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Case Report

Surgical Management of Huge Neonatal Brain Tumor: A Case Report

Ahmed Osama El Ghannam,[1] Mariam Abd Elrasheed Ghanem,[2] Ahmed Mustafa Abd Elsalam[3]
[1]Town Hospital, Fifth Settlement, Cairo, EGYPT
[2]Department of Neurosurgery, Faculty of Medicine for Girls Al-Azhar University, Cairo, EGYPT
[3]Department of Pediatric Oncology, National Cancer Institute, Cairo University, Cairo, EGYPT

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BACKGROUND: Brain tumors are uncommon in infants younger than 6 months old. The most common types of neonatal brain tumors include teratomas, astrocytomas, embryonal neoplasms, choroid plexus tumors, craniopharyngiomas, gangliogliomas, ependymal tumors, meningeal tumors, and other miscellaneous malignancies. Treatment algorithms are not well-established for infants less than 6 months of age, but surgery remains a critical component of most therapeutic approaches.

CASE PRESENTATION: In this case report, we present a rare instance of a congenital massive Xanthoastrocytoma occupying the left frontal lobe in a 28-day old neonate. Following total surgical resection, a favorable outcome was obtained without the need for adjuvant chemo or radiotherapy, with follow-up for 2 years.

CONCLUSION: Brain tumors in infants are quite uncommon, especially ones as large as the one reported in this case. Adjuvant therapy is not always necessary to produce a satisfactory outcome after gross complete surgical resection.

KEYWORDS: Infants, Neonatal brain tumor, Xanthoastrocytoma.

INTRODUCTION

Brain tumors are exceedingly rare in infants younger than 6 months of age, accounting for only 0.5-1.5% of all pediatric brain neoplasms.[1] These tumors typically present within the first 60 days after birth. The most common types of neonatal brain tumors include Teratomas, Astrocytomas, Embryonal neoplasms, Choroid plexus tumors, Craniopharyngiomas, Gangliogliomas, Ependymal tumors, meningeal tumors, and other miscellaneous malignancies.[2]

In early infancy, brain tumors commonly present with macrocephaly, bulging fontanels, vomiting, and aberrant eye movements. All these symptoms are indicative of elevated intracranial pressure. Seizures, weakness, and congenital anomalies such as cleft lip/palate, cardiac defects, and urinary tract malformations may also be associated findings.[3] Cranial ultrasonography and magnetic resonance imaging (MRI) represent the mainstays of diagnostic evaluation.

Surgical management of these tumors poses immense challenges in the infant population. Treatment algorithms are not well-established for infants less than 6 months of age, but surgery remains a critical component of most therapeutic approaches, facilitating the minimization of potential harm from radiation and chemotherapy.[4]

Town Hospital Neurosurgery team handled a challenging case of huge intracranial neoplasms below the age of one month-old.

Correspondence:
Ahmed Osama El Ghannam
Town Hospital, Fifth Settlement, Cairo, EGYPT
Email: Ay.elghannam@gmail.com

CASE PRESENTATION

A male infant, 28 days old, had tonic-clonic seizures in his right upper limb, bulging fontanel, vomiting, and macrocephaly. A neurosurgical team was principally responsible for the patient's care. The first brain imaging revealed a large tumor measuring 9.5 × 9.3 × 8.8 cm in the left frontal lobe. It had a heterogeneous enhancement pattern and contained both solid and cystic components. Surgical resection was therefore recommended **(Fig. 1).**

Surgical Technique: The patient was placed in a supine position with the head fixed with a plaster headrest. A left frontotemporal craniotomy was then carried out, along with the attachment of a bone flap. The dura mater was then opened and reflected basally **(Fig. 2).** The tumor was found to be a cortical-based lesion. A full microscopic resection was accomplished by making a direct incision into the tumor mass **(Fig. 2).** Under a microscope, the tumor's large solid component was firm, while other areas showed signs of thrombosed veins and were slightly softer. A small cystic portion of the tumor was filled with a translucent fluid. After meticulous hemostasis, the bone flap was put back into place and sutured to the periosteum. The patient was extubated in the operating room and emerged fully conscious with an intact motor function.

Pathological findings: The pathological examination revealed a grade II Xanthoastrocytoma, with some areas showing grade III features. These included moderate cellularity, pleomorphic cells, vesicular nuclei, eosinophilic cytoplasm, and spindle cells arranged in sheets and nests, foci of increased cellularity with a mitotic count of 2/10 high-power fields were noted, and increased vascularity was also observed.

