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Primary calvarial cavernous haemangioma in a child. A case report

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ABSTRACT

A wide variety of bone lesions is encountered on the skull and usually with poor clinical presentations and non-specific radiological features. Primary cavernous hemangioma is one of these rare calvarial lesions that the symptoms are limited to a simple subcutaneous mass frequently neglected by the patient. Although seen in all ages from neonate to elderly; few cases were reported in the literature of patients in the first decade of life. We report a case of four years old boy with primary calvarial cavernous hemangioma in order to help to understand this pathology and to urge physicians to explore any chronic subcutaneous mass at any age.

INTRODUCTION

Skull hemangioma is a rare pathology with minimal clinical presentation and non-specific images. The first known description of this lesion in literature was in 1845 by Toynbee ^[1]. Skull hemangiomas are classified into capillary and cavernous types; with cavernous type being more frequently seen ^[1,2,3,4,5,6]. We present a case of four years old boy with primary calvarial cavernous hemangioma.

CASE PRESENTATION

The patient is a four years old boy with trisomy 21. The parents consulted for the appearance of a right fronto parietal subcutaneous mass; of barely 1 cm of diameter. A brain CT was performed objectified an extra axial right fronto parietal lesion of 42 x 29 mm; isodense, with intense homogenous enhancement after contrast injection, and bone destruction from where the lesion get through to the subcutaneous region; there was also a circumferential osteophytic bone reaction (Figure 1). The patient was operated and the lesion was totally removed through a frontoparietal bone flap. In post-operative there were no complications. Pathology study was in favor of bone cavernous hemangioma. No supplementary management was necessary. Control images performed nine years later, showed no residual tumor or recidivism (Figure 2).

Keywords hemangioma, bone tumours, skull tumours

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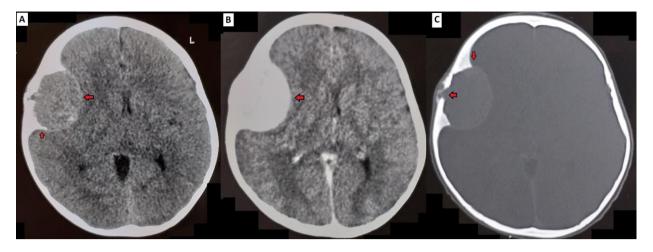


Figure 1. Preoperative brain CT; A: before injection; B: after injection; C: bone window. The images objectify an extra axial right fronto parietal lesion; it appears spontaneously isodense (A, horizontal arrow), with intense homogenous enhancement after contrast injection (B, horizontal arrow), and bone destruction from where the lesion get through to the subcutaneous region (C, horizontal arrow). There was also a circumferential osteophytic reaction from the inner calvarial bone (A and C, vertical arrows).

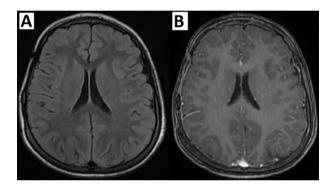


Figure 2. Nine years post-operative MRI showing no tumor recidivism. A: axial FLAIR sequence; B: axial T1 injected image.

DISCUSSION

Primary calvarial hemangiomas (PCHs) are rare lesions; they account of 0.2 % of all bone neoplasm ^[1,3,5,6,7,8]. Yang et al. reviewed 93 cases of PCH reported in the literature from 1845 to 2015; according to this paper most patients were in the fourth and the fifth decade ^[1,2]. Only 0.9 % of PCHs were seen in the first decade of life as in our case ^[2]. Frontal bone is a predilection location for PCHs with 44.1% of all cases ^[1,2]. Clinical presentation depends on the topography of the lesion. Subcutaneous mass and headaches were the most frequent reasons for consultation ^[1,2]; neurologic deficit is rarely seen ^[1,2,4,5,6,7,8]. Proptosis, impaired vision, facial nerve paralysis, tinnitus, and hearing loss are seen in orbital and skull base locations ^[1,2]. Bleeding from the lesion may be seen in the epidural or subarachnoid spaces ^[1,3,7]. On plane X-Rays and on CT scan, PCH is a lytic lesion with sclerotic rim; after contrast injection the enhancement is constant it could be heterogeneous or homogenous as in our case ^[2,3]. For us those finding were sufficient to indicate the surgery and to plan the resection strategy. Although not specific, MRI affords supplementary information about the relation of the lesion with the underlying dura, parenchyma, and vascular structures; it shows hypo to iso intense mass on T1 weighted images, hyperintense on T2 weighted images and on FLAIR sequences; there is a large enhancement after gadolinium injection ^[1,3]. PCHs are accessible lesions for surgical resection, thus total removal with acceptable margin is the treatment of choice ^[1]; this could be achieved through a skin incision and craniectomy centered on the lesion. In Adult the bone defect is correct usually with cranioplasty. As in our case the management of many other pediatric cases in the literature was limited to lesion removal without bone correction due to the young age face to future skull changing ^[4,8]; other teams preferred cranioplasty^[6] or remodeling^[5].

CONCLUSION

Subcutaneous mass could underestimate deep infiltrating and brain compressive lesion. This is the case of rare lesions like calvarial cavernous hemangioma which is seen in any age. This lesion is associated with the best prognostic if totally removed.

CONFLICTS OF INTEREST None.

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