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Imaging evaluation of diffuse abnormalities of the cranial vault (Case report)

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ABSTRACT
A variety of diffuse diseases affect the calvaria. They may be identified clinically as palpable masses or incidentally in radiologic examinations. The purpose of this study is to illustrate the main diffuse calvarial lesions starting from a case report of fibrous dysplasia of the cranial vault. A 68-year-old male patient who presented with a history of right otalgia and bloody otorrhea was diagnosed to have fibrous dysplasia based on the radiological features. Most diffuse diseases of the calvaria are benign non-neoplastic lesions of unknown origin. The radiologist has a long list of differential diagnosis and their true etiology may be puzzling when the medical evaluation is based only on imaging findings.

Keywords: calvaria, fibrous dysplasia, Paget

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Introduction

Calvaria is affected by a variety of diffuse diseases, which can be clinically identified as palpable masses or accidentally, in imaging examinations.

Case report

Patient aged 68 years, known with cranial deformity since childhood and resection of the external auditory canal polyp as a week ago, was hospitalized for right ear otalgia and bloody otorrhea. Schuller radiographic view and anterior sinuses of face radiography reveals areas of hyper transparency relatively well defined, with wiped out contour, tendency to confluence, alternating with osteocondensation areas, situated in left half of the crania; mastoid cells lack of pneumatization (Figures 1-2).

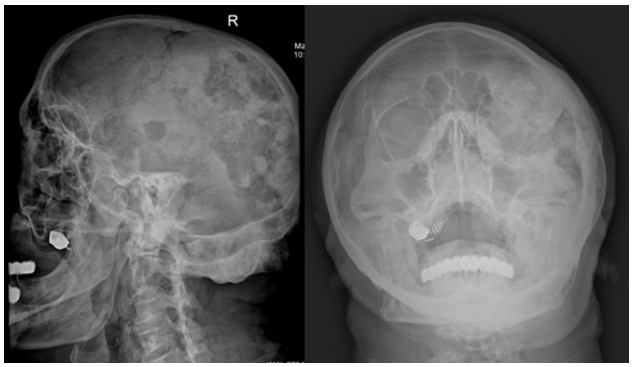

Figure 1-2

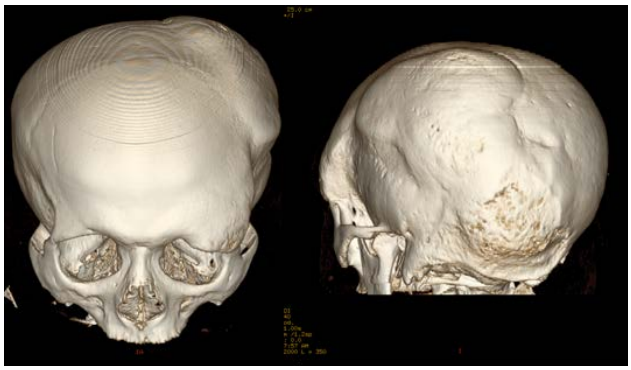

Figure 8-9

Subsequently, the patient performs a computed tomography examination showing extensive lytic areas with heterogeneous structure by amorphous nodular and linear ossification included, developed at the level of bones from the right half of the cranium, associating important diploic ballooning, without interrupting the internal and external tables continuity and cortico-spongy dedifferentiation; identical morpho-densitometry lesions developed at the level of left zygomatic bone, left sphenoidal grater wing and body, clivus, squama and basilar region of the occipital bone, petrous part of the left temporal bone and left mastoid process. Diagnosis tilts in favor of fibrous dysplasia, rather than Paget disease, given the lesion morphology and the corroboration with family medical history (Figures 3-7, 8, 9).

