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Multiple infantile-haemangiomas presenting as severe pulmonary arterial hypertension with desaturation

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Abstract
Two days baby presented with SpO2=81% and multiple cutaneous haemangiomas and systolic murmur. Echo showed Severe TR PG=64mmHg, dilated RA, RV, PFO right to left shunt. USG abdomen showed multiple haemangiomas in liver. Baby was started on Sildenafil and propranolol tablets. Review echo after one month showed No TR, PFO left to right, Normal RA, RV size. SpO2 was 93%.

Keywords: infantile-haemangiomas; pulmonary arterial hypertension; sildenafil; propranolol

Introduction

Case Report

Two days old female child weighing 2.5 kg, born by full term normal vaginal delivery, hemodynamically stable with no respiratory distress presented with SpO2=81% and multiple cutaneous haemangiomas and systolic murmur. Echo showed Severe TR PG=64mmHg, dilated RA, RV, PFO right to left shunt, normal LV, RV function, normal arch, no PDA. USG abdomen showed multiple haemangiomas in liver. Baby was started on Sildenafil @ 1mg per kg per day and propranolol @ 1mg per kg per day tablets. Baby was referred to higher centre where same treatment was continued and baby was discharged. Plan was to do CT abdomen after one month and baby was booked for percutaneous coiling of haemangiomas,

Review echo after one month showed No TR, PFO left to right, normal RA, RV size. SpO2 was 93%. Baby was clinically stable and well thriving. Sildenafil was tapered off and propranolol was continued.

At 3 months of age baby was reviewed, she was gaining weight and had normal saturation. Her ECHO was done which was normal. A decision to continue Propranolol was taken and to keep child in regular follow up.

Discussion

Infantile haemangiomas (IH) are the most common vascular tumour of infancy with an estimated 80,000 annual diagnoses in the United States. The genetic mechanisms underlying IH and the related multi-organ birth defect syndromes, PHACE (an acronym for Posterior fossa brain malformations, segmental facial Haemangiomas, Arterial anomalies, Cardiac defects, Eye anomalies, and sternal clefting or supraumbilical raphe) and LUMBAR (an acronym for Lower body haemangiomas, Urogenital anomalies, Myelopathy, Bone deformities, Anorectal malformations/Arterial anomalies, Renal anomalies) remain unsolved [1]. With advances in next generation sequencing (NGS), genomic alterations have been identified in a wide range of vascular anomaly syndromes.

During foetal life, capillary plexuses morphologically differentiate into arteries, veins, and lymph channels while unneeded vessels are destroyed through apoptosis. If any anomaly occurs in the course of such vascular construction, various types of vascular malformations may develop. For instance, anastomosis between an artery and a vein through a nidus causes an arteriovenous malformation; anastomosis between a main artery and a main vein develops into an arteriovenous fistula; an anomaly in a vein forms a venous malformation; an anomaly in a lymph channel develops into a lymphatic malformation; and an anomaly in capillaries constitutes a capillary malformation [2].

Vascular malformations are also growths of blood vessels. They also are noncancerous. They are present at birth. They're also called birthmarks. But they may not be seen for months or weeks after birth. They grow slowly throughout life. They don't shrink [3]. There are 5 types of vascular malformations. They are: Port wine stains (red or purple in colour), Venous malformations, Lymphatic malformations, Arteriovenous malformations, Mixed malformations, a combination of any of the other types [4, 5]. Treatment for vascular malformations depends on the type of malformation. If your child has a large or life-threatening growth, he or she may need a team of doctors. These can include plastic surgeons, skin doctors (dermatologists), eye doctors (ophthalmologists), and other specialists. Your child may need a combination of treatments. These may include: Laser therapy. This is used for port wine stains. Injection into the vascular malformation. This is used for arterial malformations. Injection of a clotting (sclerosing) medicine. This is used for venous malformations [6, 7].

Conclusion

Keen observation and monitoring with patience is the key to success. Although regression of haemangiomas is common with treatment yet desaturation in our case was compelling us to throw the baby into some interventions. We started oral therapy and waited for response. Baby responded dramatically as described.

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