EXPANSIVE DECOMPRESSION CRANIOPLASTY IN CRANIODIAPHYSEAL DYSPLASIA: A CASE REPORT

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NO DISCLOSURE
INTRODUCTION
CRANIODIAPHYSEAL DYSPLASIA

Very rare autosomal recessive form of craniotubular bone dysplasia mainly affecting infants and young children

Progressive hyperostosis and sclerosis of the cranial vault and intracranial foramina leading to increased intracranial pressure and cranial nerve deficits

Current literature states that mortality of most patients with this disorder is mainly due to eventual medullary compression caused by the foramen magnum obliteration

Multidisciplinary approach is required which includes low-calcium diet, calcitonin therapy, surgical decompression
This is a unique case of a 4-year-old female diagnosed with craniodiaphyseal dysplasia presenting with progressive symmetrical cranial hyperostosis with “leontiasis ossea” like features. There was also significant signs of increased intracranial pressure (ICP) manifested by progressive headache and deterioration of sensorium along with multiple cranial neuropathies such as binocular visual deterioration and hearing loss.

Serial CT scan monitoring showed significant cerebral compression in the supratentorial space with hyperostotic obliteration of the optic canal and internal acoustic foramen despite a stable foramen magnum diameter.
A bicoronal incision was done followed by Expansion Cranioplasty involving separation of the frontal bone from the temporoparietal calvaria.
Intra-operatively, the calvarium was about three centimeter thick. Operative blood loss was approximately 250ml with total duration of two hours.

Post-operative outcome noted a remarkable resolution of ICP symptoms characterized by improvement in sensorium and resolution of headache. However, visual and hearing loss persisted.
Patient was then discharged improved with monthly follow-up. However, eight months after discharge, patient developed a non-communicating hydrocephalus and pseudomeningocele requiring a ventriculoperitoneal shunt.
Supratentorial expansions cranioplasty can be used as a symptomatic surgical intervention for craniodiaphyseal dysplasia with stable foramen magnum diameter. Close follow-up surveillance is needed for early recognition of further progression of the disease.