Getting it into our thick skulls - a radiological review of calvarial thickening.

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Learning objectives

Drawing on our experience of calvarial thickening in a large general and head and neck/neuroradiological practice, the aim of this educational exhibit is to:

1) Discuss the varied aetiologies of skull vault thickening as described in the radiological literature

2) Review key imaging findings of each case

3) Explore some rare causes of calvarial thickening
Background

The separate bones which comprise the skull vault are the frontal, parietal, temporal and occipital bones. Formed entirely by intramembranous ossification, they are unfused in childhood and gradually form first fibrous and then osseous connections at the suture lines. Two bony tables or diploe (an inner and an outer) make up the calvarium.\(^1\) Between these is the diploid space, filled with diploid veins and largely 'red' or active (haematopoietic) marrow.\(^2\)

Thickening or bony changes in the inner, outer or both diploid tables can be caused by a variety of conditions. Thickening of the overall volume can also be caused by diploic space constituent changes.
Findings and procedure details

The various aetiologies can be classified roughly into intrinsic and extrinsic causes. Each group can be further split into common and rare aetiologies.

INTRINSIC CAUSES

Common:

1. Hyperostosis Interna

Most commonly seen in the frontal bone (where it is referred to as hyperostosis frontalis interna), the aetiology is unknown, but may be due to hormonal stimulation as is most often found in post-menopausal women. It was originally described in association with virilism and obesity, but these have since been shown to be spurious associations. Histologically, chaotic thickening with an increase in cancellous bone is seen, usually confined to the inner table. The condition is entirely incidental except when the hyperostosis causes brain compression due to decrease in the intracranial volume. A characteristic ‘bony nodular overgrowth, limited to the inner table’ is seen which is very unusual in other causes of skull vault thickening.\(^3\) (Figure 1.)

2. Diffuse metastatic disease

Bony metastatic disease is most commonly seen in lung, breast, renal and prostate cancers. In adults, metastatic disease is the most common bone tumour (70%).\(^4\) Metastases can be focal or diffuse, and in the skull lead to deformity or neurological compromise (especially if involved with a soft tissue component).\(^5\) Pathological fractures in the skull are uncommon due to the intrinsic stability of the calvarium, and local invasion from a primary tumour rare (although reported in squamous cell carcinoma of the skin). Of the four most common primary malignancies, breast and lung produce lytic metastases (with 25% and 15% displaying a mixed pattern, respectively). Prostate cancer is typically sclerotic, and is one of the only metastases to incite a periosteal reaction (along with some small cell cancers such as neuroblastoma and medulloblastoma).\(^6\) In our example (Fig.2), the patient presented with diffuse lytic metastases from breast carcinoma.

3. Marrow proliferation in diploid space (‘hair on end’ sign)
Massive haematopoiesis leading to marrow proliferation and expansion of the diploid space in the skull is seen most often in thalassemia. (Fig. 3) It is also seen (though rarely) in chronic anaemia/haemolysis, cyanotic heart disease and nutritional disorders. Expansion of the marrow space associated with thinning of cortical bone gives the classic 'hair on end' appearance on skull radiographs.

4. Paget’s disease of bone (osteitis deformans)

Paget’s disease (osteitis deformans) is a disorder of abnormal bone proliferation, with lytic (incipient active), mixed (active) and sclerotic (late) phases. In the skull, the lytic phase is referred to as osteoporosis circumscripta, and was often a cause of a 'change in hat size' in past times. Different phases often exist in the same patient, creating a continuum of bony changes ultimately viewed on CT as disorganised, coarse and thickened trabeculation. (Fig. 4) The inner table is often more extensively affected than the outer table. Polyosteotic disease is more common than monoosteotic. As with many of the diseases described herein, neural and brainstem compression due to bony thickening can be detected by MRI.

5. Fibrous Dysplasia

Histologically, 'expansion and replacement of normal cancellous bone with proliferative cellular fibrous stroma' is seen in this non-heritable disorder of unknown cause. It has a predilection for the craniofacial bones, ribs and long bones. When monoosteotic (75% of cases), it can be discovered incidentally. Polyosteotic fibrous dysplasia is more often symptomatic/debilitating and can involve compression to local structures e.g. cranial nerves. Radiologically, a 'ground glass' appearance replacing the usual visible trabecular pattern echoes the histological findings described above. Approximately 3% of the polyosteotic forms occur in association with endocrine abnormalities (usually precocious puberty) and unilateral cafe-au-lait spots. This is known as McClune-Albright Syndrome, and seen in our example (Fig 5).

