J. Fetal Med. (June 2015) 2:91–95
DOI 10.1007/s40556-015-0046-y

BRIEF COMMUNICATION

A Rare Aggressive Fetal Intracranial Tumor

Ravi Kapoor[1] · Ashutosh Bansal[1] · Aakriti Kapoor Aggarwal[2] · Abhinav Aggarwal[2,4] · Vandana Chaddha[3] · Sunita Kapoor[1]

Received: 3 March 2015 / Accepted: 15 May 2015 / Published online: 2 September 2015

Abstract Congenital brain tumors, especially tumors diagnosed before birth, are very rare. This article describes a case of primitive neuroectodermal tumor (PNET) diagnosed by ultrasonography at 24 weeks of gestation. Follow-up ultrasound study and MRI at 27 weeks of gestation revealed aggressive growth and complications of tumor. The diagnosis was confirmed at autopsy. Prenatal diagnosis of PNET or other congenital brain tumors is important because the presence of tumors may alter the time and mode of delivery and planning of postnatal care.

Keywords Congenital cerebral primitive neuroectodermal tumor · Fetal intracranial tumor · PNET · Fetal MRI · Prenatal ultrasonography

Introduction

Ultrasonography is the modality of choice for initial and routine examinations of fetal brain. Due to excellent soft-tissue resolution and with its safety to fetus, magnetic resonance imaging (MRI) constitutes a useful complementary, and sometimes, problem-solving tool where ultrasound is inconclusive. Brain tumors presenting in first two months of life are often described as being congenital [1]. Solitare and Krigman and later Wakai et al. classified congenital brain tumors into three categories according to period when symptoms appear after birth: definitely congenital tumors (at birth), probably congenital tumors (within first week) and possibly congenital tumors (within first two months after birth) [2, 3]. Recently, brain tumors in fetuses have been diagnosed with ultrasound and other means and have been classified as fetal brain tumors (definitely congenital tumors) [4]. Fetal brain tumors are different from other age groups with respect to localization, biological behavior, response to treatment, and histological type. These tumors can also cause spontaneous intracranial hemorrhage in utero or dystocia during pregnancy, preterm delivery, and stillbirth [5].

Primitive neuroectodermal tumor (PNET) constitutes approximately 13 % of all fetal and neonatal tumors [6]. Most patients with PNET have minor symptoms and majority are diagnosed late. Prenatal diagnosis of PNET or other congenital brain tumors is important because the presence of tumors may alter the time and mode of delivery and postnatal care planning [6, 7]. This article describes the prenatal ultrasound of a PNET at 24 weeks of gestation with follow-up ultrasound and MRI at 27 weeks of gestation revealing aggressive growth and complications of tumor with diagnosis confirmed at autopsy.

Report of Case

A 25-year-old woman, gravida 2, para 1, was referred to our clinic for routine obstetrical antenatal evaluation at approximately 24–25 weeks of gestation. She had no significant medical history and had not taken any medication during pregnancy. Ultrasound revealed an echogenic mass in the supratentorial location involving the left

