Differential diagnosis of pediatric lumps in the head

Poster No.: C-3367
Congress: ECR 2019
Type: Educational Exhibit
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Keywords: Tissue characterisation, Neoplasia, Diagnostic procedure, Ultrasound, MR, CT, Paediatric, Musculoskeletal soft tissue, Head and neck
DOI: 10.26044/ecr2019/C-3367

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Learning objectives

• To review the imaging techniques for the evaluation of head lumps in children.

• To describe image findings of lesions affecting the skull and the scalp in the pediatric population, in order to make a correct differential diagnosis.

• To present the most relevant cases of pediatric head lumps.
Background

The palpation of a lump in the head of a child is a common reason to seek medical attention. Those lumps include a wide spectrum of conditions, both benign and malignant. The clinical appearance is usually unspecific, so, imaging plays a crucial role in narrowing diagnosis and guiding potential future interventions.

Radiologists attending a child with a head lump must know the appearance of the most frequent benign lesions affecting the skull in order to avoid further unnecessary diagnostic procedures. It is also very important that radiologists recognize those features that are suggestive of malignancy or drive potential morbidity (intracranial extension, ocular disturbance, etc).
Findings and procedure details

In the presence of a lump in the head of a child, first of all, we should try to narrow the differential diagnosis with relevant clinical data, such as whether the lesion was present at birth or not, if there is a history of trauma, if it is stable or growing. Unfortunately, in the majority of cases, the clinical history is confusing and unspecific.

Imaging techniques:

Radiographic images are usually unspecific and only lesions with bone involvement can be demonstrated.

B-mode and color Doppler ultrasound (US) are the first-line imaging modalities of choice, due to its wide availability and lack of radiation. As these lesions are located superficially, they can be easily evaluated with US, using a linear probe. US provides rapid information about size, contents (solid or cystic), vascularity, location: skin, subcutaneous tissue or bone (Fig. 1 on page 11) and relationship with adjacent structures [1].

In some cases, the US and radiographic appearance can be diagnostic. If the mass is large, deep or has features of malignancy, further evaluation is required.

In the case of lytic lesions, CT may be useful to evaluate bone destruction and tumor spread. However, it has two main disadvantages, which are the use of radiation and that it offers a poor detail in the evaluation of soft-tissues.

On the other hand, MR imaging offers excellent soft-tissue contrast, anatomic and vascularization details and it's also useful in evaluating intracranial connection of small lesions [2]. Unfortunately, the lesser availability of MRI and the necessity of sedation might limit the use of this technique.

Differential diagnosis of pediatric head lumps:

To review and analyze the wide group of entities that can be presented as a head lump, we will divide them into congenital, benign and malignant lesions.
1. CONGENITAL LESIONS

- **Dermoid and epidermoid cysts**

Both lesions may overlap in imaging. Radiological features may vary depending on the content.

Dermoids are unilocular cysts that typically contain thick greasy sebaceous material, keratin debris and skin adnexa such as hair follicles.

Epidermoid cysts are inclusion cyst with keratin and cholesterol without skin elements. Sinus tracts reaching the skin surface can be seen in the nasal area.

Dermoids and epidermoids cannot be distinguished from each other with US [1]. In both cases, US typically demonstrates a well-defined hypoechoic and avascular mass with posterior acoustic enhancement shadow and bone scalloping. They may contain echogenic foci from the presence of calcification, fat or proteinaceous material [4,5].

On radiography, both dermoid and epidermoid cysts might appear as lucent lesions with well-defined sclerotic margins Fig. 2 on page 11.

MRI is not indicated unless there is a suspicion of intracranial extension or when the diagnosis is uncertain. On MRI, as a general rule, a cystic mass with lipids that has no contact with intracranial structures, suggests a dermoid cyst [5]. Epidermoids typically, but not specifically, has a restricted signal on diffusion-weighted images Fig. 3 on page 12. If the intracranial connection is seen, the diagnosis of an encephalocele should be considered.

