

PRIMARY AMEBIC MENINGOENCEPHALITIS

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ABSTRACT

This is the first case description of primary amebic meningoencephalitis (PAM) in an Iranian child. The patient was an eighteen months old boy referred to the hospital with signs and symptoms of acute meningitis. Cerebrospinal fluid (CSF) examination indicated pleocytosis with predominance of neutrophils, low sugar and high protein. Gram staining and culture of CSF for bacteria were negative. Microscopic examination of CSF revealed the motile amebae with pseudopodia. In Giemsa staining of CSF, the trophozoites of amebae were observed. The initial response to a six-week treatment course with amphotericin B, rifampin and chloramphenicol was satisfactory, but in the follow-ups the patient regressed neurologically. Although PAM is a rare and fatal disease, it should be differentiated from bacterial meningitis and treatment must be promptly carried out.

KEY WORDS: Primary amebic, Meningoencephalitis, Children.

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INTRODUCTION

Primary amebic meningoencephalitis (PAM) is a rare and fatal disease caused by free-living amebae (FLA) named *Naegleria fowleri*. This ameba is living in soil and fresh water. Most cases of PAM are seen in healthy children and young adults with a history of swimming in contaminated ponds.¹ The first case was reported by Fowler and Carter from Australia in 1965.² So far nearly 190 cases have been

reported from all around the world from which only 7 cases survived.²⁻⁸ This report describes the first case of PAM in Iran, of a patient who initially responded well to a course of amphotericin B, rifampin and chloramphenicol.

CASE REPORT

An eighteen months old boy from a rural area with a history of nausea, vomiting, fever, drowsiness and convulsion for 3 days was admitted to the children hospital in August 2001. He had the history of puddling in pond water in the southwest of Iran. In primary physical examination, he was drowsy and febrile (rectal temperature, 39.9°C), with respiratory rate of 70/min, pulse rate of 130b min, and blood pressure of 85/50mmHg. Meningeal signs were positive, but no focal neurologic signs were observed. The patient was responsive to the painful stimuli. An intact sensory, hypotonic motor and decreased deep tendon reflexes were noticed. The pupils were equally sized and reactive to light. Fundoscopy and other general examinations were unremarkable. The primary diagnosis was bacterial meningitis. The first examination

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of cerebrospinal fluid (CSF) revealed a turbid fluid with 12000/mm³ leukocytes predominantly neutrophil (83%), 1000/mm³ RBC, 238mg/dl protein and 34mg/dl glucose. Concomitant blood sugar was 100mg/dl. Gram staining and culture of CSF for bacteria were negative. In microscopic examination of CSF, motile trophozoites of amebae were observed. Fig-1 shows the amebae stained with Giemsa. 55mg/dl with no cells. The fever came down and the patient started opening his eyes, moving his hands and feet and also crying. From the day 20th, feeding was started with soup and milk. On the day 30th, he was able to sit, hold objects in his hands and also able to communicate with his parents. The treatment with rifampin, amphotericin B and chloramphenicol was continued for 6 weeks and with dexamethasone for 2 weeks. The patient took the same medications during the whole treatment period. On the day of discharge, the patient was able to keep his head straight, sitting and eating by himself, but unable to stand and walk. In the final neurological examination by the time of discharge, the patient was hypotonic with decreased deep tendon reflexes. Audiometric examination was normal. At the end of treatment; CBC, LFT, Na, K, BUN and creatinine were normal. In follow up visit after 3 months, he had no difficulties in vision, hearing, talking, walking, running and communicating with others. Six months later in second follow up, he had regressed neurologically and although his vision, hearing, sensory and motor functions were normal, he had a disturbed speech with some problems in communicating with others. In the third follow up (12 months after admission), the patient developed drop attacks and

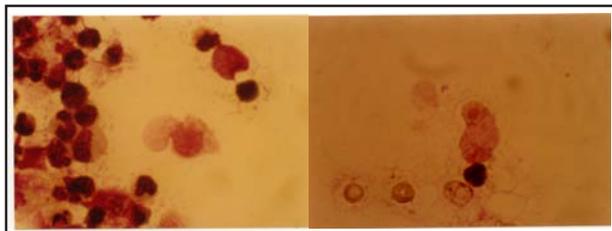


Fig-1: Trophozoites of Naegleria Fowleri and WBCs, RBCs in CSF smear.

occasional generalized tonic-clonic convulsions. Repeated EEG recordings in asleep state revealed disturbed background activity, absence of sleep patterns and frequent repetitive paroxysmal multifocal and generalized spike – wave and poly spike-wave discharge. In the last follow up (20 months after admission), despite taking sodium valproate plus pirimidone, he was still suffering from drop attacks, but less frequently, not able to eat by himself, not very well oriented to environment, no fixes and follows, although reactive pupils, had unsteady and wide base gait.

DISCUSSION

Human infection with FLA is an uncommon but life-threatening disease. FLA is responsible for 4 clinical syndromes. Granulomatous amebic encephalitis occurring in immunodeficient patients, disseminated granulomatous amebic disease (skin, pulmonary, and sinus infection), Amebic Keratitis causing chronic keratitis and PAM in children and young adults which causes acute and fatal meningoencephalitis. PAM is acquired through water entering the nasal passage through inhalation. CNS invasion by *N. fowleri* occurs after nasal inoculation with the amebae by disruption of the olfactory mucosa. The amebae penetrate the submucosal nervous plexus and cribriform palate gaining access to the CNS.² After an incubation period of 5-15 days, high fever, nausea, vomiting, headache, meningeal signs and changes in mental status may occur. The patient may rapidly develop into coma and death. The clinical manifestations are similar to bacterial meningitis and CSF examination indicates pleocytosis with predominance of neutrophils, low sugar and high protein. Definitive diagnosis of PAM is made by demonstration of amebae in wet mount preparation and stained smears of CSF or culture in enriched nutrient agar with *E. coli*.^{7,9} Several kinds of medication such as rifampin, chloramphenicol, miconazole, ketokonazole, amphotericin B and sulfisoxazole are used for the treatment of PAM.^{2,10} Despite the lack of adequate experience in treatment of this disease, our

patient was treated with chloramphenicol, rifampin and amphotericin B for 6 weeks. Survival of the patient could be due to the early diagnosis and treatment. In conclusion, although PAM is a rare disease with poor prognosis, it should be considered in differential diagnosis of bacterial meningitis specially when CSF smear and culture is negative and patient has a history of contact with contaminated fresh water. In that case, early treatment may be life saving.

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