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Exceptional Triad of Pulmonary Hemangioma, Rapid Involuting Congenital Hemangioma (RICH) and Atrial Septal Defect (ASD): A Case Report

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Abstract

Background: Congenital hemangiomas are rare vascular tumors of infancy classified as rapidly involuting congenital hemangiomas (RICH) and non-involuting congenital hemangiomas. Intrathoracic hemangiomas are extremely rare and pulmonary hemangiomas specifically, are exceptional and usually solitary lesions.

Case presentation: We present in this report the special case of a newborn with a triad of a congenital pulmonary hemangioma, cutaneous RICH and an atrial septal defect (ASD).

Conclusion: This is, to our knowledge, a first case of a triad of congenital pulmonary hemangioma, along with cutaneous RICH and an ASD that can be particularly important in clinical management given that the volume overload associated with an ASD, when added to the lung pathology of pulmonary hemangiomas, can lead long-term to possible pulmonary hypertension.

Keywords: Congenital pulmonary hemangioma; Rapidly-involuting congenital cutaneous hemangioma; Atrial septal defect; Glucose transporter protein

Abbreviations: IH: Infantile Hemangiomas; CH: Congenital Hemangiomas; GLUT-1: Glucose Transporter Protein; ASD: Atrial Septal Defect; RICH: Rapidly-Involuting Congenital Cutaneous Hemangioma; CXR: Chest Radiograph; PNET: Primitive Neuroectodermal Tumor; POD: Post-Operative Day; ISSVA: International Society for the Study of Vascular Anomalies; NICH: Non-Involuting Congenital Hemangioma

Introduction

Hemangiomas are vascular neoplasms that can occasionally cause morbidity and mortality [1]. Infantile hemangiomas (IH) occur in approximately 10% of infants, with a female predominance [2]. Their evolutive pattern comprises an actively proliferating phase followed by spontaneous regression of angiogenesis and complete disappearance of the mass [2]. Some hemangiomas fully developed at birth are called congenital hemangiomas (CH) and do not follow the typical evolutive pattern.

While IH are very common in infancy and childhood, CH are rare and have been only recently described as a distinct entity [3-5]. Hemangiomas can occur in a multitude of locations but intrathoracic hemangiomas are extremely rare [1-6]. When they occur, they arise mainly from the diaphragm, trachea, bronchi and pericardium. Hemangiomas arising from the pulmonary parenchyma are unusual [1-6]. Pulmonary hemangiomas can present as a solitary mass or diffuse interstitial process known as pulmonary hemangiomatosis [6]. There have been many case reports of mediastinal hemangiomas and pulmonary hemangiomatosis but few case reports of solitary pulmonary hemangioma. Moreover, because of the relatively new proper classification of hemangiomas along with the use of the glucose transporter protein (GLUT-1) staining, some of the case reports are indeterminate concerning the type of the hemangioma.

To our knowledge, there are only single digit number of cases of pulmonary parenchymal hemangiomas reported in the litterature of which less than five are solitary pulmonary hemangiomas [7,8]. Moreover, vascular abnormalities of the lung are rarely associated with congenital heart disease [7]. We found only 2 case reports in the literature describing atrial septal defect (ASD) in association with either hemangiomatosis of the lung [1] or solitary pulmonary IH. In this case, we describe the combination of a solitary congenital pulmonary hemnagioma, rapidly-involuting congenital cutaneous hemangioma (RICH) and an ASD. To the best of our knowledge, this is the only case describing the association of those 3 lesions.

Case Presentation

The patient is a 3 days-old female, 2660 g, was born at 36-5/7 weeks of gestation by spontaneous vaginal delivery and was vigorous with APGAR scores of 9/9 (1 and 5 minutes). Her physical exam was significant for no respiratory distress, a grade 2/6 ejection systolic murmur heard in the left precordium, significantly diminished breath sounds on the left side and multiple small hemangiomas on the right upper eyelid, the left earlobe posteriorly and on the posterior aspect of the left knee. On day of life 3, she was found to have a murmur. A chest radiograph (CXR) demonstrating a large left upper lobe mass, an electrocardiogram (right axis deviation, right ventricular dominance) and an echocardiogram (small size secundum ASD and a left chest mass) were obtained.

Tumor markers (human chorionic gonadotropin, alfa feto-protein and urinary catecholamines, dopamine, norepinephrine, epinephrine, homovanilic acid and vanilyl mandilic acid levels) were normal except for mild non-specific elevation of the urinary dopamine and urinary norepinephrine. No masses were found on abdominal ultrasound. CT

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scan of the chest with contrast demonstrated a well-circumscribed left upper lobe mass with lobular border suggestive of pulmonary blastoma measuring 4.5 × 2.9 cm. The possibilities of primitive neuroectodermal tumor (PNET) and fetal interstitial tumor were suggested. The mediastinum and rest of the lungs were normal. Due to intravenous contrast it was difficult to identify any intra tumor calcifications.