- **Encephalocele**

It is a herniation of intracranial tissue through a defect in the cranium and it is often associated with other intracranial malformations Fig. 4, Fig. 5 on page 14. In the presence of a midline nasofrontal mass, MRI is useful in differentiating a dermoid cyst from a frontoethmoidal encephalocele or nasal glioma, which is a congenital benign lesion composed of dysplastic heterotopic glial tissue, with signal intensity similar to the brain, not communicated with the subarachnoid spaces.

- **Vascular**
Vascular anomalies of the head and neck region constitute approximately 60% of vascular anomalies diagnosed in children. Vascular lesions are divided into two categories, vascular tumors, and vascular malformations. The physician should exclude features of a vascular lesion with associated syndromes [6].

- **Infantile hemangiomas** are the most frequent vascular tumors, and they usually appear during the first year of life and resolve spontaneously. **Congenital hemangiomas**, in contrast, are rare and they are fully formed at birth. Classic history and presentation can help to differentiate them from other vascular lesions. During the proliferative phase, US shows a solid mass with heterogenic echogenicity and numerous venous and arterial vessels inside [7,8,9]. On MRI, the lesion appears as well-circumscribed lobulated lesion hypo- to isointense relative to muscle on T1-weighted images, and hyperintense on T2-weighted images, with an early and intense enhancement after contrast material administration Fig. 6 on page 15 Fig. 7 on page 16

- **Vascular malformations** are present at birth and grow with the child. They may undergo phases of accelerated growth during puberty. Spontaneous involution does not occur. They can be divided into low-flow (lymphatic, venous, capillary or mixed) or high-flow malformations (arteriovenous malformations and fistulas).

  - On ultrasound, lymphatic malformations appear as multiseptate cystic masses whether venous or mixed malformations are ill-defined heterogeneous lesions with anechoic areas which may present hyperechoic foci from bleeding or thrombosis or calcifications (phleboliths).
  - Venous or mixed malformations are usually visualized on MRI as a lobulated multilocular mass (venous lakes separated by septa). It shows intermediate signal intensity in T1 images and hyperintense in T2, with signal voids secondary to phleboliths and heterogeneous enhancement due to the presence of thrombi. [6,7,8,9] Fig. 8 on page 17
  - Lymphatic malformations will show a non-enhancing, thin-walled, uni- or multi-loculated cystic lesion with septal enhancement [6] Fig. 9 on page 18.
  - Arteriovenous malformations (AVM) are rare lesions in which arterial and venous systems are connected directly by a nidus. On US, AVM appears, in general terms, as a conglomerate of tortuous vessels without a well-defined mass. Doppler US can show an arteriovenous shunt, with high velocities and low resistance flow in the arteries. MRI typically demonstrates an ill-defined soft tissue mass with flow voids and early arterial enhancement. MRI is also useful to assess intracranial involvement [6] Fig. 10 on page 19.

- Another congenital lesion that rarely appears as a palpable lump is the **arachnoid cyst**. It is a benign and asymptomatic lesion, usually located within the subarachnoid space
which contains CSF. The majority of arachnoid cysts are supratentorial. On imaging studies, they appear as well-circumscribed cysts. They typically displace structures nearby and can remodel the bone Fig. 11 on page 20.

2. BENIGN LESIONS

- **Lymph nodes**

Because children have hypertrophic lymphatic tissue and little fat in the craniocervical area, it is very common to find palpable lymph nodes in this age group and location. Therefore, lymph nodes constitute a common reason for parents concern and ultrasound evaluation.

The evidence of a nodular soft tissue mass with central echogenic vascular hilum is normally enough for characterization (Fig. 12 on page 33). Only when they are bigger than 1cm or heterogeneous further studies should be carried out.

- **Langerhans cell histiocytosis (LCH)**

LCH is a proliferation of pathologic Langerhans cells. It can be classified into three groups, depending on how many sites and organs are affected. Localized LCH is the most common form, and most-frequently involves the bone in children. The calvaria is the most common location of osseous LCH, followed by the orbit, maxilla, mandible and temporal bone. It is more common in children between 1-4 years of age. On radiography, bone LCH appears typically as lytic lesions without periosteal reaction or reactive changes ("punched out" lesions). On US, it appears as a solid mass extending from the diploic surface, with poor vascularity. CT and MRI can demonstrate a soft-tissue mass with bone destruction. On MRI this lesion shows intermediate signal intensity on T1-weighted images and high signal on T2-weighted images. Diffuse and mild enhancement can be shown. MRI can better demonstrate intracranial extension [11] Fig. 13 on page 21, Fig. 14 on page 22.