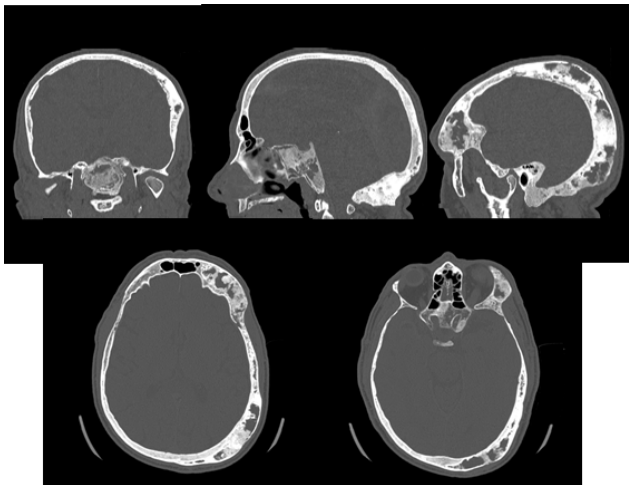

Figure 3-7

Discussions

Anatomy

The calvaria or cranial vault is the upper part of the neurocranium and is made up of the superior portions of the frontal bone, occipital bone, and parietal bones, linked together by sutures (coronal, sagittal and lambdoid). Most bones of the calvaria consist of internal and external tables, separated by diploe (Figure 10).

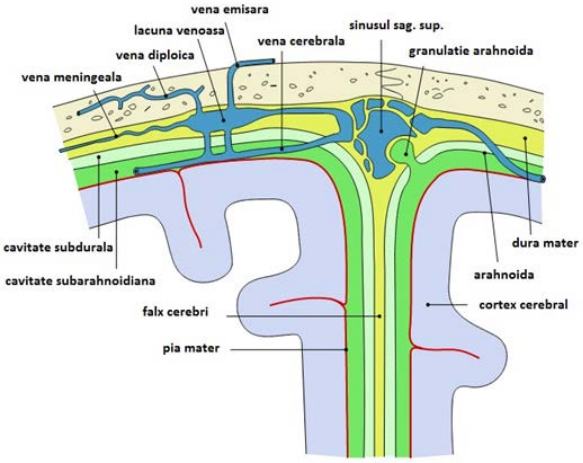

Figure 10. Skullcap (sectional) anatomy

The internal table is in contact with the meninges (dura, arachnoid, arachnoid trabeculae and Pacchioni granulations - Figure 11). The whole set is covered or in intimate contact with the vascular elements: venous sinuses, venous lakes, diploic veins (Figure 12), epidural veins, meninges veins, brain and emissary veins (Figure 13).

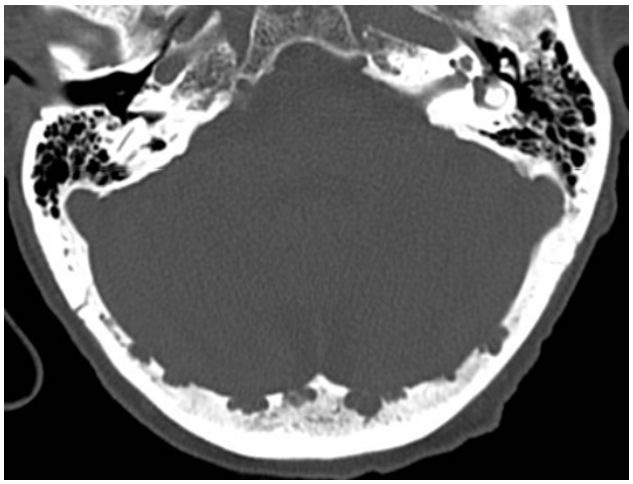

Figure 11. Skull CT, bone window. Typical Pacchioni granulation aspect placed at lateral sinuses level.

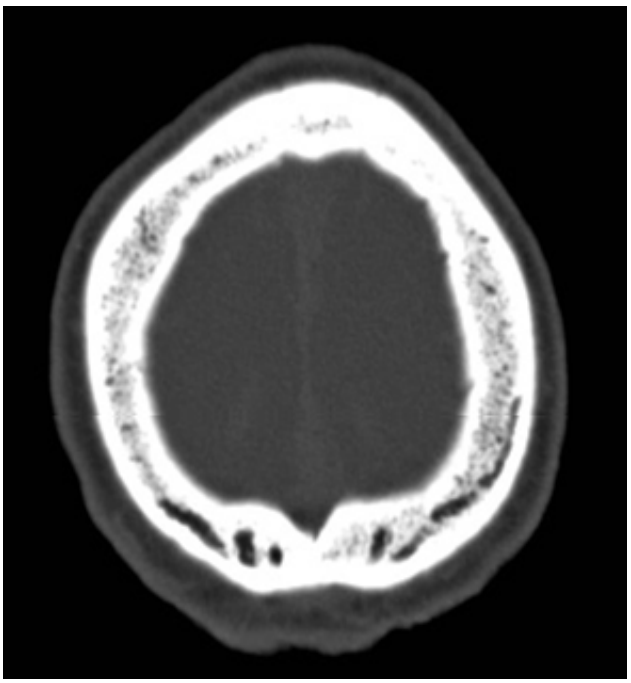

Figure 12. Skull CT, bone window. Linear and serpiginouses vascular images (diploic veins)

Imaging methods for evaluating the cranial vault

CT scan shows the best spatial resolution in lesion characterization, determining calcifications and bone tumor matrix. It can also evaluate periosteal reaction and extension at the base of the skull and orbit (holes and cracks). It represents the best method for guided biopsy [1,2].