Surgical interventions

After a multidisciplinary meeting, a left upper lobectomy was planned. The chest was approached by left thoracotomy in the 4th intercostal space. A 4 cm mass engulfed in the left upper lobe was identified. The great fissure was developed. The pulmonary artery branches including the basilar and lower lobes were cautiously dissected. The lingular and posterior branches going to the upper lobe were divided. A pulmonary vein was dissected anteriorly and was encircled with a vessel loop. The pulmonary artery branches of the upper lobe were identified and divided. The bronchus was mobilized totally and transected with an Endo-GIA stapler. The inferior pulmonary ligament was freed to mobilize the upper lobe. The rest of the lung expanded at 20 cm airway pressure with no leak. The surgical site was secured with 2 pericostal sutures for closure. One chest tube was placed. An anatomical left upper lobectomy was thus performed.

Post-operative period

She was extubated to high flow nasal cannula and weaned progressively to room air in less than 24 hours. A follow up CXR on post-operative day (POD) 2 demonstrated a pneumopericardium which resolved spontaneously on POD4 without hemodynamic compromise. The chest tube was removed on POD5. She tolerated feeds and was discharged home on POD7. At her 5 months follow-up visit, she had a normal growth curve and progressive decrease in the size of all her cutaneous hemangiomas.

Pathology

Grossly, the specimen was received from the left upper lobe and was 7 cm × 5 cm × 3 cm. The sectioning of the radiographically abnormal area revealed cystic spaces surrounded by indurated hemorrhagic tissue. Microscopically the tissue sections showed a discrete intralobular mass composed of diffuse network of sinusoidal vascular spaces that replaced the lung parenchyma. The vascular spaces show a flat endothelial lining without atypia (Figure 1). Immunohistochemistry was performed with appropriate controls, and the endothelial cells are positive for CD31 (Figure 2) and negative for GLUT-1 (Figures 3 and 4). The negative

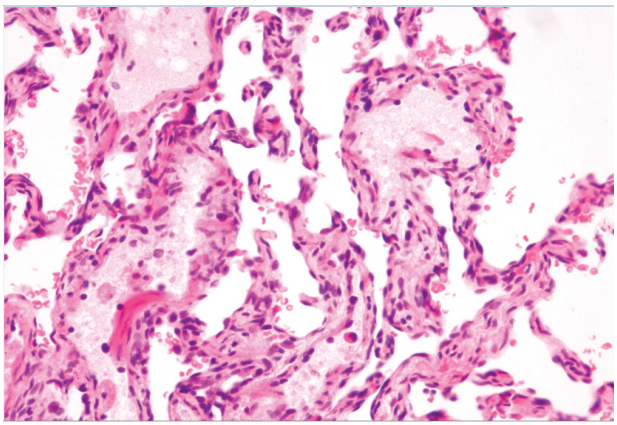

Figure 1: Higher power of pulmonary vascular lesion demonstrating flat endothelial lining (H & E).

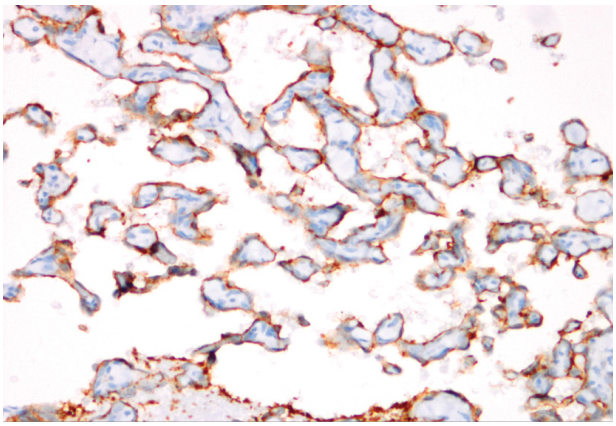

Figure 2: CD31 immunohistochemistry demonstrates positive staining of vascular endothelial lining.

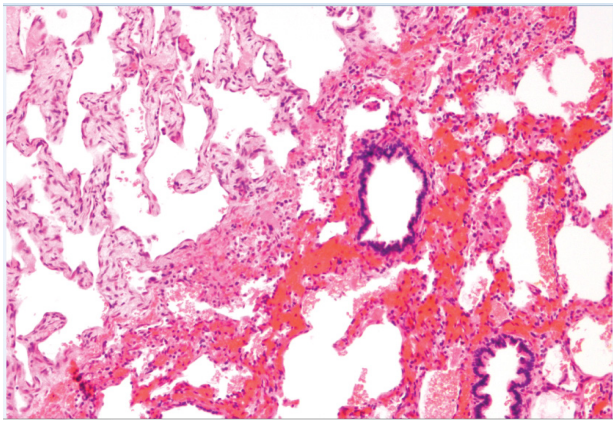

Figure 3: Intralobar pulmonary vascular lesion abutting hemorrhagic pulmonary parenchyma (H & E).