Immunohistochemical staining showed focal positivity for OLIG2 and positivity for EMA in very few cells (membranous pattern). Cytokeratin was negative, and INI-1 expression was retained. ERG highlighted the endothelial cells. The Ki-67 proliferation index ranged between 5% and 20% in the areas with increased cellularity.

Postoperative course: A ventriculoperitoneal shunt was inserted to treat the patient's hydrocephalus, which appeared four days after surgery **(Fig. 3).** With repeated follow-ups, the patient's clinical condition improved. The oncologist did not suggest adjuvant therapy because the histological analysis showed a mitotic count of 2/10 high-power fields, that is below the threshold for malignant transformation.

Serial MRI follow-up, with and without contrast, every

3 months showed no mass residual or recurrence. The patient's growth and development remained consistent with his age. This publication had been approved by the institutional review board (IRB) of the Ethical Committee, Al-Azhar University. The consent for operation and publication was obtained.

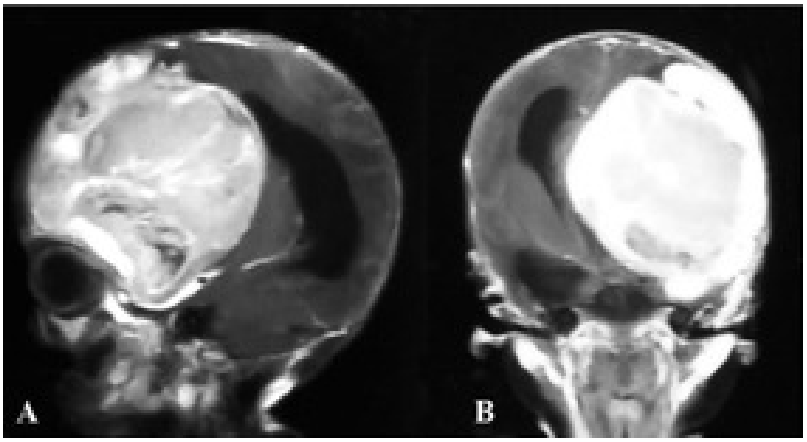

Fig 1: (A), (B) Sagittal and coronal T1-weighted magnetic resonance imaging scans with gadolinium contrast revealed a huge, heterogeneous, enhanced solid mass with a cystic lesion in the left frontal lobe, along with massive ventricular dilatation.

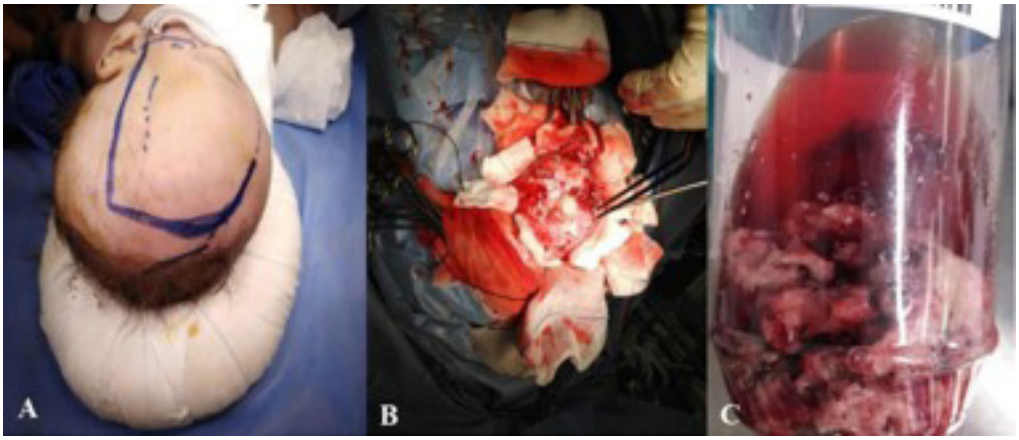

Fig 2: (A) Intraoperative photo revealed the position of the patient and marks for skin incision. (B) Intraoperative photo revealed both the attached bone flap and the dura flap that were reflected to one side; corticectomy was performed using bipolar cautery. (C) The resected tumor sample.