Rare:

1. Osteopetrosis

Osteopetrosis ('marble bone disease', 'Albers-Schonberg disease') is a disorder of osteoclast activity, leading to a failure of craniotubular bony reabsorption. There are several different types, however all produce varying degrees of unopposed
osteoblastic activity. Fractures due to poorly modelled bone (usually of the long bones), cranial deformity and cranial nerve palsies due to bony compression are all commonly seen, as well as anaemia and hepatosplenomegaly. Radiologically, the bones will be sclerotic and demonstrate multiple healed/healing fractures, possibly with an Erlenmeyer flask type deformity. In the autosomal dominant form, a classic finding of a 'bone-within-a-bone' may be seen. In both types, the inner table of the calvarium is classically more sclerotic than the outer, and if the patient is anaemic due to marrow replacement a 'hair-on-end' appearance may be present. In our example of a newborn with osteopetrosis (Fig.6), the ossicles are markedly sclerotic and the baby was born deaf.

2. Van Buchem's disease and Sclerosteosis

Closely related, both diseases are caused by disorders of sclerostin production, an anti-anabolic protein produced by the osteocyte. Radiologically, both diseases demonstrate generalised thickening and sclerosis of both skull tables, mandible, ribs and diaphyses of long bones. Cranial nerve palsies are common, as in our case of a patient with bilateral sensorineural deafness due to narrowing of the IAMs (Fig. 7). Syndactyly and increased height are also seen in sclerosteosis and distinguish the diseases from each other. Both disorders are extremely rare.

EXTRINSIC CAUSES

Common:

1. Chronic phenytoin use

Phenytoin upregulates growth factor B1 and other proteins, which stimulate osteoblast proliferation and differentiation. Calvarial thickening is often seen in association with cerebellar atrophy; the relationship is long-established in both the literature and clinical practice.

Rare:

2. Hyperostosis crani ex vacuo

Hyperostosis crani ex vacuo is seen in patients who have undergone shunting for hydrocephalus (as seen in our example of a child with a long term ventriculoperitoneal shunt (Fig. 8)). It is likely related to intracranial hypotension.
The condition is also seen in patients who have advanced cerebral atrophy and even seen unilaterally in hemiatrophy as in our example of a patient with Sturge Weber disease (encephalotrigeminal angiomatosis) (Fig. 9). Histologically, increased cancellous spaces with normal trabecular bone is seen in hyperostosis crani ex vacuo. Radiologically, in the absence of any history, the condition may possibly be distinguished by increased inner table thickening compared with the outer table or more likely the observation of a shunt in situ.\textsuperscript{20}
Images for this section:

Fig. 1: Hyperostosis Interna, with the characteristic inner table bony nodular overgrowth

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Fig. 2: Heterogenous quality to the bone of the skull vault due to diffuse metastases from a primary breast carcinoma.

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Fig. 3: Diploid expansion due to medullary hyperplasia in thalassemia. The very low signal inner and outer skull tables are thinned.

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Fig. 4: Paget's Disease of bone, displaying the coarse, thickened trabeculations characteristic of the disease

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Fig. 5: Polyosteotic fibrous dysplasia 'ground glass change' in the expanded bones. The patient also suffered from precocious puberty and had a diagnosis of McClune Albright Syndrome.

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Fig. 6: Osteopetrosis in a newborn, showing markedly sclerotic ossicles. The baby was born deaf.

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Fig. 7: Van Buchem Disease, in a patient with bilateral sensorineural deafness due to marked narrowing of the IAMs

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Fig. 8: Hyperostosis crani ex vacuo in a patient with a long term in situ ventriculoperitoneal shunt. This is believed to be related to chronic intracranial hypotension.

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Fig. 9: Unilateral hyperostosis crani ex vacuo: Hemiatrophy has occurred on the left secondary to Sturge-Weber syndrome (encephalotrigeminal angiomatosis). There is associated overlying hemihyperostosis.

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Conclusion

A wide variety of causes can lead to skull vault thickening as described. The general radiologist should be able to offer a short differential diagnosis or, in cases where the appearance is classic, a confident diagnosis. A pictorial review collating some of the major and rarer causes of skull vault thickening has been prepared for this purpose.

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