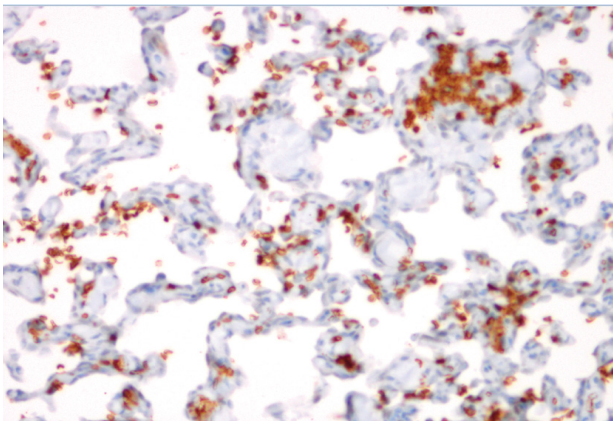

Figure 4: GLUT-1 immunohistochemistry is negative in the vascular endothelial lining with staining of red cells only.

GLUT-1 staining excludes the diagnosis of an infantile hemangioma. Based on the morphology and immunohistochemical findings, this lesion is best classified as a hemangioma-like vascular malformation.

Discussion

The current classification of vascular anomalies adopted by the International Society for the Study of Vascular Anomalies (ISSVA)

entitles two distinct entities: congenital vascular malformations and vascular tumors (IH and CH). IH and CH are fundamentally different, not only in their anatomical, histological and pathophysiological findings, but also in their clinical course [1]. IH present within a few weeks of life [6] while CH is vascular tumors that present at birth, reach their maximum size before birth (having undergone their proliferative phase in utero) and do not exhibit accelerated or disproportionate postnatal growth [3]. CH is now classified into RICH and non-involuting congenital hemangioma (NICH) [4]. Mulliken and Enjolras have characterized the growth curves of RICH and NICH compared with IH (Figure 3) [3]. CH usually present as solitary lesions at birth and rarely coexist in the same patient with a typical IH [3,7]. They constitute a rare group of benign vascular tumors, with a combined incidence of less than 3% of all IHs [3].

GLUT-1, an erythrocyte-type glucose transporter protein, has emerged as a valuable immunohistochemical marker for distinguishing different types of hemangiomas [7] and is expressed by IH of the skin and extracutaneous sites such as liver and breast, but not by vascular malformations, CH or by the normal vasculature of the skin [7]. Absence of GLUT-1 expression in CH, in contrast to IH, provides further evidence that CH and IH represent distinct entities. Intrathoracic hemangiomas are extremely uncommon [6]. These have previously been described as arising from the diaphragm, bronchus, trachea, and pericardium [6,7]. They may also very rarely arise within the lung manifesting as pulmonary hemangiomas [6,7]. The most common entity is pulmonary capillary hemangiomatosis, comprising multiple infiltrative foci of capillary proliferation [7].

Our patient had an exceptional triad of a pulmonary CH, cutaneous RICH and an ASD. Only 5 cases of pulmonary CH were described in the literature and only 1 case was associated with cutaneous CH. As previously mentioned, CH usually present as solitary lesions at birth. In our case, the pulmonary CH co-existed with cutaneous CH that are most probably RICH as described by their clinical evolution, although a biopsy with immunostaining would have been of better value for diagnosis.

In this case, the chest mass was an incidental finding while the usual presentation of the pulmonary CH is with respiratory symptoms like cough, and respiratory distress. On another hand, a pulmonary CH and an ASD co-existed in our patient. Association of vascular malformations with congenital heart disease (CHD) has been described in systemic disorders such as Osler-Weber-Rendu syndrome as is the association between capillary proliferation and CHD in PHACES syndrome [7]. ASD have been reported in patients with hepatic and sacral hemangiomas, cutaneous and visceral hemangiomatosis but has been reported only once associated to pulmonary CH [7]. Although the presence of ASD is a common finding in infants and therefore it is possible that this is a pure coincidence of coexistence of 2 lesions in the same infant; the co-existence of ASD and pulmonary CH might be an association and is worth investigating especially that the volume load on the right side of the heart in the context of an ASD when associated with a pulmonary lesion may lead to pulmonary hypertension. In our case, the ASD was small and there was no pulmonary hypertension; but highlight on this association is worth it as lager ASD along with pulmonary hemangiomas may lead to pulmonary hypertension.

Conclusion

Resection was undergone with left thoracotomy with no need for sternotomy and cardio-pulmonary bypass since its size and its blood supply (only from the hilum) are in favor of this surgical technique. This of course substracted not only the risks associated with cardio-pulmonary bypass but the overall risk of surgery. We have reported the case of a newborn with an incidental finding of a congenital pulmonary hemangioma, along with cutaneous RICH and an ASD. This triple association has never been reported to our knowledge.

Declarations

Acknowledgments

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Availability of data and materials

Data other than presented in this case report and that can be anonymized are available from the corresponding author on reasonable request.

Author's contributions

SG and LD performed the clinical examination and interpreted the patient data together with LS and TM, who were responsible for the evaluation of the immunohistochemical reactions on our specimen.

SG and LD were major contributors to the writing of the manuscript. LS and TM participated in the review of the manuscript and helped with editing. All authors read and approved the final manuscript.

Ethics approval and consent to participate

This report was approved by the Ethics Committee of Driscoll Children's Hospital, Texas, USA.

Consent for publication

Verbal informed consent was obtained from the patient's legal parent for publication of this case report and any accompanying images.

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