CASE REPORT

CEREBELLAR CRYPTOCOCCOMA IN AN IMMUNOCOMPETENT ADULT PATIENT

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Abstract. Isolated cerebellar cryptococcoma in an immunocompetent patient is a rare condition. We report a case of an immunocompetent adult with isolated cerebellar cryptococcoma. The patient presented with chronic headaches for one year and was found to have multiple cerebellar abscesses on imaging. Our patient underwent resection of the lesion and the cryptococcoma was subsequently diagnosed by histological examination. We initiated treatment with antifungal medicine and was successfully treated after 6 months of therapy. A cryptococcoma may be a cause of isolated cerebellar abscess in immunocompetent patients despite not finding cryptococcal antigen in the serum.

INTRODUCTION

Cryptococcosis of the central nervous system (CNS) is a disseminated opportunistic fungal infection and is found mainly in compromised patients with cell mediated immune defects, particularly in acquired immunodeficiency syndrome or in patients receiving corticosteroid and immunopressive therapy. In recent years, this infection has become increasingly noted to cause disease in immunocompetent hosts. It still remains a rare condition, particularly as an isolated cerebellar cryptococcoma which has never been described. We report a case of isolated cerebellar cryptococcoma in an immunocompetent patient.

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CASE REPORT

A 23-year-old man was admitted to our hospital with a complaint of chronic progressive headaches for one year previously diagnosed as migraines before this visit. The patient was a fisherman, with no particular exposure to birds or woodworking. He also gave a previous history of several non-bilious vomiting episodes. Two weeks prior to admission, he developed ataxic gait and worsening headaches. On the day of admission, his vital signs were normal and he had no clinical evidence of compromised immunity or clinical signs of localized infection in his body. A neurological examination revealed an alert, interactive, cooperative patient. Scanning speech, clumsy tandem walking, dysmetria and dysdiadokinesis were noted on neurological examination.

The patient was investigated using a contrasted-enhanced computed tomography (CT) scan of the brain, which demonstrated multiple brain abscesses in both hemispheres of the cerebellar vermis with adjacent edema and mass effect on the fourth ventricle with moderate non-communicating hydrocephalus (Fig 1). A complete blood count, urine analysis and clinical chemistry were normal. Human immunodeficiency virus antibody was repeatedly negative and the CD4 and CD8 T-cell lymphocyte counts were normal. A chest roentgenogram was also normal.

Six days after admission, the patient underwent neurosurgical resection of the cerebellar abscess. Histopathologic examination showed acute inflammation with necrotic areas, and the presence of budding yeast like organisms staining with Gomori methenamine silver. Results from the mucicarmine stain were consistent with cryptococcal infection (Fig 2). CSF analysis including cryptococcal antigen (CrAg) was normal. Blood, CSF and cerebellar debris tissue cultures were negative for fungal isolation or other organisms.

After the pathological report were obtained, a serum CrAg was reported as negative. The patient was treated with amphotericin B over 28 days. The headache and cerebellar signs gradually improved. A contrasted-enhanced CT scan of the brain was performed at follow-up, which showed a decrease in the size of the lesion and surrounding edema. His medication was changed to oral fluconazole (at 800 mg/day) which was continued for 6 months. At 6-months the patient had no residual symptoms and was well.

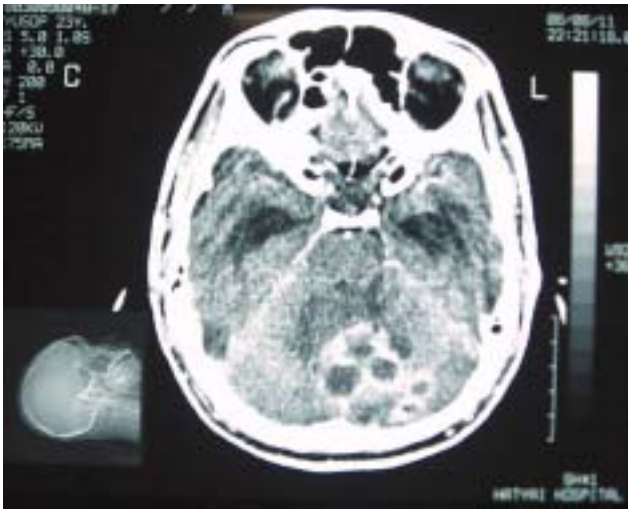

Fig 1–Contrasted-enhanced CT scan of the brain showing multiple brain abscesses in the posterior fossa region possibly in the cerebellar vermis and in both hemispheres, causing surrounding edema with effacement of the 4th ventricle and moderate hydrocephalus.

