

Multicompartmental congenital intracranial immature teratoma

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Abstract

Congenital intracranial tumors are rare and account for 0.5 to 1.5% of all childhood tumours. We report a case of a 3 week old baby presenting with multi compartmental congenital intracranial immature teratoma, first of its kind in the literature. The child had gross total excision in two stages with aid of neuronavigation. The short term outcome was good. The four years of follow-up with serial imaging showed no tumour recurrence with a stable hydrocephalus after shunting. However, there is global developmental delay with full time dependence of care giver.

INTRODUCTION

Congenital intracranial tumors are rare conditions and account for 0.5 to 1.5% of all childhood brain tumor.¹⁻² Teratomas are the most common type of congenital central nervous system tumor³ accounting for 28.8 to 50 %² of these tumors. We present a case of an infant with massive multicompartmental congenital intracranial immature teratoma, first of its kind to be reported in the literature.

CASE REPORT

A three week old baby boy presented with increasing head circumference. Computer tomography (CT) of the head revealed hydrocephalus with a large intracranial tumor containing multiple foci of calcification. Germ cell tumor markers were negative. Magnetic resonance imaging (MRI) scan revealed a multicompartmental heterogeneous intraventricular tumor with non-homogenous enhancement. (Figures 1a, 1b, 1c)

The child underwent image guided craniotomy and gross total excision of the tumor in two stages. The *MEDTRONIC AXIEM™* image guidance system which employs electromagnetic waves for probe tracking was utilized. The navigation system assisted in defining the ideal entry point along the natural CSF clefts in order to devascularise the tumour prior to debulking. A staged procedure

was needed due to blood loss. Gross total excision was achieved.

The histology demonstrated a heterogeneous tumor composed of an admixture of tissues of the three germ cell layers consisting of mature teratomatous elements. Immature teratomatous elements include some cartilaginous primitive mesenchyme and neuroepithelial tubules seen in at least four foci (Figure 2a, 2b). The histological picture was consistent with immature teratoma (Norris grade II).

Post operative period was complicated by seizures, central diabetes insipidus and subdural collection which were managed appropriately.

Four months after surgery, at the age of six months the child is attempting to turn and is able to move all his limbs. The postoperative MRI at 4 months after surgery confirms the near total excision with multi septated hydrocephalus (Figure 3a, 3b, 3c). The infant underwent ventriculoperitoneal shunt insertion after endoscopic fenestration of several fibrotic septa. The child continued to have clinical follow up and serial imaging at least yearly. The child is now four years old, the recent imaging showed no tumour recurrence with a stable ventricular size (Figure 4a, 4b, 4c). He has significant global developmental delay with epilepsy controlled with medication. He is fully dependant on a care giver.

