

## Management of Hemispheric Cerebral Mature Teratoma in New Born

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### Abstract

Teratoma is a congenital tumor composed of an admixture of different tissue types representative of ectoderm, endoderm and mesoderm. Their intracranial location is rare, preferentially arise in the midline structures, and more diagnosed in pediatric population. We presented an unusual case of a huge mature intracranial teratoma in a female new born. In MRI, the lesion occupied the place of the totality left hemisphere and deformed the normal cerebral structures. The patient underwent great total resection of the tumor after two stages. On per surgery the tumor was lobulated, containing heterogeneous solid and cystic components. Histological study has established the diagnosis of mature teratoma. Follow-up at 6 months did not show any evidence of recurrence.

**Keywords:** New born; MRI; Two stages; Mature teratoma

### Introduction

Teratomas are a rare congenital tumors representing approximately 0.5% of all intracranial tumors [1]. They are derived from pluripotent cells and composed of tissue originating from all three germinal layers: endoderm, mesoderm, and ectoderm [2]. Pathologically, teratomas are classified into mature, immature and malignant types. Mature teratomas are composed of well-differentiated elements, whereas immature teratomas contain components resembling fetal tissues [3]. Teratomas are the most common brain tumor in new-borns and they most frequently found in the rear of the third ventricle near the pineal gland and above the pituitary gland.

In this report, we described an unusual case of a huge mature teratoma in new-born female. In combination with literature review, we discussed the pathogenesis, imaging features and treatment of intracranial teratomas.

Surgery is the standard treatment for accessible tumors and could be curative.

### Case Report

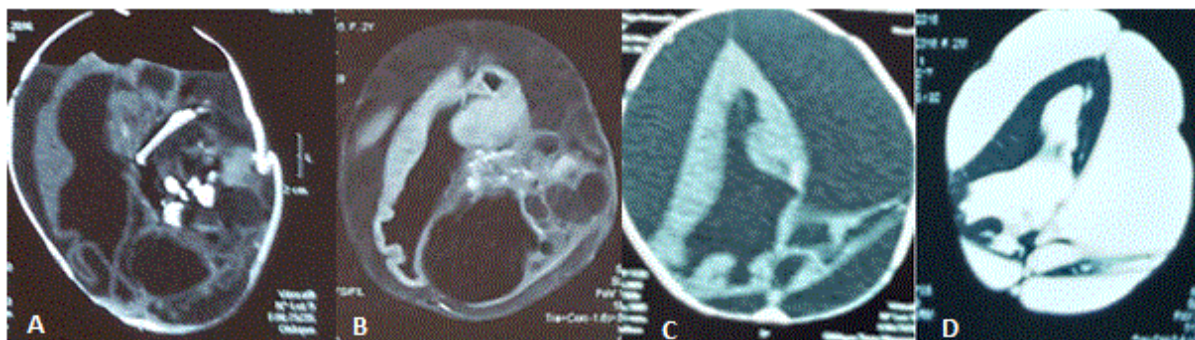
A 22-days-old new born female patient without family history of congenital malformations presented with rapid head enlargement. She was macrocephalic with a normal appearing body. Physical examination revealed newborn female with signs of maturity. She was lethargic and has bulging anterior fontanel, setting sun sign in eyes, prominent veins over forehead and widely opened cranial sutures with head circumference at 45 cm (Figure 1).



**Figure 1:** Photos showing new born before surgery and 6 months after surgery.

All other physical findings were normal. Congenital infections were ruled out.

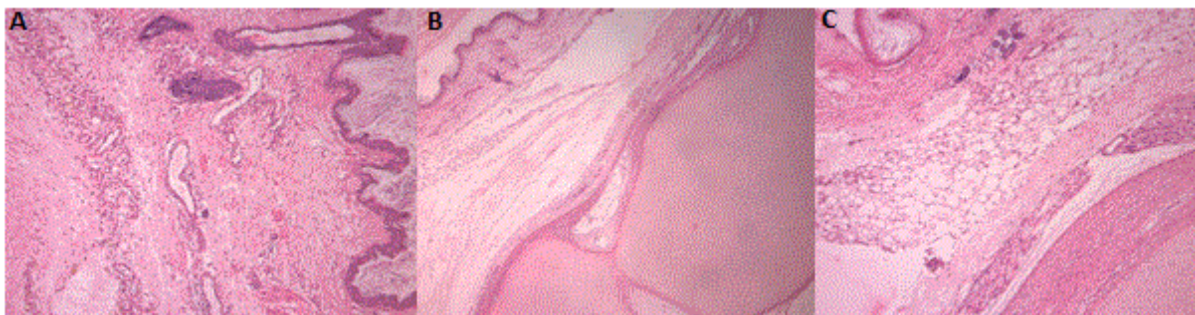
The CT (Computed Tomography) scan also MRI (Magnetic Resonance Imaging) study showed a large solid cystic lesion in left fronto-parieto-temporal side with mixed density: cystic, solid, fat, associated with organoids macrocalcifications, measuring 10/7 cm of transverse axis, and extended over a height of 12 cm. It was compressing ipsilateral and third ventricle, with severe reduction of the cerebral parenchyma; responsible of an acute huge hydrocephalus. The tumor strongly enhanced with gadolinium and was multilobulated. It has an extension towards the posterior cerebral fossa (Figure 2).



