Chronic mucormycosis manifesting as hydrocephalus

Sir: Fungal infections of the brain usually present as chronic meningitis. Mucormycosis of the brain, however, typically presents as acute rhinocerebral disease and is fatal. A search of the English literature revealed only three cases of chronic meninitis due to mucormycosis. All had rhinocerebral involvement and underlying metabolic disease. Fungal meninitis presenting primarily as hydrocephalus is rare. We here report such a patient. There were three uncommon features: (1) The primary presentation as hydrocephalus, (2) The presence of chronic basal meninitis without rhinocerebral involvement, (3) The occurrence in an immunocompetent host.

A 22-year-old female was admitted to hospital on 5 January, 1984 with complaints of convulsions, vomiting and headache of three months duration together with diplopia and unsteadiness for 15 days. She denied fever, cough, expectoration, pain in the neck or discharge from the ears. She was conscious and oriented. The pulse rate was 50/min, the BP 140/80 mm Hg and respiratory rate 30/min. There was bilateral papilloedema. The cranial nerves and the motor and sensory systems were normal. The tendon reflexes were all present and the plantar responses were extensor. Brudzinski and Kernig signs were negative. There was bilateral incoordination of the limbs, truncal ataxia and nystagmus. Romberg’s sign was negative. Examination of the abdomen and the cardiovasculard and respiratory system did not reveal any abnormality. The possibility of an intracranial space occupying lesion with raised intracranial pressure was entertained. The bilateral cerebellar deficit was considered to be a false localising sign.

Investigations revealed Hb 12 g%, total white count 6900/mm<sup>3</sup>, differential leucocyte count: polymorphs 50%, lymphocytes 42% and eosinophils 8% and ESR 77 mm in the first hour. Blood sugar was 108 mg/dl fasting and post prandial 120 mg/dl. The blood urea, creatinine level and liver function tests were within normal limits. The chest radiograph was normal but radiographs of the skull revealed osteoporosis of the posterior clinoid processes. A CT scan showed hydrocephalus.

Lumbar puncture was not performed because of papilloedema and the absence of signs of meningeal irritation. The patient was put on anticonvulsants and mannitol. After the report of the CT had been received, steroids and antituberculous therapy were started. The patient deteriorated and died on 22 January 1984 from cardiorespiratory arrest following presumed tonsillar herniation.

At necropsy the brain was oedematous and weighed 1200 g. There was bilateral tonsillar herniation. The basal meninges were thickened and pearly white in colour. Whitish gelatious exudate was seen filling the basal cisterns and covering the front of the pons and medulla. The cranial nerves and the basal vessels were entangled in the exudate. Sections revealed dilated lateral ventricles whose walls were smooth. There were no infarcts or abscesses in the brain substance. Microscopically the brain showed granulomatous lesions consisting of giant cells and epitheloid cells with necrotic centres containing fungi with broad non-septate hyphae. Sections from the nose and sinuses did not show evidence of necrosis or mucormycosis. A methanamine silver stain of brain confirmed that infection was mucormycosis. Gomori methanamine silver stains all forms of fungi black. Mucormycosis shows broad non-septate hyphae. The diameter of these hyphae is 10–15 µ, which differentiates them from Aspergillus, whose hyphae are septate and of 3–4 µ in diameter. The uniform dichotomous pattern of branching of hyphae in tissues also helps to differentiate Aspergillus from mucormycosis.

Rhinocerebral mucormycosis usually presents as acute meningitis, meningoencephalitis or cerebral abscess. Only three cases of chronic mucormycosis have so far been reported. These presented with typical sinus and rhinocerebral involvement and histologically showed chronic granulomatous inflammation. All had underlying metabolic disease. Our patient, however, had no such underlying illness. The history of convulsions, headache and vomiting suggested a space occupying lesion. The CT scan, however, only revealed hydrocephalus. In India tuberculous meningitis is the commonest cause of acquired hydrocephalus and our patient was given anti-tuberculous treatment. Of four reported patients with fungal meninigitis, primarily presenting as hydrocephalus, three had cryptococcal infection and one had aspergillosis. The diagnosis especially of cryptococcosis has become easier, but the diagnosis of mucormycosis remains notoriously difficult owing to the altered haemodynamics of CSF flow following basal meningitis and the fact that the CSF is nearly always devoid of the relevant organisms. Our patient died before any shunt operation could be performed. The conclusion would seem to be that fungal infection should be excluded in all patients with hydrocephalus of unknown aetiologie before shunt operations are undertaken.

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Impaired neurotransmitter amine metabolism in arginase deficiency

Sir: Arginase deficiency, an inborn error of the urea cycle leading to accumulation of arginine, causes a severe progressive neurodegenerative disorder characterised by mental retardation and a spastic diplegia. The mechanisms responsible for the neurological damage are uncertain. It is unlikely to be due to hyperammonaemia alone since only moderate ammonia accumulation occurs in this disorder which more closely resembles an aminoacidopathy such as phenylketonuria than the other urea cycle disorders. In a patient with arginase deficiency we have recently observed a disturbance of cerebrospinal fluid (CSF) catecholamine and serotonin metabolism similar to that in patients with "classical" phenylketonuria. The findings are consistent with the view that inhibition of aminoacid uptake by the brain is a com-