Cerebral phaeohyphomycosis presenting as an intraventricular mass

Sir,

Phaeohyphomycoses are subcutaneous and systemic infections caused by dematiaceous fungi with dark walled hyphae due to the presence of melanin in the cell wall. Cerebral phaeohyphomycosis presents most commonly as a cerebral abscess and *Cladophialophora bantiana* accounts for the majority of the reported cases thus far.\(^1\)

A 20-year-old woman presented with headache, vomiting and progressive weakness of right upper and lower limbs for three months. Physical examination revealed bilateral papilloedema, right sided hemiparesis and spastic gait. Magnetic resonance imaging (MRI) of the brain showed an enhancing lesion in the left lateral ventricle causing dilatation of the ipsilateral lateral ventricle. The choroid plexus of the ipsilateral ventricle was thickened [Figure 1]. She underwent a left frontal craniotomy and subtotal excision of the mass. At surgery the mass was intraventricular and was attached to the choroid plexus and had papillary projections. Anteriorly the mass was adherent to the caudate nucleus and the thalamostriate vein. The histopathological examination showed a circumscribed nodule in the choroid plexus consisting of several discrete necrotising granulomata composed of multinucleated giant cells, histiocytes and lymphocytes. Pigmented branching hyphae were seen within the necrotic centers of the granulomata and within some of the giant cells [Figure 2]. The biopsy was reported as necrotising granulomatous inflammation consistent with phaeohyphomycosis. Species identification was not possible, as cultures had not been submitted. She was advised systemic antifungal treatment with amphotericin B, but was discharged at request on oral ketoconazole. Two weeks later she presented with symptoms of raised intracranial pressure. A computed tomogram of the brain showed an increase in the dilatation of the left lateral ventricle. She underwent an emergency left ventriculoperitoneal shunt. CSF fungal and mycobacterial cultures done at this point were negative. She was started on intravenous Amphotericin B and a cumulative dose of 1.5 gms was given. She was discharged on a six-month course of oral itraconazole.

Cerebral phaeohyphomycosis was first reported in 1952 by Binford and since then several cases have been reported. Apart from *Cladophialophora bantiana* other agents responsible are *Exserohilum* spp, *Exophiala jeaneslmai*, *Ramichloridium mackenziei*, *Fonsecaea pedrosoi*, *Ochroconis gallopavum*, *Chaetomium globosum* and *Bipolaris spicifera*.\(^{2-4}\) *Cladophialophora bantiana* is neurotropic in nature and may cause brain abscess in both normal and immunosuppressed patients. The portal of entry is not well established. The possible sources include trauma, iatrogenic, contiguous spread from paranasal sinuses and hematogenous dissemination. To the best of our knowledge there are no reports of an intraventricular mass lesion. Fontana-Mason stain for histologic identification is only indicated early on in the disease process as the hyphae may not appear pigmented leading to false identification and under reporting. Treatment must be individualized with surgical resection and optimal antifungal therapy. The most appropriate drug therapy is not clearly defined. Varied results are reported with systemic administration of miconazole, potassium iodide, ketoconazole, amphotericinB, flucytosine, fluconazole and itraconazole.\(^5\) Itraconazole is the preferred antifungal agent for phaeohyphomycosis. Relapses after therapy with other drugs, specifically ketoconazole and amphotericin B, have been treated successfully with oral itraconazole.

In conclusion this is a case of cerebral phaeohyphomycosis presenting at an unusual site. The feasibility of surgical resection should be considered in all patients with cerebral

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**Figure 1:** Magnetic resonance imaging (MRI) of the brain, T1W post gadolinium image showing an enhancing lesion in the left lateral ventricle causing dilatation of ipsilateral lateral ventricle.

**Figure 2:** Photomicrograph showing the hyphae within the giant cells of the granuloma (H/E, 90x)
Neurofibromatosis type I with occipital encephalocele

Sir,

Neurofibromatosis Type I is an autosomal dominant disorder associated with diverse intracranial and calvarial lesions. We present a case report of a patient with NF Type I with occipital encephalocele, an unusual association that has been described in only 1 case so far in the literature to the best of our knowledge.

A 28-year-old male patient with classical cutaneous stigmata of NF – I (neurofibromas and café-au-lait spots) with plexiform neurofibroma of the scalp presented with history of gradual increase in the size of his scalp swelling over the last three months. He was subjected to a pre and postcontrast cranial MRI to know the nature of the scalp swelling, after routine skull radiographs [Figures 1a and b] and CT [Figure 1 c].

MRI was suggestive of occipital encephalocele with plexiform neurofibroma of the scalp and neck [Figures 2 and 3]. Subsequent biopsy of the scalp lesion revealed neurofibrosarcoma on histopathologically.

NF-I is the commonest of all the phakomatoses (one in 2000 to 3000 live births), is usually inherited as autosomal dominant disorder. CNS manifestations associated with NF-I are gliomas (opticochiasmatic, hypothalamic, brainstem), non-neoplastic hamartomas of white matter and basal ganglia, macrocephaly, hydrocephalus, skull and meningeal dysplasias and plexiform neurofibromatosis, neurofibromas of spinal nerves.

None of the studies have reported occipital dysplasia in NF 1 except by Renshaw et al in 2003. These bony defects are because of progressive mesodermal dysplasia and rarely due to neurofibromatosis tissue in the scalp causing secondary erosion.

On the contrary, in congenital cephaloceles the bone defect is in occipital bone plus the posterior arches of adjacent cervical spine or in the occipital bone alone either superior or inferior to the external occipital protuberance. They can be associated with Chiari II and III malformation, Dandy Walker malformation, Cerebellar dysplasias, Diastematomyelia and Klippel-Feil Letter to Editor

Infection due to phaeohyphomycosis must be considered in fungal granulomatous inflammations.

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