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Congenital Hemangiomas

Developed by Emilie Wang and Dr. Joseph Lam for PedsCases.com.
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Introduction

Hello everybody, and welcome to this podcast on congenital hemangiomas, brought to you by PedsCases. My name is Emilie Wang, and I am a medical student at the University of British Columbia. In this episode, we will be discussing congenital hemangiomas, their pathogenesis, presentation, and management. I'd like to thank Dr. Joseph Lam for helping to create this podcast with me. Dr. Joseph Lam is a pediatric dermatologist at BC Children's Hospital.

Hemangiomas are benign vascular tumors commonly found in infants. These tumors are characterized by an abnormal proliferation of blood vessels. Infantile hemangiomas are the most common type of hemangiomas in children. They are typically not present at birth and appear after birth. They grow during the first few months of life. PedsCases has a separate podcast on Infantile Hemangiomas. In contrast, congenital hemangiomas are a separate entity that are fully formed at birth. This is the focus of our podcast today!

Learning Objectives

After listening to this podcast, learners should be able to:

1. Define hemangiomas and congenital hemangiomas.
2. Explain the genetics and environmental factors associated with congenital hemangiomas.
3. Describe the pathogenesis of congenital hemangiomas.
4. Classify congenital hemangiomas.
5. Discuss the diagnosis, differential diagnosis, and management strategies for congenital hemangiomas.

Clinical Case

Let's introduce an example case to better understand congenital hemangiomas. Anna is a 2-day-old neonate born at term. Her parents noticed a large purple lesion on her left thigh at birth. The lesion is well-circumscribed, firm, and measures about 5 cm in diameter. It does not appear to be growing, but it has a raised central area with a pale halo around it. Anna's parents are concerned, and her pediatrician refers her to dermatology for further evaluation.

What conditions could present with a large, purple lesion on a newborn's thigh? Take a moment to consider your differential diagnoses.

We'll discuss the diagnosis later in the podcast!

What are congenital hemangiomas?

Congenital hemangiomas (CHs) are a distinct subtype of hemangiomas, which are present at birth and have a different growth pattern than infantile hemangiomas. They do not undergo postnatal growth, and their clinical course differs significantly.

Congenital hemangiomas are classified into two main types: (1)

1. Rapidly Involuting Congenital Hemangiomas (RICH): These tumors involute rapidly within the first 6-14 months of life, often leaving behind excess skin or a fibrofatty mass. (2)
2. Non-Involuting Congenital Hemangiomas (NICH): These lesions do not regress and may even grow proportionally with the child.

A third rare variant is Partially Involuting Congenital Hemangiomas (PICH), which display characteristics of both RICH and NICH, with partial involution over time. A fourth rare variant is tardive expansion congenital hemangioma, shortened as TECH. This type of hemangioma arises prenatally and exhibits a proportional growth pattern similar to that of NICH, but a tardive expansion of the lesion, where "tardive" means a late development or progression, happening later in childhood. (1)

Genetics and Environmental Associations

The exact genetic causes of congenital hemangiomas are not fully understood. However, mutations in certain signaling pathways, such as the Ras/Raf/MAPK pathway, may contribute to their development. Environmental factors, including hypoxic injury during fetal development, have also been suggested as potential contributors.

Pathogenesis

Congenital hemangiomas are thought to result from abnormal vasculogenesis during embryonic development. Unlike infantile hemangiomas, which stem from endothelial cell proliferation, congenital hemangiomas develop fully in utero and do not proliferate after birth. (3) They can be detected in utero through prenatal ultrasound by the end of the first trimester or the beginning of the second trimester. RICH and NICH cannot be distinguished on ultrasound. (4) These lesions do not express GLUT1, which is a hallmark of infantile hemangiomas. (2) The angiogenesis in congenital hemangiomas is more organized, contributing to their different clinical behavior.