- **Other lesions**

  - **Pilomatrixoma**

The pilomatrixoma or pilomatrixoma, which arises from the hair matrix, is a common benign lesion in children. Up to 68% of pilomatrixomas are found on head and neck. They are small subcutaneous lesions that frequently calcify. On US, a pilomatrixoma appears as a small and well-circumscribed soft-tissue mass that may contain echoic foci from the presence of calcification (Fig. 15 on page 23). On MRI it is iso or hypointense in all MRI sequences, with a patchy or peripheral enhancement. Peritumoral edema
or inflammation can be seen at MRI imaging in pilomatricomas, emulating other more aggressive pathologies [12] (Fig. 16 on page 24).

- **Subcutaneous granuloma annulare**

It is an uncommon and idiopathic benign inflammatory condition most commonly seen in kids aged 2-5 years. It can affect the scalp, usually the occiput.

US shows a solid, poorly-defined, hypoechoic, fascial based mass. On MRI, an ill-defined lesion can be seen, iso-intense to muscle on T1-weighted images and heterogeneous hyperintense on T2-weighted images with fascial tail and variable enhancement [9] (Fig. 17 on page 25).

- **Myofibroma**

Myofibromatosis (multicentric) or myofibromas (solitary) are the most common fibrous tumors in infancy, mainly in children under 2 years. Myofibromas are benign tumors characterized in imaging by presenting a low signal at T1 and variable at T2-weighted images, with peripheral enhancement after administration of intravenous gadolinium (“target” sign) [12,13] (Fig. 18 on page 26)

- **Angiomatoid Fibrous Histiocytoma**

Angiomatoid Fibrous Histiocytoma is a rare low-grade soft-tissue tumor with origin in dermis a subcutaneous tissue. It is most commonly seen in children and young adults. It has slow growth and sometimes it can simulate a bruise. Although it is not specific, on imaging this tumor can appear as a multicystic mass with internal fluid-fluid levels that indicate intralesional hemorrhage and enhancing solid parts. It also may be calcified. [14] (Fig. 19 on page 27).

3. MALIGNANT LESIONS

**- Sarcomas**

Soft tissue sarcomas account for 4-8% of cancers in patients up to 14 years of age. 35% of all sarcomas in the pediatric population manifest in the head and neck. Rhabdomyosarcoma is the most soft-tissue sarcoma in children [2]. In the head and neck, it is classified as orbital, parameningeal or superficial. Clinically, they can mimic other soft-tissue lesions or even an infection. At imaging, they appear as a poorly defined mass, isointense on T1-weighted images and hyperintense on T2-weighted images, with bone destruction and moderate enhancement (Fig. 20 on page 28).
-Osteosarcoma and Ewing sarcoma

Malignant tumors of the skull are unusual, but osteogenic sarcoma and Ewing sarcoma have been reported (Fig. 21 on page 29).

-Metastatic Lesions

Metastatic lesions of the calvarium in children include leukemia, neuroblastoma, and small round cell tumors. Neuroblastoma commonly metastasizes to the lateral orbital walls, skull base, and calvaria Fig. 22 on page 30, Fig. 23 on page 31.

4. TRAUMATIC LESIONS

A variety of traumatic lesions can occur during the neonatal period due to delivery :

-Caput succedaneum

Diffuse swelling of the scalp that contains fluid and blood. Medical support is not required.

-Subgaleatic hematoma

It consists on hemorrhage located subjacent to the aponeurosis covering the scalp. It also crosses sutures lines. It can appear up to 4 days postpartum. In order to assess potential intracranial injuries, cranial US should be performed.

-Cephalohaematoma

This is a blood collection between the periosteum and the skull. It does not cross sutures. Associated fractures may be found. Cephalohaematomas can mimic epidural hematomas (mirror artifact). Color Doppler ultrasound is helpful by showing normal vessels within the suspicious epidural hematoma [15]. Chronic cephalhaematomas may calcify. (Fig. 25 on page 34), (Fig. 26 on page 35), (Fig. 27 on page 36).