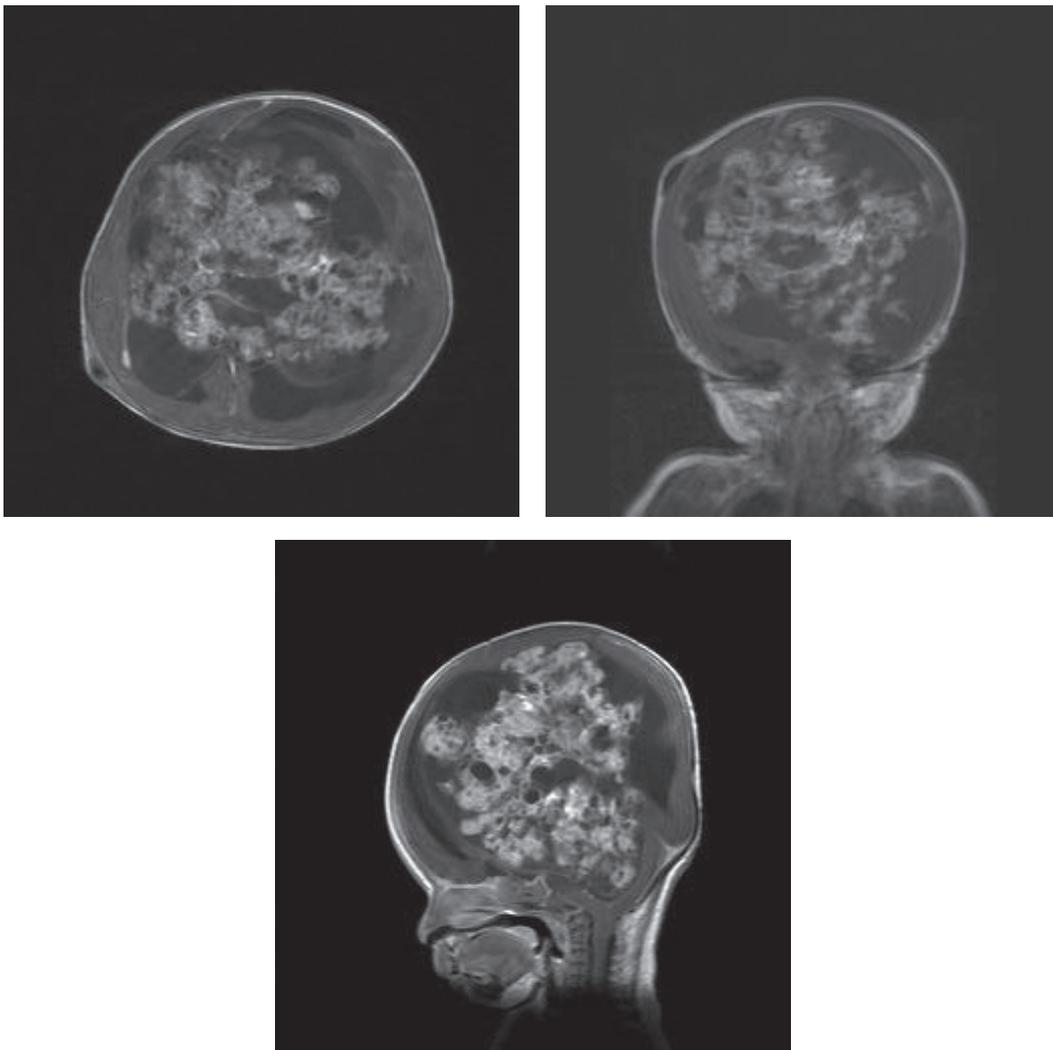


Figure 1. MRI brain, contrast enhanced T1 weighted (a) axial image, (b) coronal image, (c) sagittal image showing a large intraventricular heterogenous mass with cystic and solid areas

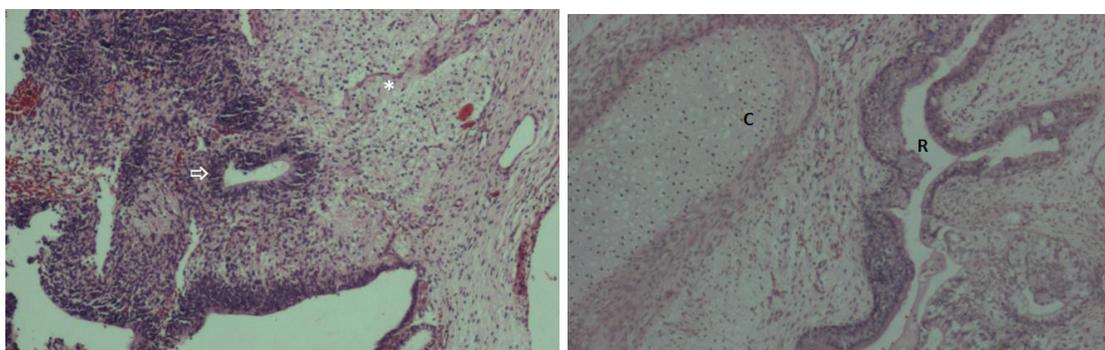


Figure 2. (a) Teratoma showing immature elements comprising neuroepithelial rosettes [↔]. The tissue to the right of the immature element is mature neural tissue [*] (Hematoxylin & Eosin, magnification x40). (b) Area of tumour showing hyaline cartilage (C) and ciliated respiratory type epithelium (R) amidst loose stroma (Hematoxylin & Eosin, magnification x40)