✉ Abhinav Aggarwal
abhinavagg@gmail.com

1 City X-Ray & Scan Clinic, Vikas Puri, New Delhi, India

2 Shri Ram Murti Smarak Institute of Medical Sciences, Bareilly, Uttar Pradesh, India

3 Lajpat Nagar, New Delhi, India

4 A-1/237, First Floor, Janak Puri, New Delhi 110058, India

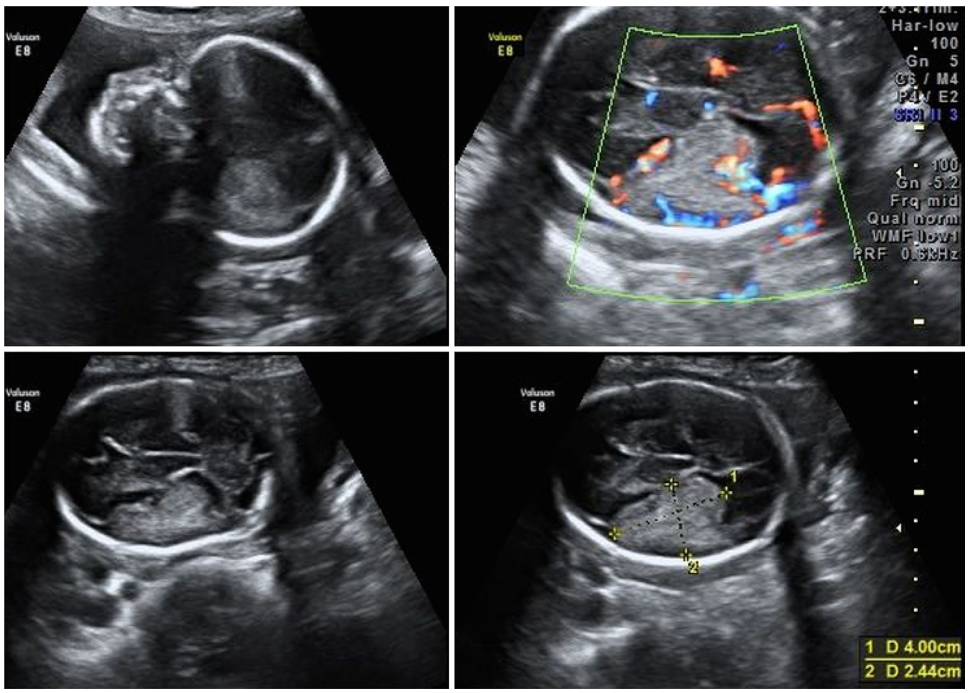

Fig. 1 Echogenic mass in left supratentorial region with intralesional and perilesional vascularity

thalamic and temporal region measuring approximately 40 × 24 mm. The lesion was minimally irregular in margins and on color and power Doppler imaging revealed intralesional vascularity (Fig. 1). No other significant finding was seen at the time of scan. Possibility of intracranial space occupying lesion was given, and patient was also advised fetal MRI examination for further evaluation to define the extent of lesion and rule out posterior fossa involvement, if any. The patient refused MRI examination as well as any further treatment at initial request citing cultural and familial reasons. The patient was then advised a follow-up scan after 2–3 weeks. A repeat ultrasound was done after three weeks which revealed an enlarged-for-gestational-age fetal head (BPD = 8.7 cm) corresponding to 35 weeks of gestational age with a large heterogenous mass involving the left cerebral hemisphere measuring approx 9.1 × 6.5 cm and showing multiple irregular cystic areas within the mass with increased intralesional and perilesional vascularity (Fig. 2a, b). Falx was displaced to the right side with enlarged right lateral ventricle seen. Associated polyhydramnios with placentomegaly, pericardial effusion and pleural effusion in fetus were noted (Fig. 3a–d). On explanation of possible aggressive malignant intracranial lesion, the patient agreed for further evaluation and a fetal MRI was done which revealed a large lesion involving the left hemisphere with the right-sided hydrocephalus. The lesion displaced the falx to right side with attenuation of right cerebral cortex (Fig. 4). There was no infratentorial extension of the tumor. However, the mass was seen to compress and displace the cerebellar hemispheres inferiorly (Fig. 5).

The patient underwent spontaneous preterm labor within the next few days. Autopsy and histopathological examination of lesion revealed an intracranial tumor with proliferation of neoplastic cells which are small plump ovoid arranged in a perivascular arrangement with focal pseudorosetting having fine granular chromatin and inconspicuous nucleoli. Mitotic activity appeared scanty with focal necrosis suggestive of PNET (Fig. 6a, b, c).

