Spontaneous Regression of Congenital Posterior Fossa Lesion – Radiologically Mimicking Teratoma – Case Report

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Abstract: We report a case of spontaneous regression of a congenital intracranial lesion presumed to be a teratoma. Various radiological features of teratoma mentioned in the literature are vague complex echogenic mass on prenatal ultrasonography, clump like calcification on CT scan, usual size being more than 5cm, suprasellar and pineal region location. Radiological features of this lesion were consistent with teratoma. The child was managed conservatively with oral steroids as an antiedema measure. Follow up scans over 2years showed more than 80% regression in size of the lesion. This is the first reported case of presumed teratoma, on radiological imaging that regressed spontaneously.

Keywords: Calcification, regression, teratoma.

INTRODUCTION

Most of the neoplasms detected at birth or during the first month of life have their beginning in the intrauterine period. It comprises of 2.5% of all tumors in the pediatric age group [1]. Teratomas are the commonest tumors in this group [2,3]. They are usually sporadic with some cases diagnosed antenatally. The outcomes of such lesions are generally poor, though surgical resection, if possible may give long term survival benefit [4]. However there is no case report so far, mentioning spontaneous regression of such congenital intracranial lesions. We report a case where spontaneous regression of congenital intracranial lesion radiologically resembling a teratoma occurred.

CASE REPORT

A one-month old male child presented to us with the complaint of large head-size and an episode of tonic posturing fifteen days before. Prenatal Ultrasound done at 36 weeks of gestation revealed large fetal head with dilated ventricular system. Anticipating cephalo-pelvic disproportion an elective Caesarean section was done. Apgar score was normal at birth. The birth weight was 2.8 kg and his head circumference was 40 cm at birth (macrocephaly for his age). Child had normal sucking reflex. There was no history of poor feeding, irritability, excessive crying, recurrent vomiting, failure to thrive or excessive sleep. On examination, his head circumference was 42 cm (which was greater than 2SD expected for his age), and anterior fontanelle was open and full. His general physical examination was unremarkable. Child was alert, playful and had good cry with no discernible neurological deficit.

Retrospective evaluation of prenatal ultrasonography images done at 36 weeks of gestational age (Fig. 1A) showed a complex echogenic mass with hydrocephalus.

CT scan of brain at the age of 1 month (Fig.1B) was done at the referred hospital which showed a large heterogeneous predominantly hypo-dense lesion (4.6 cm x 5.6 cm x 5.7 cm) with chunks of calcification located in posterior fossa. It was extending from pineal region to occipital convexity. There

Fig. (1). A: Ultrasonography at 36 weeks of gestation showing large fetal head with dilated ventricular system (white arrow). B: Contrast enhanced CT scan brain showing large heterogeneous density lesion with clump like areas of calcification. Hydrocephalus is evident with gross ventriculomegaly.

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Spontaneous Regression of Congenital Posterior Fossa Lesion

We report a unique case of a lesion which radiologically seems to be a teratoma and showed features of regression on serial neuroimaging. Congenital intracranial tumors are rare and only account for 0.5% to 1.5% of all childhood brain tumors [5-7]. Various intracranial tumors presenting in neonates reported in literature in order of frequency are as follows - Glial tumor (includes astrocytoma, glioblastoma, choroid plexus papilloma and ependymoma), teratomas, PNETs and rarely, medulloblastoma, gangliogioma, meningeal neoplasms [8]. Among the tumors present at birth, teratomas account for 28.8% to 50% of central nervous system (CNS) tumors [9]. They are usually detected during prenatal period as complex echogenic masses as compared to hypoechoic medulloblastoma and hyperechoic angioblastic meningioma [8]. On CT scan, teratomas usually show clump like calcification [8], and measure more than 5 cm in diameter [5]. The radiological feature of this lesion thus favored a teratoma, owing to its presence in the prenatal ultrasonography, clumped calcification as well as its size, and location at pineal region. Another feature that favors teratoma is its extra axial location unlike – medulloblastoma, PNET, choroid plexus papilloma. Most teratomas are midline tumors located predominantly in the sellar and pineal regions [10]. There are case reports describing teratoma occurring at other locations e.g. lateral ventricle [11]. The prognosis of congenital intracranial teratomas have been poor as these lesions are extensive when they are identified.

Spontaneous regression of malignant tumors have been seen generally in one of 60,000 to 100,000 patients [12]. There have been sporadic reports of this phenomenon of intracranial tumors identified radiologically, and this list includes intracranial metastasis of renal cell carcinoma [13], malignant lymphoma [14,15] and glioma associated with neurofibromatosis [16,17].

The possible mechanisms that have been suggested for this phenomenon include an immunological (cell mediated and humoral) mechanism, bacterial or viral infection, apoptosis, and surgical or other trauma [18,19]. Of these mechanisms, apoptosis is thought to play a role in most spontaneous regressions involving neoplasms, and an apoptosis-triggered immunological response is thought to be the main effector mechanism [20]. Corticosteroid treatment may change the patient’s total immunological defense mechanism, which then is sufficient to eradicate the intracranial tumor [13,21]. In our case too patient had received a tapering dose of steroids. Spontaneous regression is known to occur in patients with seminomas [22], however there wasn’t any report showing regression of congenital teratomas spontaneously. Histological confirmation would have been desirable to confirm the radiological diagnosis.

CONCLUSION

This case report highlights a very aggressive congenital lesion detected on imaging which mimicked a teratoma radiologically. However, histological confirmation would have been desirable to confirm the precise nature of the lesion.

CONFLICT OF INTEREST

The authors confirm that this article content has no conflicts of interest.

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REFERENCES