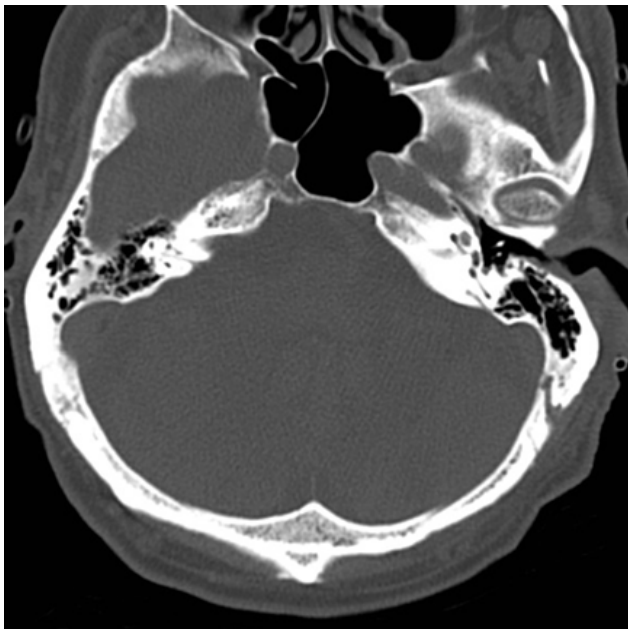

Figure 13. Skull CT, bone window. Emissary vein (arrow).

Magnetic resonance imaging (MRI) is the best resolution in contrast. It represents the method of choice for lesional tissue component characterization, delimitation, vasculature and kinetics loading analysis. Regarding the pre- and post-contrast analysis of diploe it should be noted that only normal venous elements and Pacchioni granulations loads physiological. By using MRI, loco regional endo- and exo-cranial balance (meninges, vascular elements, soft parts) can be performed [3].

Standard radiography of skull remains in third place regarding cranial vault lesions diagnosis. There is no routine, unless multiple myeloma balance and it can only diagnose lesions showing typical character (sickle cell anemia, epidermoid cyst) [4].

The diffuse anomalies of the cranial vault

They are: frontal internal hyperostosis, diploë thickening, fibrous dysplasia, and Paget's disease.

Frontal internal hyperostosis

It most commonly affects women near menopause and it has an incidence of up to 5%. It represent the thickening of the inner table of the calvaria in the form of endofrontal bone appositions arranged relative symmetrical and radial. The extension is limited to posterior by the ascending branch of the middle meningeal artery, being able as well to reach parietal bone. It shows a good table-diploe differentiation [5]. The Morgagni-Stewart-Morel syndrome was described representing the frontal internal hyperostosis association with obesity and hormonal disturbances (Figures 14-16).

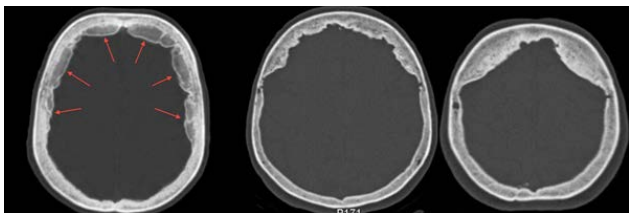

Figure 14-16. Skull CT, bone window. Internal frontal table bone appositions with symmetrical and radial disposal and mamelon-like borders.

Diploic thickening

There may be isolated or integrated into certain clinical presentations: severe anemia (thalassemia, sickle cell anemia), chronic renal failure, hypervitaminosis A, hypo- and hypervitaminosis D (Figures 17-19) [6].