Morphology

Congenital hemangiomas typically present as well-defined, raised lesions with a pale or purplish hue. RICH lesions tend to be large, bulky, and may have a central area of involution with a surrounding pale rim, while NICH lesions remain stable in size and color over time. (5) Both types are fully developed at birth and do not show the same proliferative phase as infantile hemangiomas. (5)

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Extracutaneous Manifestations

Although congenital hemangiomas are primarily cutaneous, they can occasionally have extracutaneous involvement. These include cardiac overload due to high-output heart failure, especially if the hemangioma is large or highly vascular. (6) In this circumstance, the hemangioma acts as a left-to-right shunt, increasing the volume of blood returning to the right heart, leading to volume overload. (7)

Diagnosis

The diagnosis of congenital hemangiomas is primarily clinical, based on their presence at birth and characteristic appearance. In some cases, imaging such as Doppler ultrasound or MRI may be used to assess the extent of the lesion and any involvement of deeper structures. (6) Biopsy is rarely required but may be performed in ambiguous cases. (6)

Differential Diagnosis

It is important to differentiate congenital hemangiomas from other vascular anomalies such as infantile hemangiomas, kaposiform hemangioendothelioma, and pyogenic granulomas and malignant soft-tissue tumours with prominent vascularization such as rhabdomyosarcomas and myofibromas. We'll talk about a few more details here, and you should be able to recognize common patterns of congenital hemangiomas versus more concerning vascular tumors like kaposiform hemangioendothelioma or rhabdomyosarcoma. However, because some of these distinctions can be subtle, and given the potential for misdiagnosis, referral to dermatology (or possibly pediatric hematology-oncology) will probably be the safest if there is any uncertainty.

1. Infantile Hemangioma

How does it present? Unlike congenital hemangiomas, infantile hemangiomas usually appear after birth and grow rapidly during the first few months.
How can we differentiate it? Ask parents if the lesion was present at birth or developed later. If it developed a few weeks postnatally and showed a growth phase, it's more likely to be an infantile hemangioma. (6)
Here's a learning point! Remember that congenital hemangiomas are fully formed at birth, while infantile hemangiomas typically develop later and go through a growth phase!

2. Kaposiform Hemangioendothelioma (KHE)

How does it present? A rare, locally aggressive vascular tumor that often presents with a firm, purplish mass, which can be associated with Kasabach-Merritt phenomenon (severe coagulopathy). (8)
How can we differentiate it? Look for clinical signs like coagulopathy (petechiae, bleeding) and assess for rapid growth or complications. Imaging may reveal a more infiltrative pattern, and lab tests may show thrombocytopenia. This is a rare, but can't-be-missed diagnosis.

Here's a learning point! When a lesion is associated with systemic symptoms like bleeding, always consider KHE and order a coagulation profile.

3. Pyogenic Granuloma
How does it present? Pyogenic granulomas are small, bright red, and bleed easily. They often develop rapidly after minor trauma. (9)
How can we differentiate it? Ask about recent trauma or irritation at the lesion site, and assess if the lesion bleeds with minimal provocation. Pyogenic granulomas are typically smaller and more friable compared to congenital hemangiomas. Friable refers to tissue or lesions that are easily broken, crumbled, or bleed with minimal manipulation or trauma. In a clinical context, a friable lesion is one that can bleed with only a little bit of rubbing or contact. For instance, a pyogenic granuloma is often described as friable because it tends to bleed easily when touched or slightly disturbed. This characteristic can help differentiate between different types of vascular lesions or growths. (10)
Here's a learning point! Pyogenic granulomas usually appear in response to trauma and rarely, can be congenital. If the lesion bleeds excessively, suspect this differential.

