression [9]. Clinically if the tumor is in the superficial dermis the skin is raised, crimson in color and firm and rubbery to palpation. If the tumor is in the deeper dermis and sub cutis, the lesion appear raised with a bluish hue. As it regresses it becomes softer and erythematous color changes to a grey hue. After involution is complete there may be redundant atrophic skin, yellow discoloration or scarred patches in instances of previous ulceration, residual fibro fatty tissue and telangiectasia [7]. Multi focal Hemangiomas involving visceral organs, if large can divert significant intra vascular volume leading to high output cardiac failure [10]. A dermatomal distribution in V1, V2, V3 should rule out PHACES Syndrome (Posterior fossa malformations, Hemangiomas arterial anomalies, coarctation of aorta, cardiac defects and eyeabnormalities) [11].

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Periorbital hemangiomas can block the visual axis causing deprivation amblyopia or may extend into retro bulbar space leading to ocular proptosis [12]. Subglottic Hemangiomas are life threatening lesion presenting with symptoms such as hoarseness, biphasic stridor and airway obstruction [13]. Ulceration occur in 5% of cutaneous Hemangiomas and are more common in lesion involving lip, perineum, anogenital area and extremities [14].

Congenital Hemangiomas

Rapidly involuting Hemangiomas are more common in the trunk, head or neck region. Three morphologic variants of RICH exist, a) A lesion with characteristic red purple color with a coarse telangiectasia. b) A flat infiltrative tumor with violaceous overlying skin. c) Raised grayish tumor with multiple tiny telangiectasia with pale halo [15]. Histologically RICH is composed small to large lobules of capillaries with moderately plump endothelial cells and pericytes [16]. NICH are also more common in neck, trunk or limbs. They tend to be plaque like with a pink to purple color with prominent coarse telangiectasia. Lesion are warmer to palpation than the surrounding skin. Histopathology reveals lobular collections of small thin walled vessels with large stellate shaped central vessels [17].

Pathogenesis of Hemangiomas

The placental theory for the origin of Hemangiomas was proposed by North *et al.*, [18] who reported that infantile Hemangiomas may originate from either invading angioblasts that differentiate towards a placental phenotype or from embolised placental cells. GLUT 1 is expressed throughout the three phases of infantile Hemangiomas making it a valuable tool in diagnosis of infantile Hemangiomas. On the contrary both RICH and NICH are GLUT 1 negative [19]. Dadras *et al.*, showed that lymphatic endothelial hyaluronan receptor LYVE-1 a marker was strongly expressed in proliferating Hemangiomas, diminished in involuting and absent in involuted Hemangiomas [20].

Imaging

The key imaging required for the diagnosis of Hemangiomas are ultra sound, doppler ultra sound and MRI. CT fails to adequately delineate soft tissue planes. MRI produces high signal intensity representing blood as well as focal heterogeneities representing areas of fibrosis, thrombosis or calcification. Doppler shows arterial or venous waveforms with high vessel density >5 vessels/cm² with high doppler shift >2kHz [21]. A study detailing the imaging tendencies of congenital vs infantile Hemangiomas including heterogeneity (72% of NICH, 62.5% of RICH vs 92.31% of infantile Hemangiomas), visible vessels (72% NICH, 62.5% RICH vs 15.4% of infantile Hemangiomas) and calcification (17% NICH, 37.5% RICH vs none in infantile Hemangiomas) [22]. Angiography of RICH demonstrates a well circumscribed areas with an intense tissue staining in a lobular pattern with enlarged surrounding systemic artery branches [23].

Management

The treatment of choice for Hemangiomas depend primarily on the site and growth stage of the lesion [24]. The rationale behind treating a Hemangiomas to prevent or improve functional impairment or pain, to prevent or improve scarring or disfigurement or to avoid life threatening complications. Small, isolated or multiple skin lesions on face in infants are treated early to avoid progression into proliferation.

