EUROPEAN JOURNAL OF BIOMEDICAL AND PHARMACEUTICAL SCIENCES
http://www.ejbps.com

ISSN 2349-8870
Volume: 6
Issue: 12
287-288
Year: 2019

DYSEMBRYOPLASTIC NEUROEPITHELIAL TUMOR

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Article Received on 25/08/2019 **Article Revised on 14/09/2019** **Article Accepted on 04/10/2019**

ABSTRACT
Dysembryoplastic neuroepithelial tumors (DNTs) are benign lesions affecting pediatric population and are associated with epilepsy. The tumors histologically resemble gliomas but behave as stable lesions. They are associated with medically intractable epilepsy and a favorable prognosis after surgical resection. We present to you a case of a 4 year old Asian male child, diagnosed with Dysembryoplastic neuroepithelial tumor and treated surgically for the same.

KEYWORDS: Epilepsy, Congenital brain tumor, Generalised tonic clonic seizures.

INTRODUCTION
Dysembryoplastic neuroepithelial tumors (DNT) are classified as mixed neuronal-glial tumors.[1] They are rare tumors characterized by a supratentorial cortical location preferentially involving, in decreasing order, the temporal, frontal, parietal and occipital lobes.[2–5] Besides their cortical location, they can also involve extracortical areas such as the caudate nucleus, lateral ventricle, septum pellucidum and fornix, which suggests a putative origin from the secondary germinal matrix.[6–8] They usually occur in children and young adults with a long-standing history of drug-resistant complex partial seizures.[2,4,5,9] These slow-growing tumors usually show an indolent course with a favorable prognosis.[4, 7, 9–11] They are the most common curable cause of complex partial seizures and epilepsy surgery usually results in complete recovery from seizures.[7, 9–11]

CASE REPORT
A 4 year old Asian male child presented with episodes of generalized tonic clonic seizure with 1 and half month duration not controlled by medicines started by primary doctor. He had no history of trauma to head or fever prior to this. Parents reported that he had normal birth but slightly delayed milestones and was noted to have aggressive nature with easy irritability, anger tantrums, tendency to throw objects, tendency to hit others and tendency to even hurt self in anger. He needed extra and special care. He was first of two siblings and the other child was normal. On examination, he revealed normal vital signs but he was extremely aggressive and uncooperative. He was violent at times and was constantly crying. He was speaking a jargon with improperly developed speech. He was having good motor function with apparently no major motor deficits or cranial nerve deficits. He was able to recognize parents and strangers but was apparently mentally retarded with no social, cognitive and adaptive milestones as per age. Detailed examination was not possible due to extreme un-cooperation and violent attitude but other systems apparently were normal. The child was investigated with a plain and gadolinium enhanced MRI scan, and the scan revealed a right mesialtemporal parenchymal lesion of 4x3x3 cm size, which was inhomogenous, mildly hyperintense on T2 weighted image and mildly hypointense on T1 weighted image. There was no perifocal edema and no intra lesional hemorrhage. The lesion was inhomogenously enhancing on contrast administration. The MRI morphology was suggestive of DNT. The other less likely differential diagnosis were ganglioglioma and low grade glioma. He was put on preoperative steroids, antiepliptics and antibiotic prophylaxis. His Hb was 12.6 gm%, total count was 11,500/mm^3, RBS was 94.6 mg/dl, Na 138.6 mmol/l, K 4.8 mmol/l and serum creatininine 0.82. He tested negative for HIV, HBsAg, HCV. The child was subjected to a brain tumor surgery by performing right temporal craniotomy and microsurgical near-total excision of the tumor. Child tolerated the surgery well and was intensely monitored postoperatively for any complications. Postoperative period was uneventful and the child recovered to near normal neurological status with minimal left hemiparesis. The stitches were removed in time and the child was discharged with advice to continue antiepileptic medicines and physiotherapy. The biopsy report of the tumor was WHO Grade I DNET, right temporal tumor.

DISCUSSION

Dysembryoplastic neuroepithelial tumors DNTs are highly distinct tumors that arise during embryogenesis. They are preferentially, but not exclusively, located in the supratentorial cortex. Histologically, they may mimic any categories of low-grade or even of high-grade gliomas, but from a carcinological point of view, they behave as stable lesions. Their differential diagnosis from gliomas, is obviously important to spare these young patients with a normal life expectancy the long-term deleterious effect of radiation or chemotherapy. The diagnosis of DNT must be considered when all the following criteria are present: partial seizures with or without secondary generalization, no neurological deficit or a stable congenital deficit, cortical topography on MRI, absence of peri-tumoral edema and of mass effect. In other locations, the diagnosis of DNT has to be suspected in case of discordance between the neurological status of the patient and the topography of the tumor or of bizarre radiological features such as contrast enhancement but no mass effect and no edema. Supratentorial cortical DNTs tend now to be identified more methodically by imaging soon after first seizures. In most instances, epilepsy can be cured by gross total surgical removal. Surgery also allows to prevent the risks of intratumoral hematoma or infarct. DNTs should therefore be operated soon after diagnosis. However, exemplary results can also be obtained by epilepsy surgery in patients with long term drug resistant partial seizures.

CONCLUSION

Ablation of DNETs and, when present, adjoining dysplastic cortex was highly effective for seizure control. Exquisite seizure-free outcomes and tumor control were seen with lesionectomy. Electrocorticography with extended resection is useful for patients with pharmacoresistant epilepsy.

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