-Ping-pong fracture:

A ping-pong fracture is a depressed skull fracture without loss of bony continuity. It can appear in newborns, after a localized skull compression (maternal pelvis, forceps..)[15]. In the physical examination, the non-depressed part of the skull may be confused with a head bump (Fig. 28 on page 37).
Fig. 1: Scheme of scalp layers.

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Fig. 2: Suprachiliary epidermoid cyst in a 1-year-old infant (arrows). Radiography shows a lucent lesion with well-defined sclerotic margins. US shows a well-defined hypoechoic mass, avascular, with posterior acoustic enhancement and bone skalloping.

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Fig. 3: Epidermoid cyst in a 1-year-old girl. Sagittal STIR MR image (A) and sagittal diffusion-weighted MR image (B). Midline nasofrontal mass

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Fig. 4: Newborn with occipital meningoencephalocele. Cranial contrast-enhanced CT image with VR reconstruction (A), MIP reconstruction (B) and bone window (C). Axial T2-weighted MRI image (D). Images show herniation of meninges, intracranial tissue, and dural venous sinuses through a defect in the cranium (arrow).

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Fig. 5: 11 year-old boy with occipital meningoencephalocele. Axial CT image (A) and VR CT image (B). Imaging demonstrated herniation of intracranial tissue through a defect in the cranium (arrow) as well as other associated brain anomalies (heterotopia and hydrocephalus).

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**Fig. 6:** 1 year-old girl with infantile orbital hemangioma (arrows). B-mode ultrasound (A), Doppler ultrasound (B), multiphasic dynamic contrast-enhanced MRA image (C), coronal fat-suppressed T2-weighted MRI image (D). Ultrasound images show a solid mass with prominent internal vascularity in close proximity to the ocular globe (red arrow). MR was performed to define intraorbital extension and intracranial vascular relationship.

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**Fig. 7:** Congenital hemangioma (PICH). B-mode and Doppler ultrasound images (A,B), a clinical picture at birth (C), a clinical picture at 3 years of age.

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Fig. 8: Multiple venous malformations in a 13-year-old boy with Blue Berry Bled Nevus Syndrome and a painful head lump. CT images with VR reconstruction (A) and bone window (B); sagittal (C) and axial (D) FLAIR brain MRI; STIR whole-body MRI images (E,F,G), a clinical picture at birth (H). Head images show soft tissue occipital venous malformation with prominent bone scalloping (arrows in A and B). Whole body MRI demonstrated many other venous malformations in the gastrointestinal tract and in the soft tissues of the neck and extremities. The clinical picture at birth shows a huge frontal venous malformation.

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**Fig. 9:** Lymphatic malformation in the scalp. US images show a multi-loculated cystic lesion (arrows) in the scalp without evidence of intralesional doppler signal.

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**Fig. 10:** Frontal arterio-venous malformation in a 14-year-old boy. Doppler ultrasound (A,B), coronal T2-weighted MRI image (D), multiphasic dynamic contrast-enhanced MRA image. Ultrasound shows a vascular nidus causing bone remodeling in the frontal area with intracranial extension. MRI demonstrates feeder branches of the external carotid artery and venous drainage to superficial cervical and intracranial cortical veins.

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**Fig. 11:** Arachnoid cyst in a 5-year old girl. AP radiograph (A), Doppler US image (B), axial T1-weighted MRI image (C), coronal T2-weighted MRI image. Arachnoid cyst (arrows) with bone scalloping.

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Fig. 13: LCH in the left temporal bone (arrows). Doppler and B-mode US images (A,B), contrast-enhanced cranial CT, soft-tissue (C) and bone window (D), coronal FLAIR and axial contrast-enhanced T1-weighted MRI images (E,F).

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**Fig. 14:** LCH in the right frontal bone. AP radiograph (A), Doppler US image (B), contrast-enhanced cranial CT, bone window (C) and soft-tissue (D), axial T1-weighted and coronal contrast-enhanced T1-weighted MRI images. Images reveal an aggressive soft tissue mass eroding the occipital bone with avid contrast enhancement.

