HIGH-FLOW ARTERIOVENOUS MALFORMATION WITHIN ENLARGED FETAL LEG (Congenital Hemangioma vs Parkes Weber Syndrome)

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VASCULAR ANOMALIES

- Most common cause of pediatric soft tissue masses
- In utero detection/prenatal diagnosis possible
- Solid / Cystic / Mixed
- Gray scale and color Doppler USG, MRI
- Old confusing nomenclature
- Effective diagnoses and therapeutic approaches
## ISSVA Classification of Vascular Tumors and Malformations

<table>
<thead>
<tr>
<th>Tumors</th>
</tr>
</thead>
<tbody>
<tr>
<td>Infantile hemangioma</td>
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<tr>
<td>Congenital hemangioma</td>
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<tr>
<td>Rapidly involuting congenital hemangioma</td>
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<tr>
<td>Noninvoluting congenital hemangioma</td>
</tr>
<tr>
<td>Kaposiform hemangioendothelioma</td>
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<tr>
<td>Tufted angioma</td>
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<tr>
<td>Hemangiopericytoma</td>
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<tr>
<td>Pyogenic granuloma</td>
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<tr>
<td>Spindle cell hemangioendothelioma</td>
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</tbody>
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<table>
<thead>
<tr>
<th>Malformations</th>
</tr>
</thead>
<tbody>
<tr>
<td>Simple (ie, venous, lymphatic, capillary, and arterial)</td>
</tr>
<tr>
<td>Combined (eg, arteriovenous)</td>
</tr>
</tbody>
</table>
Growth

Vascular tumors

Infantile hemangioma

NICH

RICH

Vascular malformations

Fetus  Birth  1  5  Years
VASCULAR ANOMALIES

✓ **Vascular tumors**: neoplastic/proliferative growth of vascular endothelial cells

✓ **Vascular malformations**: structural developmental anomalies of hematic or lymphatic vessels

✓ Isolated / together / within the phenotype of a syndrome

PROGNOSTIC PARAMETER → FLOW VELOCITY
Vascular anomalies

Vascular Tumors
- Hemangioma
- Other tumors:
  - Kaposiform hemangioendothelioma
  - Angiosarcoma
  - Others

Vascular malformations
- Venous (VM)
- Lymphatic (LM)
- Arterial (AM)
- Capillary (CM)
- AVM
- AVF
- Combined (CLVM, VLM, CLAVM, CVAVM)

Vascular anomalies based on type of flow

Low-flow
- Vascular malformations: VM, LM, CM, CLVM, VLM

High-flow
- Vascular tumors
- Vascular malformations: AM, AVM, AVF, CLAVM, CVAVM
VASCULAR ANOMALIES
(Flow Velocity)

✓ **Prenatal follow-up:**
  - High flow lesions $\rightarrow$ High output congestive heart failure (fetal cardiomegaly, atrioventricular regurgitation, abnormal ductus venozus Doppler, hydrops), Kasabach Merritt sequence

✓ **Postnatal treatment:**
  - High flow lesions $\rightarrow$ embolization
  - Low flow lesions $\rightarrow$ sclerotheraphy
Flow

Artery

Mass

Phlebolith

1. Vascular tumor (including infantile hemangioma)
2. Other soft tissue tumor

High-flow vascular malformation (including AVM)

Venous malformation

1. Low-flow vascular malformation (including VM before appearing of phlebolith)
2. Other soft tissue mass

1. Lymphatic malformation
2. Other cystic lesions

solid vs. cystic

high flow vs. low flow

tumor vs. malformation
PARKES WEBER SYNDROME

✓ 1907, Frederick Parkes Weber
✓ Similar to Klippel Trenaunay Syndrome
✓ Very uncommon, exact prevalence ???
✓ Multiple arteriovenous connections (AVC) (malformations, fistulas)
✓ Overgrowth of one limb (most commonly a leg)
✓ Capillary malformations
✓ RASA 1 gene (5q14.3), sporadic
✓ Life threatening even fatal
<table>
<thead>
<tr>
<th>Differential diagnosis</th>
<th>Pulsed Doppler US findings</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Low-flow sonographic pattern</strong></td>
<td>No detectable flow</td>
</tr>
<tr>
<td>a) Low-flow vascular malformations</td>
<td></td>
</tr>
<tr>
<td>1. Classic pattern for LM, VLM, VM and CM</td>
<td></td>
</tr>
<tr>
<td>2. Combined low-flow vascular malformation with limb hypertrophy or hemimegalencephaly</td>
<td></td>
</tr>
<tr>
<td>b) Other lesions, e.g., cystic teratoma, dermoid cyst</td>
<td></td>
</tr>
<tr>
<td><strong>High-flow sonographic pattern</strong></td>
<td></td>
</tr>
<tr>
<td>a) Vascular tumors: congenital hemangioma, kaposiform hemangioendothelioma</td>
<td>Low impedance arterial flow</td>
</tr>
<tr>
<td>b) Tumors with vascular appearance: teratoma, congenital-infantile fibrosarcoma, infantile myofibromatosis</td>
<td>Low impedance arterial flow</td>
</tr>
<tr>
<td>c) High-flow vascular malformations</td>
<td>Low impedance arterial flow and pulsatile veins</td>
</tr>
<tr>
<td>1. AVF, AVM (no mass associated)</td>
<td></td>
</tr>
<tr>
<td>2. Combined high-flow vascular malformation with limb hypertrophy</td>
<td></td>
</tr>
<tr>
<td>d) Low-flow vascular malformation simulating a high-flow lesion</td>
<td>Low impedance arterial flow</td>
</tr>
</tbody>
</table>

CASE REPORT

• 24-year-old, primigravid
• Referred to our clinic upon detection of a fetal leg mass on regular sonographic evaluation at 29th week of her gestation
• No follow-up during pregnancy: including aneuploidy screening and detailed midtrimester scanning
• No significant maternal risk factor
CASE REPORT

• Ultrasound examination at 29-weeks 3-days revealed 45 x 44 mm soft tissue lesion on posteromedial side of left fetal calf demonstrating high-flow arterial feeders, varicose-tortuous venous drainers reaching up to and widening fetal thigh and turbulent AV connections in between.

• With progressing gestational age crural size and intercrural discrepancy increased and the lesion size reached to 70 x 55 mm at term.
CASE REPORT

• Doppler measurements stayed within normal limits with no sign of fetal cardiomegaly or hydrops.
• The parents were informed about the probable diagnoses of Congenital Hemangioma (CH) or PWS.
• At 39w3d, 3520 g female baby was delivered.
• MRI confirmed an isolated CH, but with AVC, and the lesion was treated successfully via microcatheter arterial embolization.
• Karyotype analysis was normal. RASA 1 → ?
CONCLUSION

- The complex spectrum of vascular anomalies → misleading / confusing for physician / parents.
- Prenatal presence of an arterial (high) flow with clues of AVC should evoke the suspicion of 3 major conditions:
  - CH → if prominent exophytic mass
  - PWS → if predominant limb overgrowth and multiple AVCs
  - Isolated AV malformation → if absence of both
- A meticulous and thorough prenatal workup is necessary for correct diagnosis and rewarding prognostic counseling.
Thank you...