bitone, Primodone, Carbamazepine and Amitriptyline. Spector et al² reported that phenytoin could induce total external ophthalmoplegia irrespective of oral or intravenous administration. Incomplete⁴ as well as complete ophthalmoplegia⁵ 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 microgram 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 The phenytoin was withdrawn and he was given loading dose of phenobarbitone, followed by its maintenance dose.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 then 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 al² 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 and the spinal cord. Spector et al² 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 oculovestibular response in the present case could be explained with the above said hypothesis.

The cerebellum has an inhibitory role over the vestibular-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 oculocephalic and oculovertical 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


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Primary malignant fibrous histiocytoma (MFH) of the central nervous system (CNS) is uncommon. We report cases of two young patients of MFH arising from the cranial meninges and involving the adjacent skull and scalp. There was infiltration of the brain in one case. Both the lesions were excised and primary scalp repair was performed.

**Key Words:** Malignant fibrous histiocytoma, scalp, skull

### Introduction

Malignant fibrous histiocytoma (MFH) is a common soft tissue tumor in adults occurring in the 5th to 7th decades of life and accounts for 10.5% to 21.6% of all soft tissue malignant neoplasms. The primary incidence involving the central nervous system (CNS) was described by Gonzalez-Vitale et al in 1976. Since then there have been only few sporadic case reports of MFH involving the CNS.

We report two young patients with malignant fibrous histiocytoma presenting as rapidly growing skull tumors.

**Case No. 1**

A 25-year-old female presented with a swelling over the left parieto-occipital area for the past five months. The swelling showed ulcerative changes two months prior to admission in hospital. There was intermittent bleeding and purulent discharge from the swelling. There was local pain and mild fever with occasional vomiting. There was no history of trauma.

On examination the patient was anemic and had no neurological deficit. There was a 15cm x 20cm globular mass in the left parieto-occipital region with area of ulceration and necrosis of the overlying scalp. The swelling was firm, non-tender, involving the scalp and the underlying bone. It had well-defined margins and was fixed to the bone. Skull roentgenogram revealed a soft tissue mass in the occipital area with area of ulceration and necrosis of the overlying scalp. The swelling was firm, non-tender, involving the scalp and the underlying bone. It had well-defined margins and was fixed to the bone. Skull roentgenogram revealed a soft tissue mass in the occipital area with bony destruction in the left parieto-occipital region (Figure 1). CT scan showed a mixed density mass with bony destruction. The heterogeneously enhancing mass showed involvement of the left occipital cortex (Figure 2). Fine needle aspiration cytology (FNAC) was suggestive of a giant cell tumor.

At surgery, the fungating scalp tumor was excised with a rim of healthy bone all around including the intracranial extension and along with the involved dura. The tumor was very vascular, and was seen infiltrating the left occipital lobe. Growth of the tumor into the superior sagittal sinus, left transverse sinus and torcular was seen at surgery. The tumor infiltrating into the occipital lobe was excised but the extension into the venous sinuses was not removed. The defect created by the dural excision was patched with fascia lata. Skin cover for a 10 x 15-cm scalp defect was provided by a rotation flap, based on the superficial temporal artery. The secondary defect in the scalp was covered by a split thickness skin graft. Cranioplasty was not performed due to the presence of areas of ulceration and infection. Microscopic examination showed the tumor to be poorly encapsulated. There was a prominent plump spindle cell stroma with focal storiform pattern. Marked nuclear pleomorphism and atypia with a few multinucleated giant cells were seen. Areas of necrosis were present. There were no lipoblasts or cells with striations. Inflammatory cells such as lymphocytes and neutrophils were present focally. The combination of pleomorphism and spindled stroma with focal storiform pattern suggested a diagnosis of malignant fibrous histiocytoma. Postoperative radiotherapy was advised but the patient refused further treatment and did not follow up.

**Figure 1:** Plain X-ray skull (Case no. 1) demonstrating a large lytic lesion in the parieto-occipital region.

**Figure 2:** Contrast CT Scan (Case No. 1) showing a heterogeneous enhancing scalp lesion with erosion of underlying bone and involvement of the occipital cortex.

