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Craniolacunia: A Case Report with CT Imaging Findings

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Abstract | **Case Report**

Background: Craniolacunia, also known as Lückenschädel or lacunar skull, is an uncommon developmental abnormality of the calvarium characterized by multiple irregular osseous lucencies separated by residual bony trabeculae. It is frequently associated with congenital abnormalities of the central nervous system and may be encountered in infants undergoing neuroimaging for hydrocephalus or other neurological manifestations. ***Case presentation:*** An 11-month-old female infant with a history of hydrocephalus treated with a ventriculoperitoneal shunt presented with signs of raised intracranial pressure. Non-contrast brain CT revealed severe tetraventricular dilatation. Bone-window images demonstrated multiple irregular calvarial lacunae of variable sizes, separated by preserved bony trabeculae, producing a characteristic reticulated appearance suggestive of craniolacunia (Lückenschädel). Further MRI evaluation may be considered to investigate associated congenital anomalies ***Discussion:*** CT is particularly useful for characterizing calvarial abnormalities and evaluating associated intracranial findings. Recognition of craniolacunia should prompt a systematic assessment for associated congenital malformations, particularly neural tube defects and abnormalities of the craniocervical junction. Differential diagnoses include prominent convolutional markings and other causes of calvarial remodeling. ***Conclusion:*** Craniolacunia is an important radiological finding in infants with neurological abnormalities. Careful assessment of the skull vault on bone-window CT images, combined with a comprehensive evaluation of the intracranial structures, is essential for accurate diagnosis and appropriate investigation of associated abnormalities.

Keywords: Craniolacunia; Lückenschädel; calvarial abnormality; computed tomography; hydrocephalus; infant.

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1. INTRODUCTION

Craniolacunia, also known as Lückenschädel or lacunar skull, is an uncommon developmental abnormality of the cranial vault characterized by multiple irregular areas of thinning or deficient ossification, separated by residual bony trabeculae. It is predominantly identified during infancy and early childhood, when the calvarium is still undergoing development and ossification.

Craniolacunia has been classically associated with congenital abnormalities of the central nervous system, particularly neural tube defects and certain craniocervical junction malformations. Although its precise pathogenesis remains incompletely understood, it is generally considered a developmental disturbance of calvarial ossification.

Computed tomography (CT) plays an important role in its identification because it provides detailed assessment of the skull vault and allows simultaneous evaluation of intracranial abnormalities. Recognition of this finding should therefore lead to a systematic examination of both the calvarium and the underlying brain.

The aim of this report is to describe the CT findings suggestive of craniolacunia in an infant and to highlight the importance of evaluating associated intracranial abnormalities and relevant differential diagnoses.

2. CASE PRESENTATION

An 11-month-old female infant with a history of hydrocephalus treated with a ventriculoperitoneal (VP) shunt was evaluated for clinical features of raised intracranial pressure. Brain CT was performed to assess ventricular size, cerebral parenchymal abnormalities, and the position of the shunt catheter.

2.1. CT Findings

Non-contrast brain CT revealed marked dilatation of the entire ventricular system, involving the

lateral, third, and fourth ventricles, consistent with severe tetraventricular hydrocephalus.

On bone-window images, the calvarium demonstrated multiple well-defined, irregularly shaped osseous lacunae of variable sizes, separated by preserved bony trabeculae, producing a characteristic reticulated or honeycomb-like appearance of the cranial vault. These findings were suggestive of craniolacunia (Lückenschädel)

The combination of the calvarial findings and the intracranial abnormalities warrants further evaluation for associated congenital malformations, guided by the clinical findings and, when appropriate, brain and spine MRI.