Discussion

Congenital brain tumors represent only 0.5 %–1.9 % of all pediatric brain tumors [8]. Isaacs in his review of 250 perinatal intracranial tumors reported the incidence of PNET to be around 13.2 % [6]. Rickert et al. reported that PNET was the most common tumor in first year of life, astrocytoma and ependymoma were highest in first two years of life and astrocytomas were highest in first four years of life accounting for one-third of all tumors in their epidemiological survey [9].

Congenital PNETs are rare in fetal and perinatal period. Most arise infratentorially from cerebellar mid-line and if large enough, extend into supratentorial compartment [7]. A diagnosis of PNET in a fetus is also important due to seldom documented familial occurrences [10]. The present case is unusual due to supratentorial location and its aggressive behavior. The tumor was initially seen as an

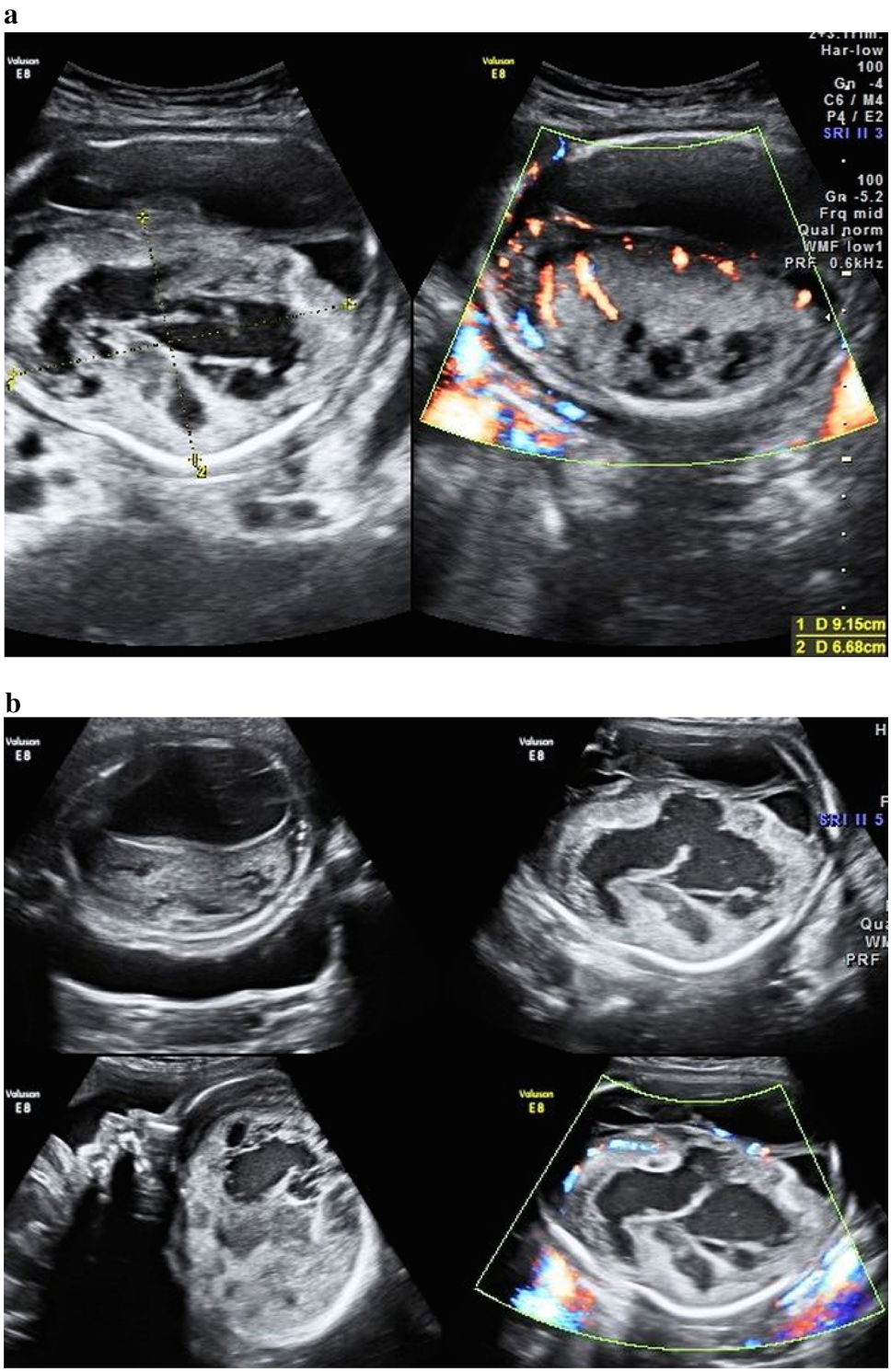

