Skull Vault Tumors and Tumor-like Lesions: A review

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Learning objectives

The aims of this pictorial review are to:

- Describe the imaging features of the main tumors of the cranial vault
- Clarify the place of each imaging method in the diagnostic approach
Background

Skull vault tumors and tumor-like lesions are a rare entity that represents a diagnostic challenge for the radiologist. They are commonly discovered incidentally in imaging or during the exploration of a painful or not swelling. The recognition of their pathologic characteristics requires a good knowledge of the normal anatomy and its variants. The main role of imaging is the detection and the characterization of these lesions, with evaluation of the extent of invasion of adjacent critical organs. Computed tomography (CT) and magnetic resonance imaging (MRI) are the preferred modalities for the evaluation of skull lesions. In deed, CT is a reference examination, which was essential for a good bone study, useful for detecting of mineralized component. However, actually, it is increasingly being replaced in first intention by MRI, because its high resolution which has made it possible to characterize tissue mature , to specify the intracranial extension, to assess the infiltration of the diploe and to show vascularity. So, the appearances of some lesions is often complicate the process of radiological diagnosis and despite a rigorous diagnostic procedure, a certain number of lesions are not characterized before the biopsy.
Findings and procedure details

A wide spectrum of congenital and acquired lesions is commonly encountered in skull. A variety of these tumors have typical imaging appearances. Several classifications have been proposed. In this review skull vault tumors may be classified into focal, multifocal and diffuse lesions.

# Solitary lesions: they can be lytic or sclerotic lesions

# Osteolytic solitary lesions: osseous hemangioma, ossifying fibroma, hydatic cyst, dermoid and epidermoid cysts....... 

1. **Osseous hemangioma:** it is benign slow-growing vascular bone tumor accounts for less than 1% of primary bone lesions [1]. The skull is a second most frequent location after spine. It is principally located in the frontal and parietal bones. **CT:** well-circumscribed intradiploic lesion with possible peripheral sclerosis (figure1). It erodes the outer table and spares the inner table. It enhances homogeneously after contrast injection. **MRI:** it is isointense on T1 and hyper intense on T2-weighted images; it may contain hypo- or hyper intense spots related to the presence of fat or iron. After gadolinium administration, the enhancement is initially focal and then diffuses.

2. **Ossifying fibroma:** it is a rare, benign primary bone tumor that occurs most commonly in the jaw but it is extremely rare in calvarial bones [2]. On CT, it usually appears as a dense bony expansion, located mainly in dipole. The lesion has lower density than the surrounding structures and older areas tend to be calcified. On MRI: it appeared as a low intensity mass with a high intensity margin (figure2).

3. **Solitary plasmocytoma:** it is a hematological malignancy with the same histology as myeloma, but with a unique character. In imaging, it may mimic that of a meningioma [3]. The diagnosis is most often histological, but the importance of osteolysis without any osteosclerosis and associated with a bulky mass must make evoke this diagnosis (figure 3).

4. **Hydatid cyst:** cranial hydatidosis is predominantly seen in young adult males. In CT scan and MRI, it appears as a solitary, homogeneous and spherical cyst with well-defined borders and without perifocal edema. The density and the signal of the cyst fluid is the same as that of cerebro-spinal fluid. The lesion may later break through the tables and produce a soft-tissue mass.

5. **Dermoid and epidermoid cysts:** this lesion is secondary to congenital or post traumatic epidermal or dermal inclusions within the diploe. Parietal and frontal bones are frequently involved [4]. **CT:** well-demarcated osteolytic lesions, homogeneously hypodense with sclerotic borders, which tend to expand into both the inner and outer tables. **MR:** the signal intensity depends on the
content of the cyst. Epidermoid cyst has pure fluid signal intensity. Typically, it has bright signal intensity on isotropic diffusion-weighted MR images. A more complex signal intensity (hyperintense on T1-weighted images, hypointense on T2-weighted images) in a dermoid cyst. There is no enhancement after gadolinium administration.

6. **Aggressive tumors of the scalp:** tumors which are located in scalp and showed intracranial extension have been reported to be very rare. Malignancies which have been described to result in extensive intracranial extension include sarcoma, lymphoma, carcinoma (figure 4), and lymphoma. The extent of local recurrence and rapid intra cerebral spread emphasize the potentially aggressive nature of the scalp and the need for early diagnosis and complete excision [5].

7. **Primary malignant tumors:** primary malignant tumors of the skull are rare. Some cases have been reported in the literature. They have been reported to account for around 0.8 to 1% of all bone tumors [6]. Most tumors occur in the context of Paget disease, fibrous dysplasia, or prior therapeutic irradiation. Malignancy resulting from these predisposing factors is also more likely to appear at a more advanced age. Imaging with MRI and CT may reveal bone growth with lytic regions and periosteal remodeling, as seen in bone tumors of other areas.

8. **Sclerotic solitary lesions:**

1. **Osteoma:** it is made up of compact and trabecular bone in variable proportions. It develops from the outer table, mainly of the frontal or parietal bone, and is either sessile or pedunculated. In rare cases, it develops from the inner table or in the diploë. The presence of multiple osteoma of the skull should prompt the search for Gardner syndrome. **CT:** small, well-defined round or oval lesion that is usually dense and homogeneous. **MRI:** homogeneous low signal intensity on T1-weighted images with variable appearance in T2-weighted images, that depends on the relative content of cortical and trabecular bone [4].

2. **Intraosseous Meningioma:** it accounts for less than 2% of all meningiomas. It sites near the sutures, particularly the coronal suture. **CT:** osteosclerotic lesion which is frequently associated with destructive irregular borders suggestive of the diagnosis. **MRI:** low signal intensity on T1-weighted images and variable appearance on T2-weighted images and does not enhance after gadolinium administration but a meningeal enhancement is possible by adjacent irritation [7].