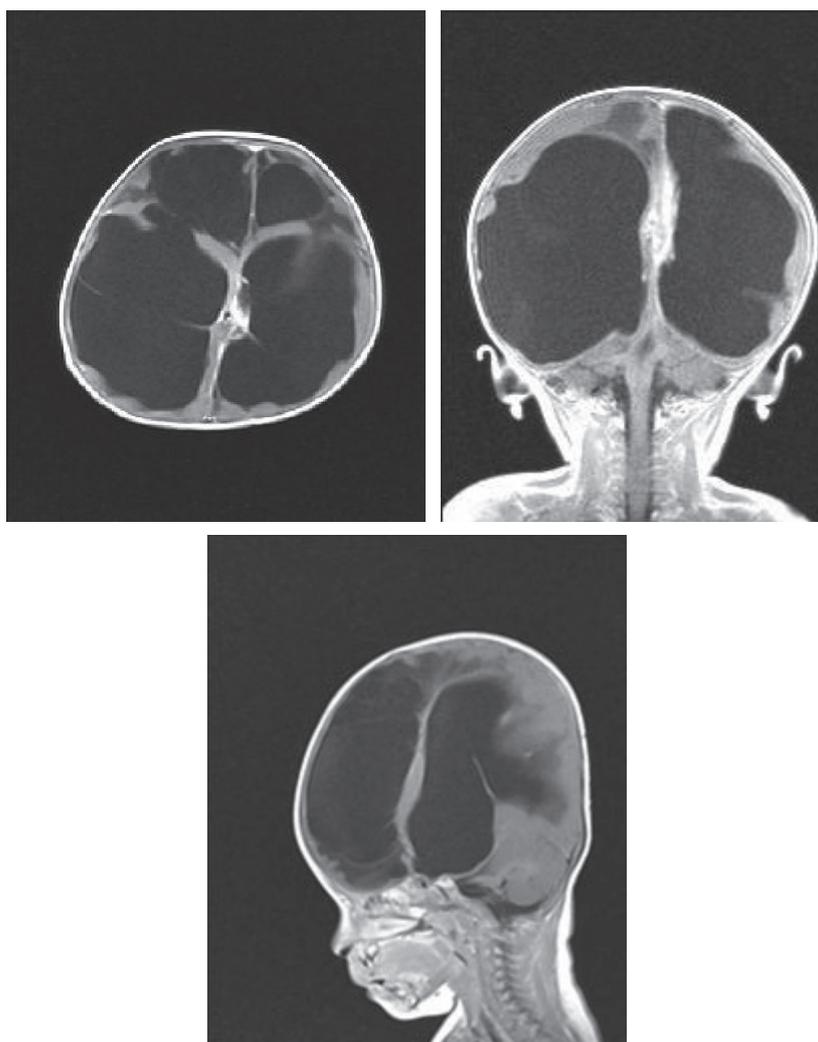


Figure 3. MRI brain, T1 weighted with contrast enhancement (a) axial image, (b) coronal image, (c) sagittal image; 4 months after surgery revealing near total excision of tumor with hydrocephalus and multiple septa.

DISCUSSION

Congenital intracranial tumors account for approximately 0.5-1.5% of all childhood brain tumors.² Germ cell tumors comprise 0.4-3.1% of all intracranial tumors and teratomas constitute 9-30% of this germ cell tumors.⁴

Immature teratomas have been reported to have an unpredictable biologic behaviour and the value of chemotherapy in their treatment is not yet established.⁵ However, there is a case report of recurrent immature teratoma being successfully treated with chemotherapy.⁵ The prognosis of immature teratoma has been correlated to age, primary site, histologic grading and extent of excision.⁵ There are reports of spontaneously differentiation and maturation of

immature teratomas to the mature teratomas on recurrence.

Surgical excision has been the mainstay in managing intracranial immature teratomas in children.⁶ In our case, the utilisation of the *MEDTRONIC AXIEM™* electromagnetic tracking technology permitted the application of neuronavigation on an infant. The neuronavigation aided in determining the ideal entry zone. Therefore, allowing circumferential dissection of the tumor off the ventricular wall and devascularising the tumor as much as possible prior to debulking. By this approach we managed to remove at least two thirds of the tumor at the first stage and the remainder at the second stage.