Fig. 2 **a** Large mass at 27–28 weeks involving left cerebral hemisphere and right hydrocephalus. **b** Large heterogenous mass in left cerebral hemisphere with cystic areas and increased intralesional and perilesional color flow

echogenic mass with intralesionals vascularity on initial examination. On subsequent examination after three weeks, the lesion was seen to occupy most of the left cerebral hemisphere with hydrocephalus. Associated placentomegaly, pericardial effusion, pleural effusion, and polyhydramnios suggest high fetal cardiac stress due to aggressively growing tumor. This finding seems to contrast with that of teratomas where normal brain tissue is replaced by tumor and normal brain structures are not clearly identifiable. MRI findings complemented the ultrasound findings and also helped in better visualization of the compressed cerebral mantle. Also, the heterogeneous

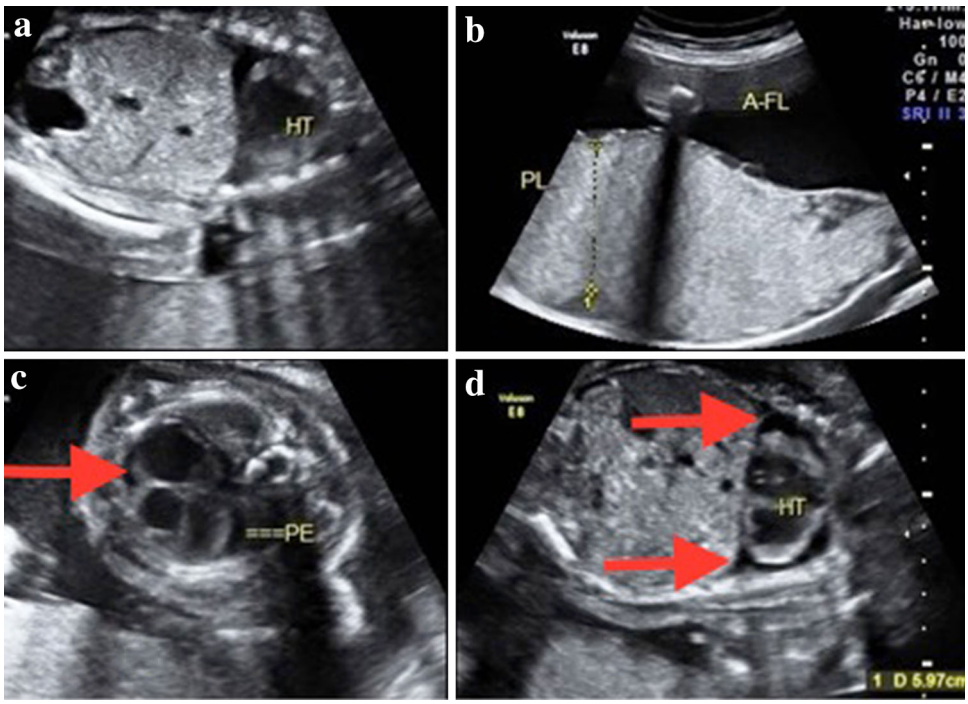

Fig. 3 27–28 weeks scan. Cardiomegaly (**a**, **c** & **d**; marked by *red arrow* in **c**); placentomegaly (**b**); pericardial (**c**); and bilateral pleural effusion (**d**; marked by two *red arrows*)