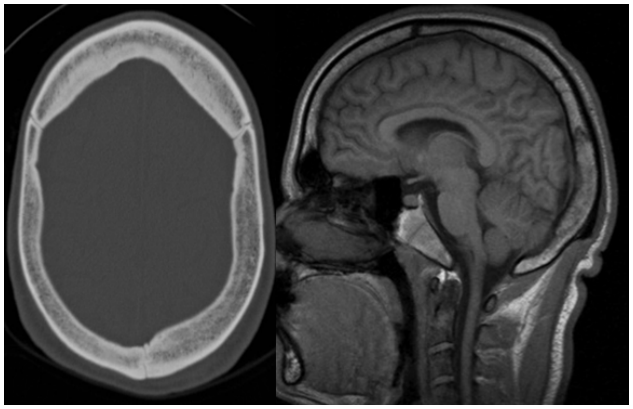

Figure 17, 18. Skull CT, bone window (left). Cranian MRI, T1. Isolated diploe and internal table thickening without known metabolic abnormalities. Hypersignal T1 (fatty).

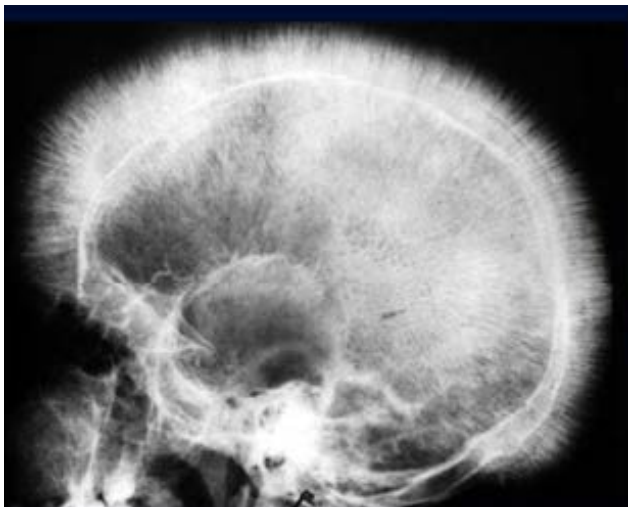

Figure 19. Brush like diploe thickening (evolved thalassemia)

Fibrous dysplasia

It shows replacement of normal bone by fibrous tissue, trabeculae disorganization with possible inflammatory and hemorrhagic cystic reshuffle. It is most commonly found in young adults, sometimes by chance. In terms of evolution, it shows an active phase until puberty and a passive phase into adulthood [7,8]. Shows frequently hemicranian touch, possibly focused, rare malignant degeneration (0.4 to 4% of cases) and may be associated with aneurysmal cyst. [9] Depending on fibrous tissue mineralization can be of three types:

a) condensed, compact (dense, homogeneous bone lesions, ground glass opacities); (Figures 20, 21)

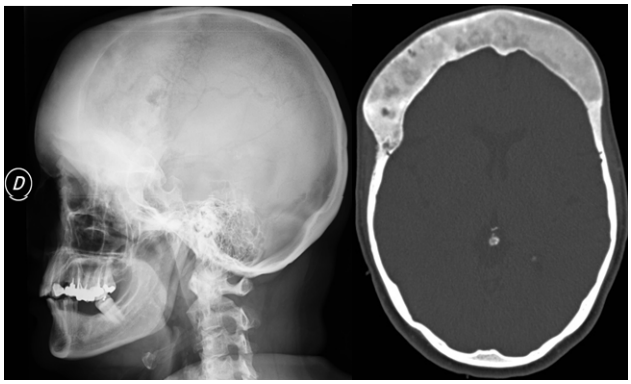

Figure 20, 21. Skull x-ray profile view (left). Skull CT bone window (right). Frontal bone condensed type fibrous dysplasia

b) pseudochistic (few bays, lacunae with tissue density); (Figures 22-24)

