Case Report

The Brain Eating Amoeba! An Unusual Case Report

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ABSTRACT

Acanthamoeba is a eukaryotic protist that occurs world-wide and can potentially cause infections in humans and other animals. Acanthamoeba cause the insidious and mostly fatal disease, granulomatous amoebic encephalitis (GAE), particularly in immunocompromised or otherwise debilitated individuals. Acanthamoeba meningoencephalitis, also known as GAE is a rare, but nearly always fatal disease, caused by infection with Acanthamoeba species. The organism enters the nasal cavity when water contaminated with amoebae is aspirated. Subsequently, it invades the central nervous system through the olfactory neuroepithelium, disseminates via haematogenous spread, and migrates into CNS at the blood brain barrier, and causes a fatal infection.

We report a case of granulomatous amoebic meningoencephalitis (GAE) in an eleven year old male child, who was immunocompromised, a known case of AML (Acute Myeloid Leukaemia-type II), on chemotherapy, presenting with fever, headache, altered sensorium and seizures, with positive meningeal signs, where on wet mount cytology of CSF suggested Acanthamoeba.

Key words: AML, Acanthamoeba sp, Granulomatous Amoebic Meningoencephalitis, Rare disease.

INTRODUCTION

Acanthamoeba is an opportunistic protist pathogen that is ubiquitously distributed in the environment. [1] Acanthamoeba encephalitis is a fatal brain infection with a case fatality of more than 90%, despite advances in antimicrobial chemotherapy and supportive care. Natural infection is associated with haematogenous spread leading to Acanthamoeba entry into the brain, most likely via the blood-brain barrier, although blood-cerebrospinal fluid barrier has also been suggested as a possible route. [2] Once in the CNS, severe injury occurs, with most patients dying quickly from increased intracranial pressure and brainstem herniation. It is clinically indistinguishable from bacterial and viral causes of encephalitis, therefore we believe, that one should be familiar with the epidemiology, clinical presentation, diagnostic modalities, and treatment options that exist for this serious infection. The rapid progression of the disease process and limited awareness among the clinicians, pathologists and the microbiologists makes this disease a diagnostic challenge. We report a case of granulomatous amoebic encephalitis (GAE) due to Acanthamoeba sp. where the amoebae were detected in cerebrospinal fluid of an 11 year old boy, with a past history of AML.

CASE REPORT

Eleven year old male child, who was a known case of AML on chemotherapy, admitted in our hospital with history of fever on and off since four days, associated
with joint pains, headache, neck stiffness, vomiting, seizures and irritable sensorium. He had no other significant history.

**Investigations**

Complete blood count showed haemoglobin of 11 gm/dl, Leukocyte count of 3,400 /cu.mm, and platelets were 2.55 lakhs/cu.mm. No malarial parasites were detected in the peripheral blood smear examination. Chest X-ray and Urine examination was insignificant. CT brain showed no relevant finding. Serum SGOT and SGPT levels were within normal limits and CRP (C-reactive protein) was normal. Laboratory data of CSF revealed 2cc, colourless, hazy fluid, with specific gravity of 1.010. No cobweb was present. The total WBC count of CSF was 4,200 cells/cu.mm and RBC count was 1,800. DLC revealed predominantly polymorphonuclear leukocytes (90%) and lymphocytes (10%). Biochemical analysis showed glucose at a concentration of 27 mg/dl and proteins 960 mg/dl, with ADA being 13.5 and CSF lactate was markedly increased to 4.7 mmol/L. Rapid tests for HIV (J Mitra-Tridot), anti HCV and HBsAg were non-reactive. Gram stain showed scanty pus cells and bacteria, and culture was sterile after 48 hrs of incubation. Cryptococcus antigen detection was negative ruling out Cryptococcus neoformans, and Cytomegalovirus DNA detection done with real time PCR technique was found to be negative.

Pap and Gimesa stained smears of CSF showed predominantly neutrophils, few lymphocytes and few RBCs. No malignant cells were seen in the smears studied.

Microscopic examination of wet preparation of CSF, revealed amoeboid movements with the presence of acanthopodia along with scanty pus cells, suggestive of trophozoites of Acanthamoeba species. Following the demonstration of amoebae in the CSF, a diagnosis of Granulomatous Meningoencephalitis was made.

**DISCUSSION**

_Acanthamoeba_, a free living amoeba is commonly found in water resources such
as swimming pools, lakes and rivers. [3] It has also been isolated from contact lens cases and as contaminants in cell cultures. Despite the ubiquitous nature of the organism and common exposure, the incidence of human disease from *Acanthamoeba* is low. [4] GAE from *Acanthamoeba* occurs almost exclusively in immunocompromised hosts or debilitated patients. HIV disease, systemic lupus erythematosus, diabetes, chronic liver disease, chemotherapy, prolonged steroid, and organ and marrow transplantation have all been described as potential associations. [5] The symptoms of GAE are generally nonspecific and include headache (53%), meningismus (40%), fever (53%), mental status changes (86%), visual disturbances (26%), ataxia (20%), hemiparesis (53%), and seizures (66%). The exact incubation period is unknown but is probably weeks to months. The duration of disease is reported to range from 7 to 120 days (average, 39 days). The clinical course of GAE is often fatal. Laboratory Diagnosis of GAE is based on CSF analysis, using CSF wet mount and Geimsa-Wright staining technique, culture, and PCR. [6] PCR is especially useful in cases with low parasite density. [7] *Acanthamoeba* grows in culture on nonnutritive agar overlaid with Gram-negative bacteria. If a culture is positive, susceptibility testing should be done. [8] *Acanthamoebae* is sensitive to multiple antimicrobial agents. The GAE has been described rarely in the immunocompetent host. Granulomatous amebic encephalitis is uncommon and it is likely that it goes undiagnosed in many cases. Two other amebic genera are known to cause CNS infections, *Naegleria fowleri* and *Balamuthia mandrillaris*. While infections with *Balamuthia mandrillaris* are clinically very similar to infections with *Acanthamoeba, Naegleria fowleri* causes primary amebic meningoencephalitis, usually in immunocompetent individuals and children. To the best of our knowledge, this is the first case of GAE in childhood occurring during treatment of AML in which diagnosis of Acanthamoeba was confirmed. *Acanthamoeba* more commonly gain access to the CNS hematogenously either through the respiratory tract or skin, organisms can also enter through the olfactory route. They have also been isolated from the nasopharyngeal passages of healthy persons and may be considered normal flora. [9] Serologic surveys have detected serum antibodies against *Acanthamoeba* in healthy individuals. [10]

In this report, CSF showed pleocytosis with plenty of neutrophils and lymphocytes along with low glucose and high protein levels. Wet mount and special stains confirmed *Acanthamoeba* meningitis.

**CONCLUSION**

It is of paramount importance to identify patients with *Acanthamoeba* infections of the CNS as early diagnosis prevents mortality and serious complications, which is the norm in these infections. Recent studies show the importance of suspecting and evaluating patients for amoebic causes of CNS infections in order to prevent misdiagnosis and inappropriate treatment.

**REFERENCES**