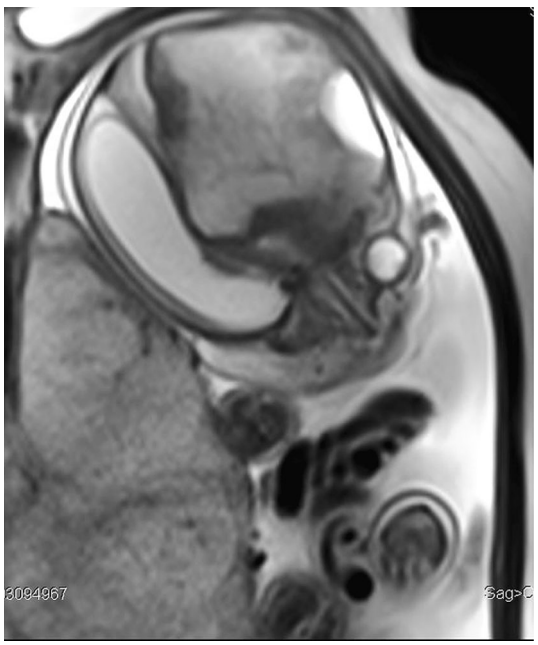

Fig. 4 Fetal MRI. A large heterogenous lesion involving most of left cerebral hemisphere with right-sided hydrocephalus and attenuation of right cerebral cortex

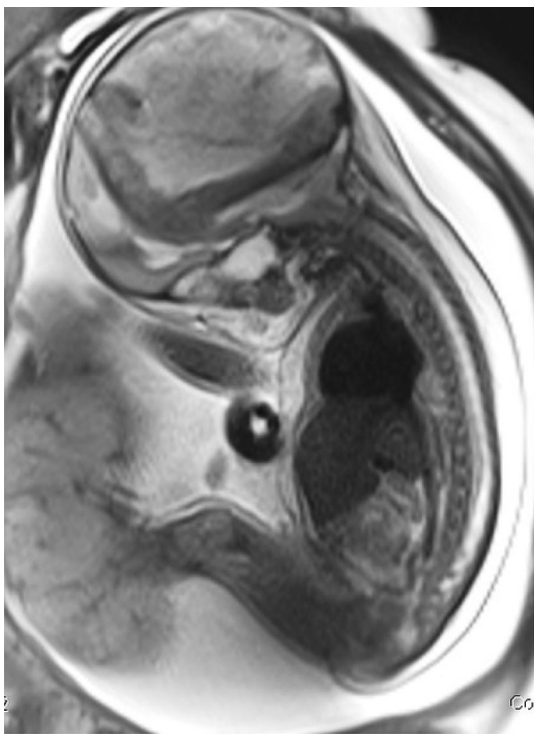

Fig. 5 Fetal MRI. Supratentorial location of tumor with compression of tentorium and posterior fossa structures inferiorly

signal intensity of the lesion suggested intratumoral bleed. MRI is useful for detection of lesions questionable on ultrasound such as ischemic and hemorrhagic lesions. In the present case, prenatal MRI helped in better tumoral localization, as well as evaluation of tumor margin and intratumoral bleeding. MRI was also helpful in parental counseling as it provided an image of fetus which was more visually comprehensible to the patient and family.

Management of the case is dependent on the gestational age and tumoral extent at time of diagnosis. Most of them