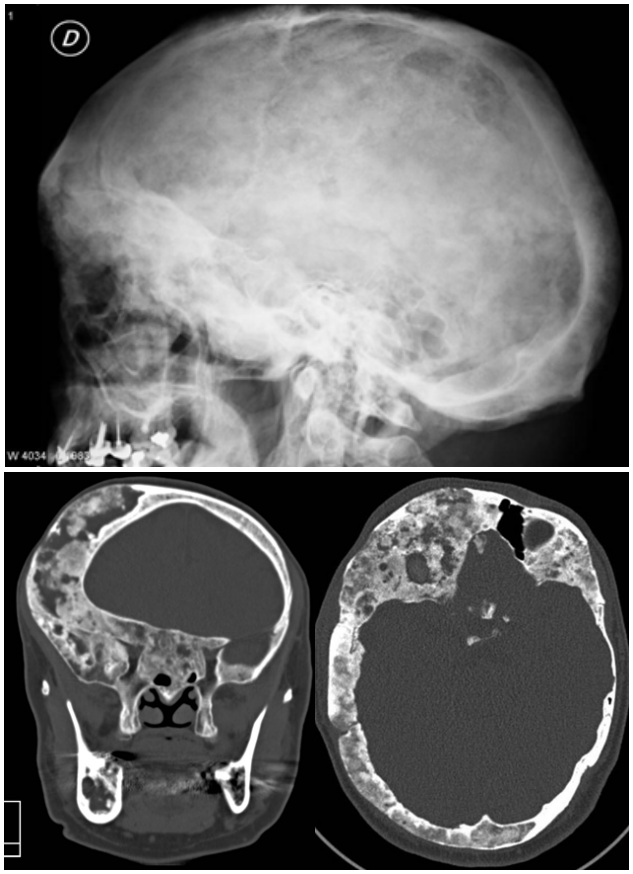

Figure 22-24. Skull CT bone window. Pseudocystic type fibrous dysplasia with skull base extension.

c) Mixed (predominates condensation, cortical thinning, pagetoid appearance) [10,11].

Morphological, shows thickening of newly formed bone (skull dysmorphia), cortico-cancellous dedifferentiation (tables-diploe) with absence of clear demarcation between dysplastic lesions and normal bone and suture failure. In order of frequency, appears impairment of frontal, sphenoid, ethmoid, maxillary bone (and more rare of: temporal, parietal, malar, mandible, occipital bone).

CT examination shows expansive diploic lesion that appears thickened, with external table blown and internal table usually thinned. It is necessary to evaluate the holes at the base of the skull, which can be narrowed (stenosis rarely occurs complete). The MRI aspect is of a hy-posignal T1 and T2 lesion, with post contrast loading. It is also possible to evaluate nerve compressions [12].

Paget's disease

Represents unknown cause of deforming osteitis with excessive and anarchic reshaping of the skeletal parts. It affects 10% of the population over 85 years old, predominantly males. It is generally asymptomatic and may involve the skull in 25-65% of cases [13].

The classic appearance is circumscribed Schuller osteoporosis, with hypertrophic type bone reshaping progression and cortico-medullary dedifferentiation. It affects the whole skull with a fluffy aspect and diffuse irregular tables thickening (Figure 25).

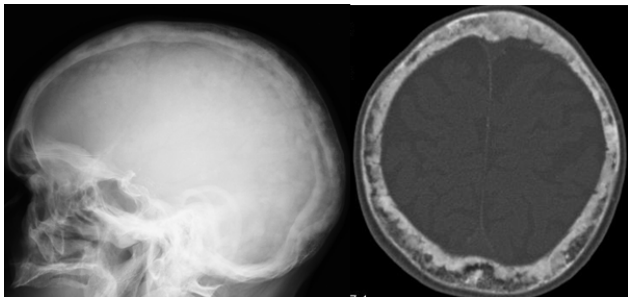

Figure 25, 26. Skull x-ray profile view (left). Skull CT bone window (right). Diffuse calvaria Paget's disease

CT examination reveals the thickening of skull tables, tables-diploe dedifferentiation and "wax stains" appearance of external table (Figure 26). MRI examination with T2 ponderation and fatty suppression is useful in locating active zones, which appear in hypersignal.

In case of pain or swelling sarcomatous degeneration is suspected [14].

Conclusion

Calvaria diffuse diseases are benign, non-neoplastic lesions with unknown origin, frequently asymptomatic. Knowing the different ways of

developing and presenting results in making a correct diagnosis, which can be difficult to achieve just imaging, without knowing the data and patient history. Multiple imaging methods can be used to highlight these injuries, but CT scanning is considered the best method for evaluating bone (Table I).

Table I. Comparison of Paget's disease and fibrous dysplasia

Paget's disease	Fibrous dysplasia
Affects elderly people	Discovered either puberty or young adulthood
Whole calvaria touch	Hemicranian touch
Irregular cortical thickening	External table blown, internal table thinned

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