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**Multifocal lesions:**
1. **Metastases**: they can be encountered at any age, with a higher prevalence during the sixth and seventh decades of life. They are usually secondary to osteophilic tumors in adults and to neuroblastoma or sarcomas in children. Metastases of thyroid or renal neoplasia are single and expanded. However, diffuse infiltration of the skull is encountered less frequently [4]. In imaging, well-circumscribed osteolytic or osteosclerotic lesion, which generally extend into the adjacent soft tissue. The signal intensity in MRI is not specific. Mostly, areas of lesion replacing the normal diploidic space and cortical bone. Enhancement after gadolinium can be homogeneous, heterogeneous, peripheral, ring enhancement or lack of enhancement (figure 5).

2. **Langerhans histiocytosis**: parietal bone is frequently affected. It's a rare disease and most common in children, particularly those between 6 and 10 years old [4]. **CT**: oval or round isolated lytic lesions that begin in the diploe. The margins appear circumscribed, and the edges may be beveled. The commune characteristic is the Bony sequestrum (which represent the residual intact bone). Multiple lesions may expand and coalesce, producing the appearance of a geographic map and periosteal reactions are rarely encountered. **MRI**: the signal intensity of the lesions is not specific, but usually they strongly enhance after gadolinium administration. Extra Dural and extra cranial extension are frequent. Peri lesional edema is rarely encountered (figure 6).

3. **Myeloma**: it is a malignant disease of the bone marrow characterized by a monoclonal proliferation of plasma cells. It is the most common primary malignant bone lesion, with a higher prevalence in men between the fifth and the eighth decades. **CT**: because lesions in myeloma are exclusively osteolytic, they are usually not detected with 99m Tc-based scintigraphy . **MRI**: the signal intensity of the lesions is nonspecific; a "salt-and-pepper" appearance or diffuse bone marrow replacement may be noted.

# Diffuse or extensive lesions:

1. **Paget disease**: patients are typically older than 55 years. It affects the skull in 28-42% of cases. Frontal and occipital bones are frequently affected [4]. **CT**: osteolytic activity is predominant in the beginning of the disease, and consists on circumscribed osteoporosis of the skull. The characteristic appearance of Paget disease includes homogeneous enlargement of the skull vault and thickening of the tables which leads to a loss of differentiation between these structures (figure 7). **MRI**: it depends on the stage of the disease. The osteoblastic activity of the pagetic bone develops fatty bone marrow containing hypo intense thickened trabecular and surrounded by thick hypo intense tables.

2. **Fibrous dysplasia**: it is frequently seen in childhood and adolescence. Frontal and temporal bones are frequently affected; sutures may be crossed [8]. **CT**: is the imaging modality of choice for the diagnosis because it can show the characteristic ground-glass matrix appearance (70-130 HU). This lesion is responsible of expansion of the diploe, bulging of the outer table.
and thinning of the inner table. MRI: lesions are mainly hypo intense on T1 and T2 -weighted. Sometimes, they are isointense on T1 weighted and heterogeneous on T2, depends on the fibrous tissue density, intrallesional cellularity, and hemorrhagic or cystic rearrangements. Enhancement after gadolinium is also variable (figure 9).

**Differential diagnosis:**

The radiologist has a long list of differential diagnosis which includes congenital, traumatic, inflammatory and neoplastic lesions:

# Anatomic variants: trancalvarial venous channels and venous lake, arachnoid granulation, enlarged parietal foramina….

# Congenital: sinus pericranii, encephalocele, leptomeningeal cyst…

# Inflammatory: osteomyelitis…
Fig. 1: CT scan: An intradiploic osteolytic lesion of left frontal bone with intralesional spicules. It erodes the outer table and spares the inner table of skull. It is expanded to a soft scalp. The etiological research of a primitive tumor was negative; the diagnosis of a hemangioma was retained.

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**Fig. 2:** CT shows an expansile intradiploic lesion at the right frontal bone with enhancing soft tissue component. There are areas of ground glass density and ossifications seen within. On MRI, this lesion has a low signal on T1 with homogenous enhancement post-gadolinium. The diagnosis of an ossifying fibroma is made post-operatively.

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**Fig. 3:** CT: Osteolytic lesion of the occipital bone with significant contrast enhancement. Extension of the lesion in soft scalp tissues. Biopsy discovers a solitary plasmocytoma.

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**Fig. 4:** Coronal T2 W image shows an extra cranial cyst tumor with significant osteolysis in sagittal T1 W and which demonstrated peripheral contrast enhancement after intravenous gadolinium administration on the T1W coronal image. A biopsy taken from the lesion revealed invasive epidermoid Carcinoma of the Scalp.

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**Fig. 5:** Tissue mass lysing the cortex of the frontal bone with meningeal invasion in the CT. MRI shows that this mass is hypervascular with endocranial extension. Histological examination confirmed the metastasis of a papillary carcinoma of the thyroid.

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**Fig. 6:** The CT scan of a 12-year-old girl shows an isolated lytic lesion in the diploe. In MRI, this lesion is expansive with T1 iso-signal and T2 high signal, with enhancement after gadolinium administration. Further exploration confirmed a histiocytosis.

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**Fig. 7:** Thickening of the cranial vault with loss of differentiation between the internal and external tables. Typical appearance in CT and MRI of Paget's disease.

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**Fig. 8:** On CT: lesion of vault with ground-glass appearance. On MRI: this lesion is mainly hypointense on T1 and T2-weighted with discrete enhancement after gadolinium.

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Conclusion

MRI is the reference imaging technique for exploring tumors of the vault. The CT is in second intention that brings complementary semiological elements, allowing refining the diagnostic procedure. Biopsy is often essential.
References