**Case No. 2**

A 22-year-old male was admitted with history of a rapidly growing swelling in left temporal region for the past two...
months. On examination the swelling was situated in the left temporal region just above and anterior to the pinna. It was a 7x5-cm globular, firm and mildly tender mass, fixed to the underlying bone, with well-defined margins. A contrast CT scan demonstrated an enhancing mass in the left temporal region with underlying bone destruction (Figure 3). At surgery, the temporal bone was eroded and destroyed but the cortical surface of the brain was intact. The mass was excised along with the middle fossa dura, which was involved. Repair was done using a fascia lata graft. Postoperatively, the patient had a CSF leak, which was managed by a lumbar drain. Postoperative CT showed gross total excision (Figure 4). He was referred for radiotherapy. He had a local recurrence 1 month later and was advised chemotherapy but refused treatment. Histological examination showed a pleomorphic tumor composed of short fascicles of spindle cells and plumper histiocytic cells (Figures 5 & 6). Immunostaining for skeletal muscle actin was negative, ruling out a rhabdomyosarcoma. There were no lipoblasts. Other features similar to the previous case were present. No syncytial pattern or focal whirling was noted to suggest meningeal origin. Epithelial membrane antigen (EMA) was negative.

Discussion

Malignant fibrous histiocytoma, in extracranial locations, is the most common soft tissue sarcoma in adults,\(^8,16\) the most common sites being the retroperitoneum and the deep soft tissue of the extremities.

Primary MFH of the CNS is uncommon. In a review in 1998, Akimoto et al reported one case and described 17 previously reported cases.\(^15\) In both our cases there was no evidence of a primary tumor elsewhere. MFH elsewhere with brain secondaries usually occurs in old age while primary brain MFH occurs in relatively younger patients.\(^15,16\) Cerebral MFH usually involves the parietal, frontal or temporal lobes, whereas in one of our patients the occipital lobe was involved. The lesion in this patient possibly had a meningeal origin with secondary involvement of the overlying bone and scalp and infiltration of the underlying brain.

Immunohistochemistry has provided evidence that these tumors are not derived from cells of monocytic or macrophage lineage but rather from fibroblasts.\(^17\) In case no. 2 we had performed an immunostain for skeletal muscle actin which was negative, excluding a rhabdomyosarcoma.

Plain roentgenograms in MFH involving bone show a poorly...
circumscribed lytic lesion with areas of calcification while tomograms reveal subtle permeative radiolucent changes extending beyond the poorly demarcated central area of radiolucency. Angiography shows an extensive neovascular response while isotope studies reveal increased uptake beyond the tumor margin. The differential diagnosis includes cystic gliomas, glialblastoma, meningioma, abscess, metastasis and tuberculosis. MRI findings of an intracranial MFH secondary to an extracranial primary lesion have been described.18 Here, a cystic lesion with a ring-shaped tumor rim of decreasing signal intensity in T2-weighted image was seen. MRI is superior to CT in demonstrating tumor extent and edema.

The treatment for MFH has been described as a combination of radical excision, radiotherapy and chemotherapy. However, MFH of the brain has always been a gloomy prospect with a relentless course leading to death within the 1st year after surgery.3 Radical removal in case No.1 would have been extremely formidable due to the involvement of more than one venous sinus converging onto torcular Herophili. The hazards associated with interruption or diversion of venous sinus flow and reconstruction of the torcular Herophili in an attempt to achieve complete resection of a peritumoral malignant tumor seem unwarranted.19

References


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An intramedullary tumor presenting with hyperhidrosis

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A case of a cervical intramedullary tumor is reported whose presentation was with disabling hyperhidrosis. The symptom resolved after surgical debulking of the tumor. Hyperhidrosis as a presenting manifestation of an intramedullary tumor has not been reported earlier.

Key Words: intramedullary tumor, hyperhidrosis, cervical cord tumor

Introduction

We report an unusual case of hyperhidrosis in a middle-aged woman, as a presenting feature of an intramedullary cervical tumor: We could not locate any similar case in the literature.

Case Report

A 56-year-old lady presented with difficulty in using her hands as the initial complaint for a period of 6 months. This was followed by excessive sweating involving her head and neck area, so much so that she had to use 10-12 handkerchiefs daily to wipe herself. By the time she was seen at the Clinic, she had also started experiencing difficulty in walking “with a tendency to fall forwards” and had also developed urgency of micturition. However her main disabling symptom was hyperhidrosis.