Desmoplastic supratentorial neuroepithelial tumors of infancy form a rare but distinctive group of tumors occurring in children less than 2 years of age. Vanden Berg et al (1987), named these tumors as desmoplastic cerebral astrocytoma of infancy and desmoplastic infantile ganglioglioma (DIG), on the basis of immunohistochemistry and electron microscopy. Both these groups of tumors occur in children less than 2 years of age. They have identical clinical and radiological features and both have favorable outcome following successful excision. Subsequently, these groups of tumors have been included as a distinctive group, in the revised World Health Organization (WHO) classification of brain tumors (Kleihues et al 1993).

Less than 50 cases of desmoplastic infantile ganglioglioma (DIG) have been reported in the literature. Several authors have reported single case reports and small series of cases of DIG.4,5 The youngest patient has been 4 months of age. All these children presented with macrocephaly, seizures, psychomotor delay, either in isolation or in combination. All these patients have similar Computerized Tomographic scan (CT) and Magnetic Resonance Imaging (MRI) characteristics (Martin et al 1991, Tenreiro-Picon et al 1995). The tumors commonly occur in frontal or parietal lobes, with a solid component closely attached to the dura, which intensely enhances with contrast, surrounded by a parenchymal cystic component and mass effect. Most of these tumors are amenable to surgical excision because of their surface location and distinct firmness with clear demarcation from the surrounding normal brain. No adjuvant therapy is recommended after total excision, despite the fact that the lesion has a low malignant potential. Long-term prognosis after total surgical excision has been excellent in all the reported series.

Histologically, these tumors have intense desmoplasia, with neoplastic astrocytes with occasional neuronal elements. Desmoplastic cerebral astrocytoma of infancy and DIG are difficult to differentiate under light microscopy. Immunohistochemistry plays a very important role in the diagnosis of DIG. In DIG, the tumor shows intense positivity for Glial Fibrillary Acidic Protein (GFAP) which is a glial marker, with areas positive for synaptophysin or neurofilament immunostain, which are neuronal markers. In desmoplastic cerebral astrocytoma, there is positivity for GFAP only.

References


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bitone, Primodone, Carbanazepine and Amitryptiline. Spector et al\(^2\) reported that phenytoin could induce total external ophthalmoplegia irrespective of oral or intravenous administration. Incomplete\(^1\) as well as complete ophthalmoplegia\(^2\) has been reported with phenytoin even within therapeutic range.

We present a patient with phenytoin intoxication who developed disturbances in the ocular movements.

**Case Report**

A 28-year-old mentally retarded male, with IQ-55, had generalized tonic-clonic epilepsy for two years. He was placed on carbamazepine (Tegretol) 800mg per day. However, due to financial reasons, he was irregular with his treatment. Subsequently, he was placed on Diphenyl hydantoin (300mg per day). After about two weeks of the change in the drug treatment he reported inability to move his eyes. He denied excessive ingestion of the drug. The pupils were 5mm in diameter, round, equal and reacting well to light. The gaze was fixed directly forward. The patient did not move his eyes on command and could not follow the light. The eyes could not be moved by head turning or neck bending or by irrigation of the ears with ice water. Fundi were normal. The rest of the neurological examination was normal. The cerebrospinal fluid examination was normal. The CT of the head was normal. The phenytoin drug level was 22 microgm per ml. The phenytoin was withdrawn and he was given loading dose of phenobarbitone, followed by its maintenance dose. On the fifth day, he could move his eyes about 10° in horizontal direction (laterally and medially) but was unable to move them in the vertical direction. On the seventh day, the horizontal movements had improved and he could move the eyes slightly downward but not upward. Irrigation of the left ear with ice cold water produced no movements of the eyes but irrigation of right ear produced occasional nystagmus with fast component to the left. On the eleventh day his eye movements were normal. He was seizure-free on 120 mgm of phenobarbitone per day.

**Discussion**

Phenytoin is a vestibular depressant.\(^6\) Spector et al\(^2\) observed that the return of vestibulo-ocular response in their patients with phenytoin intoxication lagged behind the return of consciousness and other reflex activities and attributed it to its depressant effect on the vestibulo-ocular motor system which may be out of proportion to its actions on other levels of the neuraxis. The oculomotor unresponsiveness to cold caloric irrigation may occur even when the blood phenytoin level is within the accepted therapeutic range.

GABA mediates the inhibition of oculomotor neurons produced by the vestibular system. Phenytoin increases the post-synaptic potentials produced by GABA in the cerebral cortex\(^6\) and the spinal cord. Spector et al\(^2\) attributed the unresponsive cold caloric irrigation with phenytoin to an increased effectiveness of GABA-induced inhibition at the synapses of the vestibulo-ocular motor system. The lag in the recovery of the oculo-vestibular response in the present case could be explained with the above said hypothesis.

The cerebellum has an inhibitory role over the vestibulo-ocular reflex through Purkinje cells. Phenytoin increases the rate of Purkinje cell discharge.\(^7\) Whether this phenytoin-augmented Purkinje cell firing would act in a manner analogous to electric stimulation of the cerebellar cortex and result in the depression of transmission through the vestibular nucleus and affect the normal function of the vestibulo-ocular apparatus is unclear. The majority of the reported cases of phenytoin-induced ophthalmoplegia recovered completely over a variable period with normalization of the phenytoin level.

The present case is unique; in spite of the drug toxicity the patient was alert and had bilateral external ophthalmoplegia with loss of oculo-cephalic and oculo-vestibular reflexes. The recognition of this entity is important to avoid unnecessary investigations in a patient on phenytoin.

**References**


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**Calvarial malignant fibrous histiocytoma**


Departments of Neurosurgery, *Pathology and **Plastic Surgery, PGIMER, Chandigarh, India.