http://jmscr.igmpublication.org/home/
ISSN (e)-2347-176x ISSN (p) 2455-0450
crossref DOI: https://dx.doi.org/10.18535/jmscr/v7i10.04

IGM Publication
Journal Of Medical Science And Clinical Research
An Official Publication Of IGM Publication

A Case Report of Infantile Hemangioma

Authors
Dr P. Pari[1], Dr Nalini MD, DM (Neonatalogy)[2], Dr N.S. Raghupathy, D.Ch., M.D., Ph.D.[3]
[1]Post-graduate in MD Paediatrics
[2]Assistant Professor, Department of Paediatrics at Aarupadai Veedu Medical College and Hospital, Puducherry
[3]HOD and Professor, Department of Paediatrics at Aarupadai Veedu Medical College and Hospital, Puducherry

Abstract
Infantile Hemangioma are proliferative, benign vascular tumours of vascular endothelium that may be present at birth or present in first two weeks of life, predictably enlarge and then involute spontaneously. We report an infant with Infantile Hemangioma in multiple sites and moderate Ostium Secundum type Atrial Septal Defect with Left to Right shunt. The baby is started on Tablet Propranolol. Clinically we can diagnose Infantile Hemangioma. But need to rule out systemic involvement like PHACES Syndrome and Kasabach Merritt phenomenon.
Keywords: *Infantile Hemangioma, Propranolol, Vascular Tumours.*

Introduction

Infantile Hemangioma are benign tumours of vascular endothelium that may be present at birth or present in first two weeks of life. Incidence is 5% of live newborns. Risk factors include Caucasians, girls, twins, preterm and Low Birth Weight. Types- superficial, deep or mixed. Favoured sites are face, scalp, back and anterior chest. Infantile Hemangiomas undergo a phase of rapid expansion followed by stationary period and then spontaneous involution. Approximately 60%lesions reach maximum involution by 5 years of age and 90-95% by 9 years of age.(1)

Case Report

A 26 years old primi mother married by non-consanguinous marriage with twin pregnancy. There was no family history of fetal anomalies. Targeted Imaging for Fetal Anomalies scan was normal with twin pregnancy. She had spontaneous onset of labour at 36weeks+6days and delivered two female babies with first twin 2.75 kg and second twin 2.35 kg by emergency LSCS. Apgar scores were 9/10 and 10/10 at 1, 5 minutes respectively. They were mildly discordant twins. Admitted in NICU for risk of sepsis due to Premature Rupture of Membranes and hypoglycaemia. Babies are discharged after 1 week. After 2 weeks parents noticed a red tinge on the dorsum of right hand and scalp of the second twin. The other twin is normal. They came to our hospital and was diagnosed as Infantile Hemangioma based on clinical examination and history. Investigations were done. Hb-12.6gm%, Platelet count - 2.36 lakhs/cu.mm. ECHO findings are Congenial Heart Disease, Situs Solitus,

Ostium Secundum Type Atrial Septal Defect with Left to Right shunt with increased Pulmonary artery flow, Normal Pulmonary Venous Drainage and Normal Biventricular function. Ultrasound abdomen - Normal. Ultrasound cranium - Normal. The child is started on Tablet Propranolol 2.5 mg bd. After starting the medication, the swelling starts to reduce in size after 1 week. The child is on regular follow up.

Etiopathogenesis

Interaction of both genetic and environmental factors. Several theories have been proposed which includes Folkman Klagsburn placental theory, endothelial progenitor cell theory, hypoxia theory and angiogenesis theory. [2]

Pathophysiology

Infantile Hemangiomasare result of endothelial cell hyperplasia. The origin of Hemangioma is due to anabolic placental angioblastic or intrinsic endothelial progenitor cells with ability to clonally duplicate in a precise milieu of cytokines and estrogen. North et al. discovered that the histology and molecular markers unique to placental tissue namely GLUT1, Lewis Y antigen, Merosin and Receptor III were also present in Infantile Hemangiomas. The placental tissue suggests that fetal progenitor cells arise from disrupted placental tissue during gestation or birth and is further supported by the increased incidence of Infantile Hemangioma found following chorionic villus sampling, placenta previa and pre-eclampsia. Yu et al, 2004 found these tumours arise from embryonic endothelial precursors identified by their specific cell-surface protein CD133+/CD34+. Environmental cues perinatal events, disordered cytokines signaling, angiogenic factors and genetic determinants have been explored [2,3]

Clinical Features

CLINICAL PHASE	FEATURES
Prodromal Phase	Premonitary lesions - circumscribed telangiectasia.
Initial Phase	Loss of typical skin structure with increasing thickness and duration.
Proliferation Phase	Bright red cutaneous infiltration. Firm and non-compressible.
Maturation Phase	Raised bosselated crimson red lesions. Ulceration seen
Regression Phase	50% have normal skin at the site of lesion. May leave fibrofatty residuum, telangiectasia, yellow hypo elastic patches or may rarely scar.Involution is usually complete by 5 yrs of age in 50% of children, 7yrs in 70% and most by 10-12 yrs. [4,5]

