Imaging Methods in Pediatric Dermatology - How to Approach and their Differential Diagnosis

Poster No.: C-1900
Congress: ECR 2016
Type: Educational Exhibit
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Keywords: Tissue characterisation, Diagnostic procedure, MR, CT, Ultrasound, Musculoskeletal soft tissue, Soft tissues / Skin
DOI: 10.1594/ecr2016/C-1900

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Learning objectives

The use of imaging methods for the evaluation of skin lesions in Pediatrics has increased over the last few years. A great number of superficial lesions in soft tissues have been observed daily and a systematized evaluation may either help achieve a definitive diagnosis or shorten the differential diagnosis.

Through an approach which associates the location of the lesion (epidermis and dermis, subcutaneous and muscular fascia / deep tissues) with the characteristics of imaging in different methods, the goal is to:

- Offer a broad view of the most important lesions in soft spots in Pediatrics
- Illustrate findings related to the imaging of these lesions and differential diagnosis

Background

The clinical history and the ectoscopy are to start the assessment and, if necessary, complemented by imaging.

The first approach by imaging in soft stop lesions in children is ultrasound with high frequency linear transducer probe. Regular US shows a hyperechoic line on the interface between the transducer and the skin, which corresponds to the epidermis. Right below, the derm, a less echogenic layer. Underneath it, it is still possible to see the hypoderm and subcutaneous tissue, the last one hypoechoic with permiating hyperechoic lines (Fig 1).

Low frequency transducers could be useful as it allows a wider field of view and depth in large lesions in deep layers. In cases like this, imaging methods such as CT scan and MRI would be important to help complement assessing the extension of the compromised area and help with differential diagnosis.

Images for this section:
**Fig. 1:** US scan of the skin showing its layers

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Findings and procedure details

We have reviewed patients who had been sent with need of investigation of superficial alterations (bulging soft spots / skin color change).

Along with clinical findings, a US assessment had been used in order to appoint the location, size, depth, vascular pattern on Doppler and infiltration signs / complication. At previous diagnosis, some of these parameters had been used for follow-up and monitoring of therapeutic response.

1. Lesions in skin appendages

# Inclusion cyst

- ectodermal sequestration at line closures
- location: intracerebral, cervical region, face around eyes and scalp
- unilocular, capsulated, slow growing due to accumulation of skin products
- size: 1 to 4 cm
- Both genders, children and young teenagers

--> Dermoid cyst (Fig 1 - 2):

- Type 1: Periorbital in the region where embryonic processes of upper jaw and lateral nasal join at nasolacrimal groove (40-70%)
- Type 2: nasal bridge at frontonasal (8-13%)
- Type 3: submentonian, floor of the mouth and suprahyoid region (9-24%)
- Type 4: Thyroid, supraesternal or suboccipital (5-14%)

* The main differential diagnosis is encephalocele (Fig 4 - 6).

The Crista Galli is essential for differential diagnosis as if it is present but split, the diagnosis is dermoid cyst, and if it is absent or eroded and blind foramen is enlarged, the possible diagnosis is encephalocele.

* Another important differential diagnosis is the sinus pericranii, a collection of nonmuscular scalp veins that communicate with the intracranial venous sinuses. It is most often found in the frontal and parietal region (Fig 7).

# Pilomatrixoma (Fig 8 - 9)

- 60-70% appear before 20 years of age
10% of skin tumors in children

Benign appendage tumor, lobulated, irregular and sometimes hard, 0.5 - 5.0 cm

Solitary, more than 50% in head, neck and upper extremities. Multiple in 12% of the cases.

75% with calcium deposits, 15-20% with ossification

Oval or round

Heterogeneous with echogenic focus that vary according to progress

Hypoechoic halo

Sparse microcalcifications esparsas or complete calcifications

Dermal, hypodermal or dermal-hypodermal

2. Mesenchimal tumors

# Lipoma (Fig 10 - 12)

- It is the most common soft tissue tumor
- Benign, made of mature adipose tissue, less than 5 cm in 80% of the cases
- Superficial (hypoderm) or deep
- The presence of capsule is variable
- Echogenic aspect is variable, determined with the water-fat rate (cellularity): hyperechoic, isoechoic, hypoechoic or mixed echogenicity
- Contourn / outlines: well defined 60%, poorly defined 40%