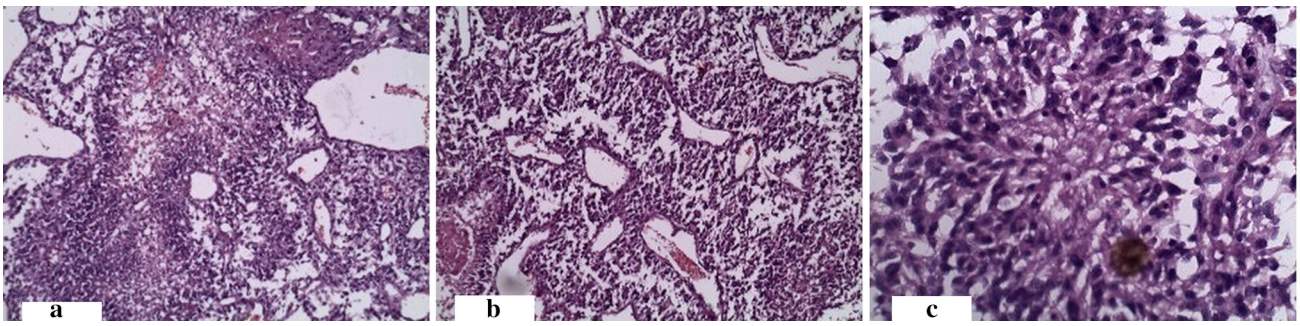

Fig. 6 Histopathological view. **a** Pseudorosseting, **b** Perivascular arrangement, **c** Plump ovoid neoplastic cells [High power (40x)]

are commonly diagnosed at advanced stage and the tumor may sometimes grow excessively where cesarean section may have to be done due to enlarged head leading to cephalopelvic disproportion [7]. In the present case, termination of pregnancy was considered, because the diagnosis was made at early stage with aggressive nature of tumor established on subsequent follow-up which is associated with a very poor outcome and postnatal prognosis.

To the best of our knowledge, there are very few cases of PNET diagnosed prenatally [7, 11, 12]. Our case is also one of the earliest diagnosed cases (24–25 weeks) of PNET with a documented aggressive growth shown on subsequent examination. Although rare, PNET is a highly aggressive tumor. Progressive hydrocephalus and enlarged for gestational age fetal head require meticulous ultrasound and if needed MR evaluation to rule out intracranial tumor.

Complaince with Ethical Standards

Conflict of Interest None.

References

1. Arnstein LH, Boldrey E, Naffziger HC. A case report and survey of brain tumours during neonatal period. J Neurosurg. 1951;8:315–9.
2. Solitaire GB, Krigman MR. Congenital intracranial neoplasm. J Neuropathol Exp Neurol. 1964;2:280–92.
3. Wakai S, Arai T, Nagai M. Congenital brain tumors. Surg Neurol. 1984;21:597–609.
4. Nozaki M, Ohnishi A, Fujimaki T, et al. Congenital germistocytic Astrocytoma in a fetus. Childs Nerv Syst. 2006;22:168–71.
5. Inwald D, Kempley S, Hird M. Congenital primitive neuroectodermal tumor presenting as obstructed labour. Arch Dis Child Fetal Neonatal Ed. 1998;78:222–4.
6. Isaacs H. Perinatal brain tumors-a review of 250 cases. Pediatr Neurol. 2002;27(4):249–61.
7. Yamada T, Takeuchi K, Masuda Y, Kitazawa S, Maruo T. Prenatal imaging of congenital cerebral primitive neuroectodermal tumour. Fetal Diagn Ther. 2003;18(3):137–9.
8. Buetow PC, Smirniotopoulos JG. Congenital brain tumors: a review of 45 cases. Am J Roentgenol. 1990;155(3):587–93.
9. Rickert CH, Probst-Cousin S, Gullota F. Primary Intracranial tumours of infancy and early childhood. Childs Nerv Syst. 1997;13:507–13.
10. Yamashita J, Handa H, Toyoma M. Medulloblastoma in two brothers. Surg Neurol. 1975;4(2):225–7.
11. Koksal Y, Varan A, Akalan N, Bostanci A, Clia A, Söylemezoglu F, et al. Congenital cerebellar primitive neuroectodermal tumor in a newborn. Am J Perinatol. 2006;23(3):173–6.
12. Sahin FK, Koken G, Cosar E, Koken R, Sahin O, Gokden M. A prenatal diagnosed case of primitive neuroectodermal tumour. Fetal Diagn Ther. 2008;23(4):267–70.