Clinical Brief

Large Choroid Plexus Teratoma : A Rare Cause of Congenital Hydrocephalus

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ABSTRACT

Teratomas form the most common type of congenital brain tumors, frequently presenting as stillbirth. The largest neonatal series of intracranial teratomas reported a 12% survival rate. Although the first teratoma of the lateral ventricle was reported in 1961 by Maier, neonatal intracranial teratoma of the lateral ventricle is an extremely rare entity. We report here a large intracranial poorly differentiated teratoma arising from choroid plexus of lateral ventricle. This typically presented at birth with a large congenital hydrocephalus. **[Indian J Pediatr 2010; 77 (4) : 452-453]** *E-mail: drashishjain2000@yahoo.co.in*

Key words : *Hydrocephalus; Choroid plexus; Teratoma*

REPORT OF CASE

A male weighing 4.6 kg was born by cesarean section, he needed positive pressure ventilation for 30 seconds and Apgars of 6, 9 and 9 were assigned at 1, 5, and 10 minutes respectively. The baby (Fig. 1) was transferred to nursery in view of the large head and respiratory distress. At 20 hours, baby had generalised tonic clonic seizures, and was loaded with phenobarbitone. Subsequently CSF analysis revealed protein-500 mg/dl, sugar-24 mg/dl, RBCs- 100/ HPF, 35 cells, with 60% polymorphs. Sepsis screen and blood/ CSF cultures were non-suggestive. On third day cranial sonogram revealed a choroid plexus mass 11×7 cm in both lateral ventricles compressing on third ventricle. MRI (Fig. 2) highlighted a huge choroid plexus mass arising from lateral ventricles. Neurosurgery consultation opined that in view of the grave prognosis baby may be managed on supportive lines. On day 10th baby had poor respiratory efforts followed by cardiac arrest. A trans-cranial biopsy was taken after consent and sent for histopathological examination. The biopsy was suggestive of poorly differentiated teratoma.

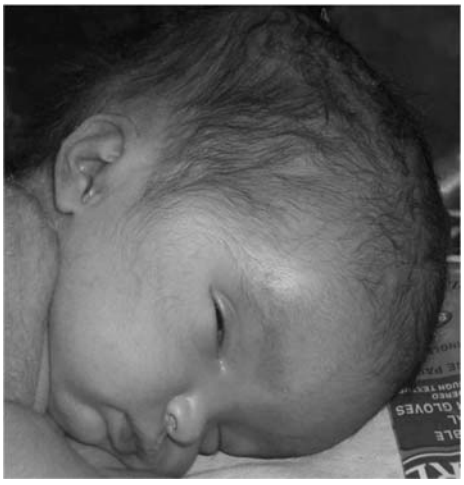

Fig. 1. Newborn baby with large head.

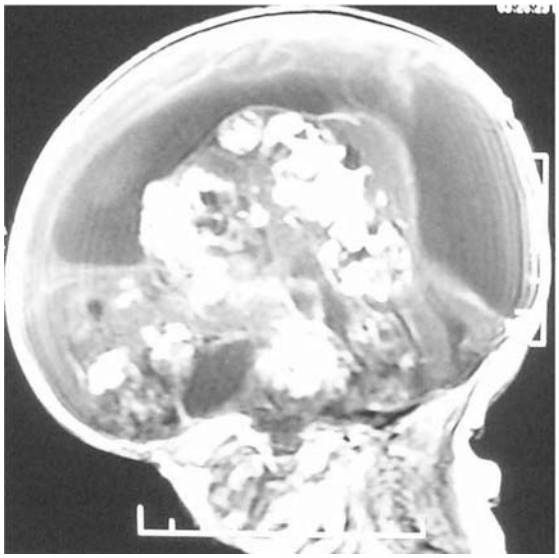

Fig. 2. Huge tumor arising from choroid plexus of lateral ventricle.