A review of 80 children with brain tumors who were operated in the first two years of life; recorded

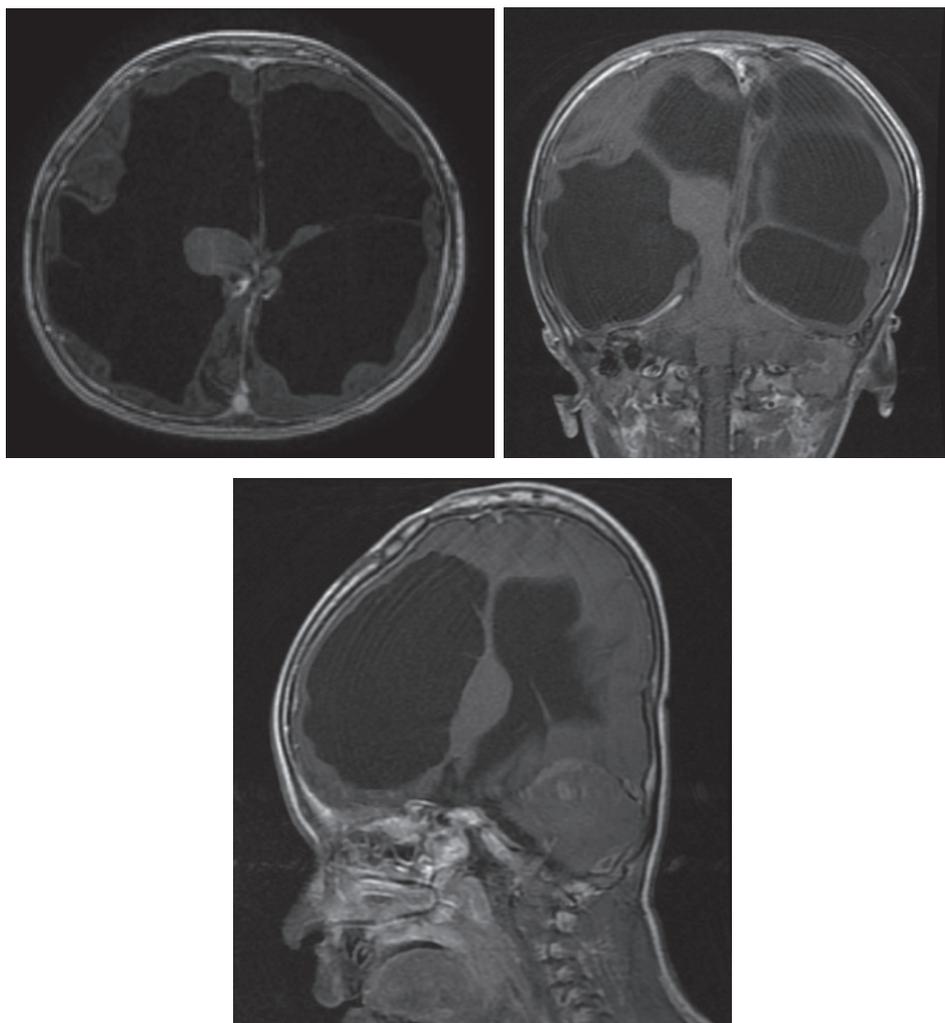


Figure 4. MRI brain, T1 weighted with contrast enhancement (a) axial image, (b) coronal image, (c) sagittal image; 42 months after surgery revealing no recurrence of tumour and stable ventricular size.

a surgical mortality of 17.4% with 46% having normal physical and intellectual performance while 75% had sufficient autonomy in daily life.¹ Although these figures are not specific for immature teratoma, they do reflect that surgical excision is a feasible option in this age group with acceptable quality of life in the long term. Sano in reviewing her 153 cases of intracranial germ cell tumor records a 5 and 10 year survival rate of 86% each for immature teratoma.⁷ However, the mental and physical developments of these children need to be monitored.

In conclusion, a multi-compartmental large intracranial immature teratoma was resected in a staged manner with the aid of electromagnetic neuronavigation. The short term results were good. After four years there is no evidence of tumour recurrence. However, despite gross total excision

and controlled hydrocephalus, there is significant delay in global development. Most probably due to the poor development of the cortic mantle resulting from congenitally raised intracranial pressure.

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