Purely intraparenchymal solitary Infantile Myofibroma of the Central Nervous System without dural attachment

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Introduction

Infantile myofibroma, the most common fibrous disorder of infancy and early childhood, is characterized by the formation of mesenchymal tumors of skin, soft tissue, muscle, bone and viscera. Intracranial central nervous system (CNS) involvement has been reported with skull or dural involvement with variable intracranial extension. We report a rare case of a purely intraparenchymal solitary infantile myofibroma with no dural attachment or bony involvement in a 14-month old child.

To our knowledge, this is the first case of solitary and purely intra-axial (no dural or osseus involvement) infantile myofibroma that is also unifocal, e.g. not associated with multiple myofibromatous involvement of the CNS, calvarium or extracranial soft tissue or viscera.

Methods

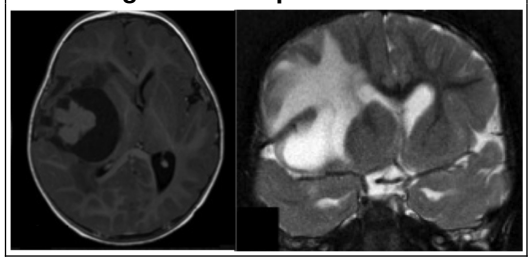
Case report and literature review

Case Presentation

Presentation

A 14-month-old boy presented with a 1-week history of left hemiparesis. Brain Magnetic Resonance Imaging revealed a large right hemispheric peri-Sylvian cystic mass with a mildly enhancing nodule with significant vasogenic oedema, mass effect and midline shift (Fig. 1). Whole-body PET scanning was negative as was chest XRAY and abdominal ultrasound.

Figure 1. Preoperative MRI

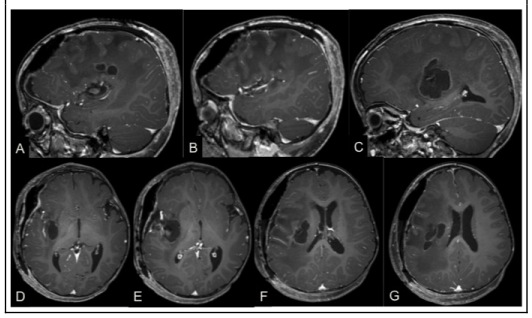


Surgery

The patient underwent gross total resection (GTR) of the purely intraparenchymal lesion (Fig 2).

Figure 2. Intraoperative photograph (A) and ultrasound (B)

Figure 3. Postoperative MRI confirming GTR



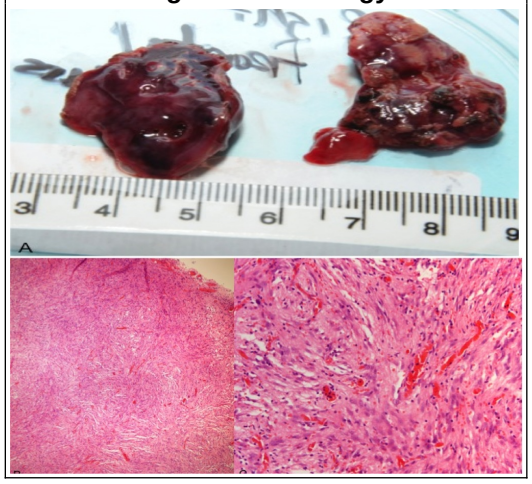
Outcome

At 5-month follow-up, the child has no deficits and MRI confirms GTR (Fig 3)

Pathology

Pathology revealed a spindle cell neoplasm of low cellularity/proliferation and smooth muscle actin expression (mesenchymal differentiation), consistent with a myofibroma (Fig. 4).

Figure 4. Pathology



Discussion

Myofibromatosis of infancy may occur in either a multicentric or solitary form. The solitary infantile myofibroma form is usually characterized by isolated myofibroma occurring most commonly in the head, neck, extremity or trunk. Involvement of the brain is rare, and when present they usually originate form the dura and extend to invade the skull and compress the brain. The multicentric form, which represents 25% infantile myofibroma, is characterized my multiple soft tissue, bony, and viscera.

Discussion (2)

Involvement of the CNS in the multicentric form is also rare, and usually has dural involvement with extension through bone and compression of parenchyma, but usually do not transgress the arachnoid or galea. We report a rare case of a purely intraparenchymal solitary infantile myofibroma with no dural attachment in a child. Clinicians should be alerted to the possible existence of this rare presentation of infantile myofibroma.

Conclusion

Myofibromatosis of infancy rarely involves the CNS, and usually has dural involvement with extension through bone and compression of parenchyma, but usually do not transgress the arachnoid or galea. We report a rare case of a purely intraparenchymal solitary infantile myofibroma with no dural attachment or multifocal myofibromas in a child. it is imperative to eliminate multi-systemic involvement as prognosis is directly related to involvement of viscera, such as the heart or gastrointestinal tract

Learning Objectives

1. Understand infantile myofibromatosis as a disease entity in children
2. Recognize that CNS involvement of myofibromatosis usally involves the skull or dura matter
3. Recognize the existence of the rare variant of a purely intraparenchymal myofibroma