Complications

Local complications	Ulceration, hemorrhage, infection and necrosis
According to location	
Periorbital and retrobulbar	Visual axis obscured, astigmatism, amblyopia and optic nerve compression
Auricular	External auditory canal obstruction, otitis externa and hearing impairment
Nasal tip and large facial hemangiomas	Permanent scarring (Cyrano nose) and disfigurement
Mandibular / beard area distribution	Airway obstruction
Perioral	Feeding difficulties, ulceration and disfigurement
Subglottic	Stridor and respiratory failure
Perineal	Ulceration
Multiple hemangiomas	Visceral involvement (liver and GIT)
Large hemangiomas	High-output cardiac failure
Associated anomalies	
PHACES syndrome	Posterior fossa malformations, hemangiomas, arterial anomalies, cardiac defects, eye abnormalities, sterna clefts and supraumbilical raphe
PELVIS syndrome	Perineal hemangiomas and external genitalia malformations, Lipomyelomeningocele, vesicorenal abnormalities, imperforate anus and skin tags
SACRAL syndrome	Spinal dysraphism, anogenital anomalies, cutaneous anomalies, renal and urological anomalies, associated angioma of lumbosacral localization

Diagnosis

It is usually diagnosed based on history and examination. Investigations are deemed appropriate in case of doubtful cases or in case of suspected associated systemic abnormalities. We have to rule out PHACES syndrome (Posterior fossa abnormalities, Hemangioma, Arterial anomalies, Cardiac defects, Eye abnormalities, Sternal cleft and supra umbilical raphe. Line of work up for large facial and scalp Hemangioma, Ultrasound head in infants <4 mon of age and MRI in infants >4mon of age.[4]. Examination and investigation should be done to rule out Multifocal Infantile Hemangioma, Kaposiform Hemangioendothelioma, Kasabach- Merritt Phenomenon, Pyogenic Granuloma, Angiokeratoma of Mibelli, Spider Angioma, Maffucci Syndrome, Hereditary Hemorrhagic Telangiectasia (Osler-Weber-Rendu Disease), Ataxia Telangiectasia and Angiokeratoma Corporis Diffusum (Fabry Disease).[1]

Management

Options for medical therapy of Infantile Hemangioma include topical agents and systemic medications. Propranolol is a non-selective beta blocker, which has become the gold standard first - line therapy for most Infantile Hemangiomas. The goal dose is 1-3 mg/kg/day. Other systemic beta blockers including Nadolol and Atenolol have been shown to also effectively treat Infantile Hemangiomas. Topical Timolol, a non-selective beta blocker can be used for Infantile Hemangiomas. Prednisolone, systemic corticosteroids were the standard therapy before Propranolol Intralesional corticosteroids (Triamcinolone) are consideration for treatment of select, small, isolated and focal Infantile Hemangiomas.[4,6,7,8]

Procedural Management of IH

Pulsed dye laser, gold standard laser treatment for Pediatric vascular lesions has been used in the treatment of Infantile Hemangiomas. Depending on the size and location of an Infantile Hemangiomas, surgical removal may be a consideration.[5]

Discussion

Any infant presenting with multiple Hemangiomas we have to rule out PHACE Syndrome and Kasabach- Merritt Phenomenon. We have ruled out these two major conditions in our case, and started with Tablet Propranolol for which the infant is responding well.

Conclusion

In the past two decades, research and clinical experience have greatly increased our understanding of Infantile Hemangiomas pathogenesis, risk factors and the clinical spectrum of disease. The discovery of beta blockers as an effective therapy has significantly altered the treatment landscape and as pathways contributing to Infantile Hemangiomas development continue to be elucidated, other therapeutic approaches may emerge. Clinicians caring for Infantile Hemangiomas patients should appreciate their natural history and potential complications and initiate treatment as early as possible in an effort to optimize outcomes and minimise morbidities.

Reference

1. Nelson Textbook of Pediatrics. First South Asia Edition. Vol.3, Page No. 3124—3128.
2. LM Burkmiller, GT Richter, JY Suen. Diagnosis and Management of Hemangiomas and Vascular Malformations of the Head and Neck. Oral Diseases (2010)16.405-418.
3. Zia Khan, Elisa Boscolo, Arnaud Picard, et. al. Multipotential stem cells recapitulate human Infantile Hemangioma in immunodeficient mice. The Journal of Clinical Investigation. 118(7),2592-2599, 2008.

4. Anil Abraham, Anupav Mary Job, Gillian Roga. Indian Journal of Dermatology. 2016; Vol.61; Issue:2, Page No. 181-186.
5. Karina L Vivar, Antony J Mancini. Indian Journal of Pediatric Dermatology - Infantile Hemangiomas: An Update on Pathogenesis, Associations and Management. 2018; Vol.19; Issue- 4. Page No. 293-303.
6. Veronique Sans, Eric Dumas da la Roque, et. al. Propranolol for severe Infantile Hemangiomas: follow-up report. American Academy of Pediatrics 124(3). e423-e431, 2009.
7. Christine Leaute - Labreze, Eric Dumas de la Roque, et. al. New England Journal of Medicine. Propranolol for severe Hemangiomas of Infancy. 358(24),2649-2651,2008.
8. Jessica Stiles, Clarissa Amaya, Robert Pham, et. al. Spandidos Publications. Propranolol Treatment of Infantile Hemangioma Endothelial cells: A molecular analysis. Experimental and therapeutic medicine. 4(4), 594-604, 2012.