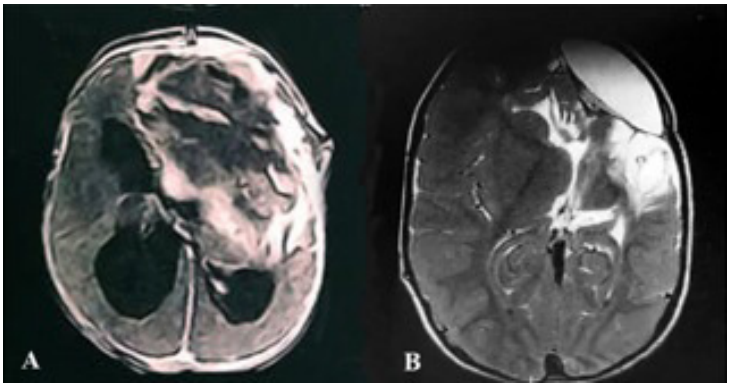

Fig 3: (A) Postoperative MRI showing postoperative hydrocephalus. (B) The last MRI follow-up, axial T2 image, 2 years postoperatively, revealed no tumor residue or recurrence.

DISCUSSION

We report an extremely rare case of congenital Xanthoastrocytoma; these brain tumors make up less than 1% of all pediatric brain neoplasms.[5] In this case report, we describe a rare case of a 28-day old neonate with a congenital massive Xanthoastrocytoma occupying the left frontal lobe. Since the tumor was incidentally discovered during fetal ultrasonography before birth, we classify this case as congenital. Within the first two weeks following birth, the patient experienced focal seizures; at four weeks, elevated intracranial pressure symptoms appeared. Previous research has divided newborn brain tumors into three categories: congenital (manifest symptoms at birth), probably congenital (manifest symptoms within the first week), and possibly congenital (manifest symptoms within the first few months).[6]

Consideration of alternative differential diagnoses, including glioblastomas, atypical teratoid/rhabdoid tumors, pilocytic astrocytomas, and primitive neuroectodermal tumors, were prompted by the heterogeneous appearance with peripheral contrast enhancement.[7,8] However, in both pediatric and adult cases of Xanthoastrocytomas, the presence of a mural nodule within a cystic component has been described as a characteristic imaging finding.[9]

Neonatal brain tumor prognosis can vary greatly, depending on a multitude of factors that affect the overall clinical outcome. The patient's age, the tumor's size and characteristics, and the intraoperative variables that arise during surgical management are of the important determinants. With a large tumor, the patient in our case was a neonate, which presented its own unique set of challenges. However, with the coordinated efforts of the multidisciplinary team, which included a skilled neurosurgeon and anesthesiologists, complete surgical excision was achieved with a favorable postoperative outcome.

Other studies had to perform subtotal excision because of extremely high tumor vascularity and massive intraoperative bleeding, which had a negative effect on the patient's outcome and made the recovery process challenging for the anesthesiologist and the infant. The infant also suffered from complications related to intraoperative bleeding, prolonged anesthesia, and blood transfusions; sadly, their young patient passed away 48 hours later.[10] Better results have been reported, though, when small tumors are fully removed. This is especially true for Choroid plexus tumors, where aggressive multimodal treatment can result in disease-free survival.[11]

Despite the histological analysis indicating a mitotic count of 2/10 high-power fields, which is below the threshold for malignant transformation, we opted for complete surgical resection of the tumor without adjuvant therapy. This decision was supported by studies suggesting that aggressive treatment, including both radiotherapy and chemotherapy, may not be necessary for Xanthoastrocytomas.[12] Normal development and growth were noted in our case during a two-year period of long-term follow-up.

CONCLUSION

Neonatal brain tumors, particularly those with a massive large size like the one described in this case, are extremely rare. Our experience indicates that a successful outcome from gross total surgical resection can be obtained without the need for adjuvant therapy, but it does require the coordinated efforts of a multidisciplinary team and the assistance of anesthesia.

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List of Abbreviations

EMA: Epithelial membrane antigen.
ERG: Erythroblast transformation specific related gene.
INI-1: Integrase interactor 1.
Ki-67: Protein marker for cellular proliferation.
MRI: Magnetic resonance imaging.
OLIG2: Oligodendrocyte lineages transcription factor 2.

Disclosure

The authors report no conflict of interest in the materials or methods used in this study or the findings specified in this paper.

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