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**Fig. 15:** US images show supraciliar pilomaticoma in a 5-year-old girl (arrow). Well defined soft tissue mass superficial to the scalp aponeurosis with tiny scattered echogenic foci.

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Fig. 16: Ruptured pilomatrixoma that shows adjacent inflammatory changes (arrows). B-mode and Doppler US images (A,B), sagittal T1-weighted and STIR MRI images (C,D), sagittal contrast-enhanced T1-weighted MRI image.

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Fig. 17: Frontal subcutaneous granuloma annulare (arrows) in a 2-years-old boy. Lateral skull radiograph (A) shows a soft tissue mass protruding in the frontal region. B-mode US image (B) demonstrates an hypoechogenic mass limited by the periosteum with fascial extension. Axial and sagittal contrast-enhanced T1-weighted MRI images (C,D) reveals a subfascial mass with intense contrast enhancement.

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**Fig. 18:** Cranial myofibroma in a 3-year-old boy (arrows). B-mode US image (A), contrast-enhanced cranial CT, soft-tissue window (B), coronal STIR and sagittal contrast-enhanced T1-weighted MRI images (C,D). Peripheral contrast enhancement is characteristic.

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**Fig. 19:** Angiomatoid Fibrous Histiocytoma (arrow). Doppler US show an heterogeneous soft-tissue mass with both solid and cystic components with intralesional hemorrhage.

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**Fig. 20:** Orbital rhabdomyosarcoma in an 11-year-old girl (arrows). Axial contrast-enhanced cranial CT, soft-tissue window (A), axial contrast-enhanced T1-weighted MRI image (B).

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Fig. 21: Ewing sarcoma of the skull. Contrast-enhanced CT images with VR reconstruction (A) and soft-tissue window (B); axial FLAIR (C) and sagittal contrast-enhanced T1 (D) MRI images. There is a big soft tissue mass in the occipital area with aggressive sunburst periosteal reaction. Images also demonstrate compression of the adjacent brain parenchyma.

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Fig. 22: Metastatic lesions of the calvarium in a 15-year old boy with Ewing’s sarcoma of the ulna. Elbow and cranial radiographs (A,B), contrast-enhanced CT images, bone and soft-tissue windows (C,D).

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Fig. 23: Metastatic lesions of the right orbit (arrows in B and C) in a 15-month infant with metastatic abdominal neuroblastoma (arrow in A). FLAIR abdominal MRI image, coronal T2-weighted cranial MRI image, axial contrast-enhanced T1-weighted MRI image (C).

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**Fig. 24:** Scheme of possible skull bleeding locations.

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**Fig. 12:** 2 year-old-boy with a palpable occipital lymph node. The vascular echogenic hilum with Doppler signal in the center of the nodular lesion is characteristic and is usually enough to define the diagnosis.

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**Fig. 25:** Cephalohaematoma in a newborn in a B-mode US image (arrow in A). Calcification of the cephalohematoma one month later (arrows in B and C). B-mode US image (B) and AP radiograph (C).

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Fig. 26: Cephalohaematoma in a newborn (arrows). B-mode and Doppler US image (A,B) show a subperiostial collection. Note the mirror artifact simulating an intracranial bleeding. The diagnostic clue is the presence of crossing vessels. Sagital and coronal contrast-enhanced CT (C,D).

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Fig. 27: Occipital cephalohematoma in a newborn. Sagittal T1-weighted MRI image (A), coronal and axial T1-weighted MRI images (B,C).

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Fig. 28: A ping-pong fracture in a newborn. AP and lateral radiographs show a depressed skull fracture without loss of bony continuity (arrow).

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Conclusion

There is a wide spectrum of conditions, both benign and malignant, that can present as a head lump in the pediatric population. Radiologists attending a child with a head lump must know the appearance of the most frequent benign lesions affecting the skull in order to avoid further unnecessary diagnostic procedures. Ultrasound (US) is the first-line modality of choice. In some cases, further evaluation with CT or MRI should be done.
References