# Lipoblastoma / Lipoblastomatosis (Fig 13)

- Present mature and immature shapes of adipose tissue and other mesenchimal elements poorly identified
- Infiltrating lipoblastomatosis committing more profound plans
- 88% in children under 3, sometimes at birth
- # > #
- Localization: extremities

4 - Vascular tumors:

# Hemangiomas (Fig 14 - 15)

- It is the most common soft tissue tumor in childhood.
- # > # (3:1), premature babies and patients with family history
- Maculas, plaques or nodules 1 mm - 20 cm.
- Only one or multiplex (visceral).
- Dynamic progress:
Absent at birth

Proliferation (3-12 meses)

Stationary period

Involution (after 2 years)

# Vascular Malformations

- Anomalies in the morphologic development of the embryonic vascular system which occurs between the fourth and the tenth week of intrauterine life
- Might # after trauma, sepsis e hormonal stimulation.
- # = #
- Do not involute and might lead to veno ischemia stasis, skeletal anomalies and local consumption coagulopathy
- Classified as high flow (MAV) and Low flow (capilar, venous, lymphatic or mixed)

# Venous Malformation

- Usually bluish lesions, soft, comprehensive, non-throbbing and increase with Valsalva manouver
- Do not involute and might require surgery
- Calcifications are usual as dystrophic mineralizations or phleboliths

* Differential diagnosis from venous fistula (Fig 18 - 19)

# Lymphangioma

- Congenit obstruction of lymphatic drainage
- Classified according to the size of the loculations:
  - capilar (rare, subcutaneous),
  - cavernous (around the mouth and tongue)
  - cystic
    - Cervical 75%, axillary 20% of the cases
    - Present at birth 50-65%, 90% diagnosis before 2 years of age
    - Acute symptoms: infection, hemorrhage or rupture
US = cystic lesion multiloculated with septums of different thickness, sometimes vascularized / cyst content of uncertain echogenicity

5 - Cutaneous metastasis (Neuroblastoma) - Fig 21 - 22

6 - Neurocutaneous syndromes

# Tuberous sclerosis (Fig 23 - 25)

- Autosomal dominant
- Distinguished for the presence of hamatomatous tumors in multiple organs
- Diagnosis: physical examination and radiological findings
- Classic triad: epilepsy/ mental retardation/ Adenome
- Cutaneous findings:
  - Hypopigmented maculas (90%)
  - Facial angiofibromas ("adenoma sebaceum") 75% "butterfly wings"
  - Nail Fibromas (20%)

- Adenoma sebaceum is the only major criterion for diagnosis that can be detected by physical examination

# Neurofibromatosis (Fig 26 - 29)

- It is the most common of the phakomatosis 1: 2,000
- Autosomal dominant (50% esporádico)
- Compromises skin, nervous system, bones, endocrine glands
- Classic tumor outside central nervous system is neurofibroma, more specifically plexiform
- 30% of the patients with solitary neurofibroma will develop NF1
- All patients with multiple neurofibromas or plexiform have the disease

- Criteria for diagnosis # 2 or +:
  - Six or more café-au-lait spots > 5 mm
  - One neurofibroma plexiform or two or more any type of neurofibroma
  - Two or more iris pigmented hamatomas
  - Freckles in axillar or inguinal regions
Optic nerve glioma

First relatives with NF1

Distinguished bone lesion (sphenoid great wing displasia, pseudo arthrosis)

--> Another findings

Subcutaneous neurofibromas

Focal Thoracic Scoliosis

Posterior vertebral scalloping

Neural foramen enlargement

Remodeling and displasia of costal arches

# Other less common syndromes:

- Stuge Weber: Multiple capillary telangiectatic plexus, thin-walled, lie along the subarachnoid space of the brain. Poverty of normal veins, lead to venous stasis, congestion and cortex hypoxia with consequent atrofia and calcification.

The most frequent radiological manifestation is intracranial, unusual calcification before two years (Fig 30).

- Cutis marmorata telangiectatica congenita: Uncommon, sporadic, congenital cutaneous vascular anomaly evident as persistent cutis marmorata, telangiectasia, and phlebectasia (Fig 31 - 32).

Images for this section:
**Fig. 1:** US scan of the skin showing its layers
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**Fig. 2:** Dermoid cyst: US - Male, 1 moth - Bulge of the right nasolacrimal groove
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Fig. 3: Dermoid cyst: CT - Bulge of nasofrontal region and a small depression anterior to the crista galli

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Fig. 4: Encephaloceles: CT - Frontonasal encephaloceles form when the fonticulus nasofrontalis remains patent.