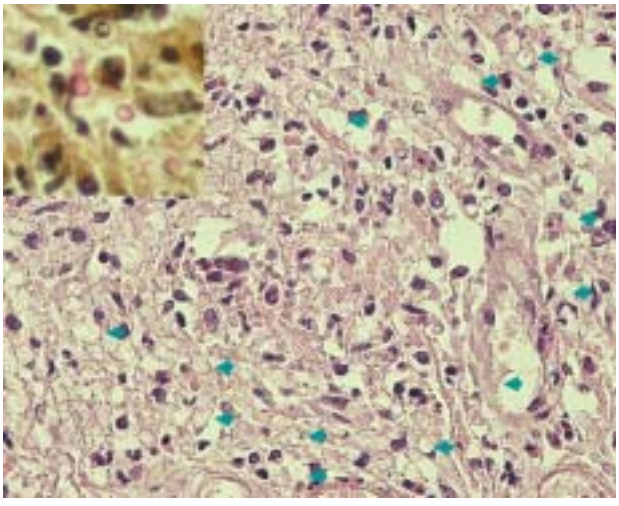

Fig 2–Histological section of cerebellar tissue revealing yeast-formed fungus (blue arrow) among the inflammatory and necrotic tissue. The fungus is about 5 to 10 μm in diameter and has a thick gelatinous capsule. (Hematoxylin-eosin stain; magnification x 40). Inset: The capsule is intensely red in color with a mucicarmine stain (black arrow). (Magnification x 100).

DISCUSSION

An isolated cerebellar cryptococcoma is a rare condition in immunocompetent patients. To our knowledge, only 2 cases of isolated cerebellar cryptococcoma have been previously reported in the English literature: one in an immunocompetent child (Gologorsky *et al*, 2007) and the other in an adult with co-existent sarcoidosis (Kanaly *et al*, 2007).

CNS cryptococcosis usually presents with non-specific symptoms of headache, lethargy, fever, weakness and anorexia, however, the clinical course is highly variable. Ecevit *et al* (2006) found the duration from onset of symptoms to diagnosis was variable, taking up to one year in immunocompetent patients. Headache is the most common presentation, Speed and Dunt (1995) reported the duration from initial presentation to diagnosis was longer in healthier host than in immunosuppressed hosts. This may be related to the effect of the host's immune response from the onset of symptoms until clinical presentation. Misdiagnoses, such as primary headache, stroke, carcinoma, chronic infection or sinusitis, also occurred in this patient group (Ecevit *et al*, 2006).

The diagnosis was challenging in this otherwise healthy patient. A neurosurgical approach is important to ensure a definite diagnosis and decompress the mass effect to prevent progressive hydrocephalus and brainstem compression. The cryptococcoma was diagnosed on histological examination even though the CSF and serum CrAg were negative. Evaluation of the CSF and checking a serum CrAg have become important methods to diagnose extrapulmonary cryptococcosis due to the high sensitivity and specificity (Frank *et al*, 1993). The diagnosis of cryptococcoma in this patient with negative serology for CrAg was a surprise.

A cryptococcoma is characterized by a localized tumor-like mass in which the fungus has invaded the brain parenchyma. Imaging of the CNS is essential to diagnose patients presenting with neurological abnormalities, however CT and magnetic resonance images have no pathognomonic appearances in people with a cryptococcoma. The findings have been found to persist on neuroimaging for up to 7 years (Hospenthal and Bennett, 2000).

Treatment for CNS cryptococcal disease is common, particularly in patients with HIV infection (Saag *et al*, 2000). In immunocompetent hosts with CNS cryptococcoma, treatment data are limited. Recommended practice guidelines should be adopted from the HIV-patient regimen (Saag *et al*, 2000). Our patient was successfully treated with amphotericin B followed by oral fluconazole. Duration of treatment is highly variable and depends on the patient's clinical status and the physician's decision.

Although most cerebellar abscesses are due to pyogenic infection, this case underlines the need to consider a cryptococcoma as a possible cause of chronic cerebellar abscesses in an immunocompetent patient. A definite diagnosis must be confirmed by histological examination for appropriate antifungal therapy to be given, even though the serum and CSF CrAg results are negative.

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