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DISCUSSION

Congenital brain tumors are present in the first 60 days of life. They include primitive neuroectodermal tumors (PNET), teratomas, astrocytomas, choroid plexus papillomas, and gangliogliomas.[2,3] Typical presentations include polyhydramnios, macrocephaly, dystocia, and stillbirth. The incidence of congenital brain tumors is 0.34 per million live births, higher incidences have been reported by Takaku *et al*.[6] Most of these tumors are supratentorial, in contrast to the infratentorial location in older children. The teratomas constitute between one third and one half of this number.[2,3,7]

Intracranial teratomas are histopathologically categorized as mature, immature, or malignant. All extragonadal teratomas are presumed to arise from misplaced primordial germ cells. Teratomas are typically cystic, well defined, and have all three germinal layers. They have no necrosis and low mitotic activity. Breslau(1864)[8] reported the first case of massive congenital intracranial teratoma. Intracranial teratomas may arise from the pineal gland, quadrigeminal plate, third ventricle, suprasellar region or cerebellar vermis. Their location in the lateral ventricles, as in the present case, is rare and probably relates to the choroid plexus.

Oiset *et al* classified tumors as 'definitely congenital' (within 1 week) as in our case, 'probably congenital' or 'possibly congenital'(within first month of life) depending upon onset of symptoms.[9] Congenital intracranial teratomas may be:[10] (i) massive teratomas, (ii) small teratoma producing hydrocephalus, (iii) teratomas extending into the orbit or neck.

These tumors can be diagnosed prenatally by sonology as early as 20 weeks. Congenital teratomas usually present clinically with macrocephaly. Hydrocephalus may be an early presentation because of obstruction of the CSF pathway. Cesarean sections are common as in our case due to the cephalopelvic disproportion, dystocia and extremely difficult delivery.

Various elements of teratomas can be identified by postnatal MRI (cystic and fatty components). The management is aimed at CSF diversions and subtotal or complete excision. Teratomas have an inherent tendency to grow; therefore, complete surgical excision should be attempted.[5] The first year mortality of excised tumors is 20%. Complete excision is often difficult where it has replaced large portions of the telencephalon.[4] Chemotherapy may help delaying the re-growth of these tumors and decrease the detrimental effects of radiotherapy. However, with improved methods of diagnosis and management, selected cases may be treated with more success in the coming future. Despite all efforts, the 5 year survival was 18% for malignant teratomas in the series reported by Ferreira. Most neonates, who have survived operation, have developmental delay and neuropsychological changes.

CONCLUSION

Intracranial teratoma arising from lateral ventricle and choroid plexus is a rare entity, but should be considered in cases of congenital hydrocephalus. These neonates are usually still-born or have a poor prognosis. A good outcome is rarely possible under optimal conditions, after resection in these complex, giant tumors. A multidisciplinary and staged surgical approach is usually essential for the successful outcome in such cases.

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REFERENCES

1. DiGiovanni LM, Sheikh Z. Prenatal diagnosis, clinical significance and management of fetal intracranial teratoma: a case report and literature review. *Am J Perinatol* 1994; 11: 420–422.
2. Isaacs H Jr I. Perinatal brain tumours: a review of 250 cases. *Pediatr Neurol* 2002; 27: 249–261.
3. Isaacs H Jr: II. Perinatal brain tumours: a review of 250 cases. *Pediatr Neurol* 2002; 27:333–342.
4. Whittle IR, Simpson DA. Surgical treatment of neonatal intracranial teratoma. *Surg Neurol* 1981; 15: 268-273.
5. Herder S, Ventureyra EC. Neonatal intracranial teratoma. *J Neurosurg* 1983; 59: 879-883.
6. Takaku A, Kadoma N, Ohara H, Hori S. Brain tumours in newborn babies. *Child's Brain* 1978; 4: 365-375.
7. Buetow PC, Smirniotopoulos JG, Done S.Congenital brain tumours: a review of 45 cases. *Am J Roentgenol* 1990; 155: 587-593.
8. Breslau, Rindfleisch E. Geburtgeschichte and Untersuchung eines. Falles von Foetus in Foetu. *Virchows Arch Path Anat* 1864; 30: 406-417.
9. Oi S, Kokunai T, Matsumoto S. Congenital brain tumors in Japan (ISPN cooperative study). Specific clinical features in neonates. *Child Nerv Syst* 1990; 6: 86-91.
10. Nanda Anil, Luis Schut, Leslie N Sutton. Congenital forms of intracranial teratoma. *Child's Nerv Syst* 1991; 7: 112-114.