4. Infantile Myofibromatosis
How does it present? Infantile myofibromatosis is the most common fibrous tumor of infancy, presenting as firm, nodular lesions that can appear on the skin, subcutaneous tissue, muscle, or bone. (11) These lesions are usually painless and can be solitary or multiple. In cases with visceral involvement, there may be symptoms related to the affected organs, such as respiratory or GI issues. (11) They can be congenital, present on neonates in the first few months of life, or infants that are less than 24 months.
How can we differentiate it? Unlike vascular lesions like pyogenic granulomas or congenital hemangiomas, myofibromatosis lesions are firmer, less friable, and do not bleed easily. They tend to grow more slowly and are not as vascular, so they lack the bright red color and propensity to bleed with minor trauma. Imaging (such as ultrasound or MRI) often shows a well-circumscribed mass with a heterogeneous internal structure, and biopsy reveals spindle-shaped cells typical of fibromatous tumors. (12)
Here's a learning point! Infantile myofibromatosis can be mistaken for other soft-tissue masses, but its firm, non-friable nature and lack of rapid growth or bleeding are distinguishing features. Additionally, cases with multiple lesions or visceral involvement require careful monitoring and may suggest a more systemic form of the disease. Another key differential to consider, is congenital primary cutaneous rhabdomyosarcoma in a neonate. This is an aggressive malignancy that grows out of muscle cells. Rhabdomyosarcoma lesions exhibit areas of increased vascularity, leading to a somewhat similar appearance to congenital hemangiomas. They are also a rare cause of the "blueberry muffin baby", which means that there are multiple blue nodules on the baby, which often include other differentials of infections. This one will grow rapidly and aggressively. Biopsies and imaging would be required, if there is diagnostic uncertainty. This one is a serious one, and although rare, it cannot be missed. (13)

The first four differentials, including congenital or infantile hemangiomas, kaposiform hemangioendotheliomas, also called Kasabach-Merritt syndrome, pyogenic granulomas, infantile myofibromatosis can be seen on DermNetNZ and Compendium Vascular Anomalies. We have attached the links for you to take a look, as learning visually is so important in dermatology. https://dermnetnz.org/topics/infantile-haemangioma-definition-and-pathogenesis https://www.compva.com/patientexamples/kaposiform-hemangioendothelioma-of-the-trunk-with-bleeding https://dermnetnz.org/topics/pyogenic-granuloma https://dermnetnz.org/topics/infantile-digital-fibroma

Management

The management of congenital hemangiomas depends on the subtype and any associated complications. RICH lesions are usually managed conservatively, as they involute spontaneously. NICH lesions may require surgical excision if they cause functional or cosmetic concerns. (14) In rare cases, large hemangiomas causing cardiac complications may need medical intervention such as embolization. (6)

Case Discussion

Returning to our case: Here are some differentials to a large purple lesion on an infant's thigh. You complete a detailed history and exam. You confirm that the lesion was present at birth, making an infantile hemangioma unlikely. She has no history of bleeding/petechiae that could be consistent with Kasabach-Merritt phenomenon. No systemic complications such as high output heart failure is observed, or any bone and visceral involvement that could be indicative of infantile myofibromatosis. There was no evidence of coagulopathy or aggressive growth. We know that kaposiform hemangioendotheliomas are firm, infiltrative, and non-involuting, while congenital hemangiomas are generally well-demarcated, and e also see some signs of involution here. With regards to pyogenic granulomas, we know that those develop later in infancy or childhood, and are not congenital. They're small, friable, and prone to bleeding, and will not spontaneously involute or resolve. Infantile myofibromatosis, as we mentioned, are skin-coloured or bluish, while Anna's case is a deep purple vascular appearance.

Because we see that it does not appear to be growing and has a raised central area with a pale halo around it, we recognize that it matches the description of a RICH, having a central area of involution with a surrounding pale rim. Anna's lesion is therefore clinically diagnosed as a rapidly involuting congenital hemangioma (RICH). Since it is not causing any functional issues and is expected to involute on its own, the dermatologist advises her parents to monitor the lesion closely and return for follow-up if there are any changes in size, color, or if Anna develops any complications.

Takeaway points

Let's wrap up with a few take-away points. For those interested, you can find images of congenital hemangiomas on DermnetNZ to help visualize the differences between RICH and NICH.

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1. Congenital hemangiomas are benign vascular tumors that are fully developed at birth, unlike infantile hemangiomas.
2. RICH and NICH are the two main types of congenital hemangiomas. RICH tends to involute, while NICH remains stable.
3. The diagnosis is primarily clinical, with imaging reserved for complicated or deep lesions.
4. Management is non-surgical for most RICH lesions, while NICH lesions may require surgical intervention.
5. The differential diagnosis includes rare but can't-be-missed diagnosis for malignancy is primary cutaneous rhabdomyosarcoma and kaposiform hemangioendothelioma.

That concludes today's podcast on congenital hemangiomas. Thanks for listening!

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