Imiquimod is an immuno modifier used for small and intermediate sized Hemangiomas with alternate day topical application for a cycle of 3-5 months [25]. The drawback of the drug is hyper pigmentation which makes it use in the highly esthetic region such as the face controversial. Laser therapy is indicated in cases of superficial proliferating Hemangiomas, the laser therapy accelerates the regression or reduces the sizes of the lesion. The choice of laser depends on location, depth and size of the lesion. Flash lamp pumped pulsed dye laser with a wavelength of 585 or 595nm is the only laser that photo coagulate the target vessels leaving the overlying skin intact thereby making it useful for superficial Hemangiomas or those at involution stage, however it is of little use in deep seated Hemangiomas because of limited penetration depth [26]. Neodymium Yttrium aluminum garnet laser (Nd:YAG) having wavelength of 1064nm and penetration depth of 5mm is the choice for larger or 2cm deep seated Hemangiomas. Percutaneous laser therapy can be used for deep Hemangiomas but cooling devices are needed to lower the temperature to prevent damage to the epidermis from thermal injury [27]. The effectiveness of laser is 77-100% in treating Hemangiomas. Drug therapy is indicated for mixed proliferative and Hemangiomas affecting vital organs or life threatening Hemangiomas. Oral prednisolone is more effective than intravenous injection of methyl prednisolone. The effectiveness rate is 84% with a significant relationship between dosage and effectiveness [28]. The regimen for oral corticosteroid is oral prednisolone (3-5 mg/kg) every morning for 6-8 wks. The dose is tapered after that for 2 or 3 wks. The treatment can be repeated for 2 or 3 cycles at an interval of 4-6 wks [29]. In case of more localized Hemangiomas such as orbital or parotid lesion, intra lesional steroid triamcinolonacetone 1-2mg/kg body weight is given at monthly intervals [30]. Pingyangmycin (bleomycin A5) having a high sclerosing effect on vascular endothelium is found to have greater than 90% success rate and 49% complete resolution [31]. It is suitable for proliferating Hemangiomas responding poorly to steroids or laser therapy, for cutaneous superficial or mucosal Hemangiomas its concentration is 1mg/ml; for subcutaneous or deep, the concentration is 1.5-2mg/ml [32]. In case of Hemangiomas unresponsive to steroids or recurrence after steroids, Vincristine 0.5 - 1 mg/ml can be given intravenously once a week over 6 weeks [33]. In case of life threatening platelet consumptive coagulopathy although more common in the trunk than the head and neck, diluted ethanol embolo therapy is reported to be effective. Alpha interferon has been effective but has serious side effects such as spastic diplegia [34]. Recently propranolol has been used to treat infantile Hemangiomas on the basis that propranolol being a β blocker induce apoptosis by antagonising GLUT-1 receptors. Leslie *et al.*, reported a protocol for usage of propranolol. Starting dose of 0.17mg/kg given at 8 hourly intervals. Vitals signs and blood glucose are monitored one hour after each dose. If the first two doses are tolerated, the amount is doubled to 33mg/kg/dose. After two more doses, the amount is doubled to 0.67mg/kg/dose. For infants less than 3 months of age, slower dose escalation is recommended to avoid the risk of hypoglycaemia [35]. The choice of a surgical resection for Hemangiomas should be made taking into account that surgical intervention would be more esthetically acceptable than that from the medical treatment or from observation. The surgical indications for proliferating Hemangiomas are, a. Hemangiomas located in the nose and lip that did not respond to other treatment. b. Eyelids that cause visual interference. c.

Repeated bleeding from the Hemangiomas [36]. Residual deformities often require surgical recontouring to improve esthetics and function. Local wound care is needed to alleviate pain and infection in ulcerated lesion. Debridement of the ulcer in conjunction with topical mupirocin, bacitracin or metronidazole is needed.

Conclusion

Hemangiomas are a complex group of endothelial tumor having a heterogenic city in their presentation and treatment of Hemangiomas have always been a controversial issue, any treatment decision whether surgical or medical must be critically evaluated in terms of cure and esthetic results, and the choice of treatment plan should be individualized rather than a fixed treatment protocol.

Conflict Of Interest & Source of Funding

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