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Fig. 5: Atretic encephalocele: Prenatal US - (a) Parieto-occipital bulge; (b) Power Doppler without blood flow

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Fig. 6: Atretic encephalocele: (a) Prenatal MRI - mid-line subcutaneous cystic lesion. (b) Neonatal MRI - Vertically oriented persistent primitive falcine vein and a prominent superior cerebellar cistern.

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Fig. 7: Sinus pericarini - CT: Female, 6 month - Nodule with soft tissue density and contrast enhancement, in the subcutaneous of parietal region, associated with prominent vascularization converging to the injury, suggesting vascular malformation. It is also observed discontinuity of the adjacent skull, observing contiguity of the lesion with the sagittal sinus, suggestive of sinus peri crani.

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**Fig. 8:** Pilomatricoma: Female, 10 years - Arm X-ray - Soft tissue calcification.

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Fig. 9: Pilomatrixoma: Female, 10 years. US - Well-defined ovoid lesion at the junction on the dermis and subcutaneous fat. It may have a blood flow at color Doppler.

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**Fig. 10:** Lipoma: US - (a) Abdominal wall and (b) retroauricular region - Variably echogenic masses, well-defined, which don't have an acoustic shadow and no or minimal colour Doppler flow.

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**Fig. 11:** Lipoma in between the muscles of the left chest wall - US: Solid lesion with ill-defined margins, heterogeneous and isoechoic to adjacent subcutaneous tissue. Extension of lesion to deep planes cannot be determined.
Fig. 12: Lipoma in between the muscles of the left chest wall - (a) MRI; (b - c) CT - CT bone reconstruction - defined image with fat-density in subcutaneous tissue, extending to deep planes with muscular tissue interposition. No bone erosion is seen.
Fig. 13: Liposarcoma: US - Ovoid well-defined lesion in subcutaneous plane. Color Doppler shows arterial and venous flow

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Fig. 14: Hemangioma - US

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Fig. 15: Hemangioma: Female, 3 months - (a) Progressive swelling of the neck after birth and laryngeal stridor. (b - c) CT and 3D CT reconstruction- An extensive vascular soft tissue mass is seen in the neck.

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Fig. 16: Arteriovenous malformations - Female, 12 years - Red lesion in the lower back. It was present at birth without significant evolution during the years, associated with hyperthermia and local sweating.

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**Fig. 17:** Arteriovenous malformations - Female, 12 years: Axial CT and 3D reconstruction

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Fig. 18: Collateral vascularization after deep vein thrombosis - Male, 1 year - Bulging on the lateral aspect of the right ankle.
**Fig. 19:** Collateral vascularization after deep vein thrombosis - Male, 1 year - Thickening of the subcutaneous tissue with serpiginosas tubular images permeating, extending to the proximal portion of the calf. The color Doppler shows flow compatible with venous spectral pattern.

**Fig. 20:** Lymphangioma - Female, 1 year - Left cervical region swelling: US - Complex fluid collection with peripheral colour Doppler flow.
Fig. 21: Neuroblastoma - case 1: US

Fig. 22: Neuroblastoma - case 2: CT

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Fig. 23: Tuberous sclerosis - Female - 4 month: Hypochromic spot

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Fig. 24: Tuberous sclerosis - Subependymal nodules (a) tranfontanelar US - (b) CT - (c) prenatal MRI

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Fig. 25: Tuberous sclerosis - Multiple bilateral renal cysts - (a) US - (b) CT

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Fig. 26: Neurofibromatosis - cafe au lait spots

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Fig. 27: Neurofibromatosis - US - Subcutaneous neurofibromas in the right supraclavicular and posterior cervical regions.

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Fig. 28: Neurofibromatosis - MRI - Dural ectasia

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Fig. 29: Neurofibromatosis- X-Ray - Focal thoracic scoliosis and pseudoarthrosis of the tibia

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Fig. 30: Stuge Weber

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**Fig. 31:** Cutis marmorata telangiectatica congenita - CT: Giriforme calcification in the cerebral cortex, especially in the left temporal lobe.

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**Fig. 32:** Cutis marmorata telangiectatica congenita - MRI: Global atrophy of the cerebral hemispheres with asymmetry, down mainly on the left.
Conclusion

The diagnosis of superficial skin lesions is easy for specialists, but deeper nodules are less specific and imaging methods help reaching diagnosis. It is extremely important that the radiologist know these pathologies and interact with the specialist.

Personal information

References