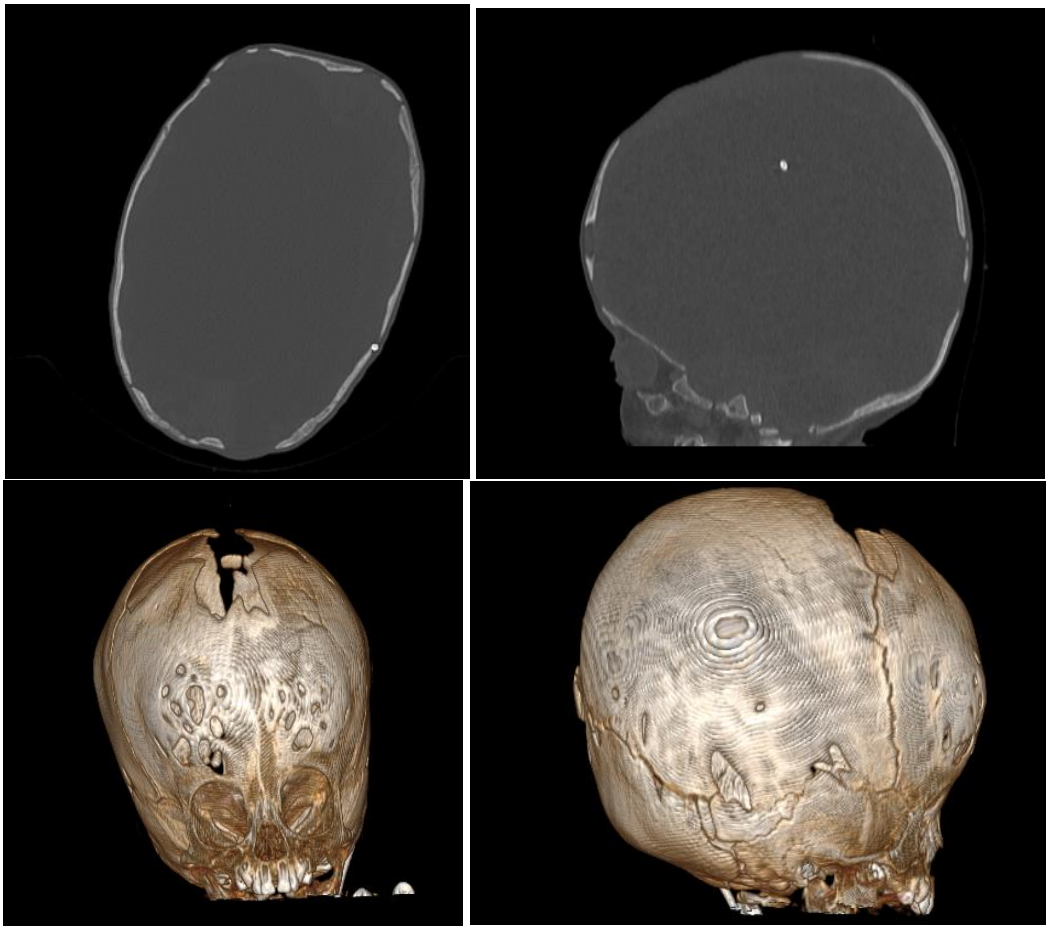


Figure 1: Brain CT images in axial and sagittal planes, with three-dimensional (3D) bone reconstruction, demonstrating multiple irregular calvarial lacunae of variable sizes, separated by preserved bony trabeculae, producing a characteristic reticulated appearance consistent with craniolacunia (Lückenschädel)

3. DISCUSSION

3.1. Definition and Pathogenesis of Craniolacunia

Craniolacunia is a developmental abnormality of the cranial vault characterized by multiple irregular areas of calvarial lucency separated by residual bony ridges or trabeculae. It is most readily recognized during infancy, when the skull is undergoing active ossification.

Its pathogenesis is not fully established. The condition is generally attributed to abnormal development or ossification of the membranous bones of the skull. Craniolacunia has been described in association with congenital central nervous system abnormalities, particularly open neural tube defects and certain malformations involving the craniocervical junction.

Although the finding may be striking on imaging, craniolacunia should not be considered sufficient, on its own, to establish a specific underlying syndrome. The clinical history and the presence or absence of associated abnormalities are essential to its interpretation.

3.2. CT Features and Diagnostic Approach

CT is particularly valuable for evaluating craniolacunia because of its high spatial resolution and its ability to characterize the calvarium in bone windows. The characteristic appearance consists of multiple irregular osseous lacunae separated by residual bony trabeculae, producing a reticulated or honeycomb-like configuration.

The radiological assessment should describe:

- The number, size, shape, and distribution of the calvarial lacunae.
- The thickness and continuity of the residual bony trabeculae.
- The extent of involvement of the cranial vault.
- The presence of associated cranial deformity or other osseous abnormalities.

Multiplanar CT reconstructions may help define the distribution of the lesions. Brain-window images should be reviewed concurrently to assess ventricular size, the cerebral mantle, midline structures, the posterior fossa, and other possible intracranial abnormalities.

3.3. Differential Diagnosis

The principal radiological differential diagnosis includes prominent convolutional markings, traditionally referred to as a "copper-beaten skull." These markings correspond to impressions produced by the cerebral gyri on the inner table of the skull. They may be physiological in children or become more prominent in association with sustained increases in intracranial pressure.

In contrast, craniolacunia is characterized by irregular areas of deficient calvarial ossification separated by residual bony trabeculae. The distinction depends on the morphology of the abnormalities, their distribution, and their appearance on bone-window images.

Other causes of calvarial remodeling should be considered according to the patient's age, clinical history, and imaging findings. In an infant with hydrocephalus and a VP shunt, comparison with previous examinations may also help determine whether calvarial changes are developmental or acquired. The possibility of acquired calvarial remodeling should not be confused with congenital craniolacunia without careful image review.

3.4. Associated Congenital Abnormalities

The identification of craniolacunia should prompt a systematic search for associated central nervous system abnormalities. These may include open neural tube defects, such as myelomeningocele, and malformations of the craniocervical junction, including Chiari II malformation.

The absence of visible cerebellar tonsillar herniation on CT does not exclude every craniocervical abnormality. MRI may be useful for assessing the posterior fossa, cerebellar tonsils, brainstem, spinal cord, and possible spinal dysraphism when clinically indicated.

In this case, the marked dilatation of the ventricular system was the main intracranial finding. The assessment of the midline structures was limited by the severity of the ventricular dilatation and the resulting distortion of the intracranial anatomy. The description of the calvarial lacunae on bone-window images remains the key finding supporting the diagnosis of craniolacunia.

4. CONCLUSION

Craniolacunia is an uncommon developmental abnormality of the cranial vault that can be recognized on CT by the presence of multiple irregular calvarial lacunae separated by residual bony trabeculae. Its identification in infancy warrants a comprehensive assessment of the intracranial structures and consideration of associated congenital malformations.

CT provides valuable information about calvarial morphology, ventricular dilatation, cerebral parenchymal abnormalities, and shunt catheter position. MRI may provide additional information when associated brain, posterior fossa, craniocervical, or spinal abnormalities are suspected.

A systematic radiological approach, combining bone-window assessment with careful evaluation of the intracranial contents, is essential for accurate characterization and appropriate further investigation.

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