**Figure 2:** (A) CT scan, (B) axial MRI showing huge teratoma, (C) CT scan, (D) axial MRI showing great total resection.

We performed surgery in two stages through a large left fronto-parietal craniotomy which result in near-total resection (Figure 2). The lesion was well delimited and enveloped by a thick white capsule. We firstly opened the capsule at the top end and evacuated its soft, yellowish xanthochromic and greasy content, the rest of content were debulked (calcification, organoid bone, tissue and fat) and then its capsule totally removed. While fat tissue attached to the splenium was left to avoid eventual arterial injury.

Histological examination of the different components removed found a well differentiated adipose tissue, mature bone and keratin-producing squamous epithelium, which were compatible with the diagnosis of a mature teratoma (Figure 3).



**Figure 3:** Histopathology of huge teratoma Color Hex 100 note: (A) a regular and keratinized epidermis with its epidermal appendages in the dermis, (B) The bronchial teat lined by a regular respiratory epithelium, (C) mature adipose, muscular and epithelial structures.

Postoperatively, the cranium collapsed and the patient recovered uneventfully and was discharged ten days later. At one month, the baby manifested an increase of the hydrocephalus and she underwent a left Ventricular Peritoneal Shunt.

In the follow-up, neither neurological deficits nor recurrence signs were found except transient dysfunction of the shunt which would not be sufficient alone and a second Ventricular Peritoneal Shunt has placed at right side.

## Discussion

Intracranial germ cell tumors are rare comprising about 0.3% to 3.4% of all intracranial tumors [4]. The prevalence is much higher in the earlier decades of life [5], with one peak in the neonatal and another peak in children ages of 5-14 years [6].

Most intracranial germ cell tumors arise from midline structures, with the pineal gland being the most common, followed by the supra sellar region [4]. Mature teratomas are well-differentiated germinal

tumors [7]. They contain fully mature tissues of ectodermal, mesodermal, and endodermal origins. Microscopically, they tend to consist of solid and cystic components of squamous epithelium with keratin debris [7].

The etiology and pathogenesis of extragonadal teratomas remain unclear, but they are thought to arise from misplaced primordial germ cells, which become embedded in or near midline structures in the head [8].

MRI combined to CT are helpful to approach the nature of the lesion and may show components of mixed density that include fat and soft tissue, cartilage, and calcified tissues such as bone and teeth. Like our case, independent of their histology, congenital brain tumors mostly present as large, displacing masses with secondary obstructive hydrocephalus [7]. As the MRI features of teratomas are obvious, the differential diagnosis of congenital supratentorial tumors should also include primitive neuroectodermal tumors, gliomas, craniopharyngioma, and papilloma.

Histological examination is the mainstay to establish a definitive diagnosis of intracranial teratomas, and its histologic subtype.

The typical treatment for mature teratomas is radical resection wherever possible, which was successfully done in our case. The persistence of macrocranium may be due to hydrocephalus which often needs drainage. Mature teratomas are benign and have been reported to have survival rates up to 93% at 10 years [9]. Since surgery is curative and no further adjuvants therapy are required. Immediate neurosurgical intervention should be indicated before neurological worsening from mass effect or obstructive hydrocephalus [2,10-12].

At the opposite of our case, the few reported attempts at total or subtotal tumor resection have had poor outcomes [7].

## Conclusion

Teratomas occur rarely in the central nervous system. Pineal region is the commonest site in brain. The available information regarding teratoma of perinatal period is limited. Then a collaborative study would be helpful to clarify the complete clinical picture and to establish a prognosis factors after their management. Total surgical resection remains the treatment of choice for mature teratoma.

A long-term follow-up period is recommended.

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