FATAL RHINO-ORBITO-CEREBRAL INFECTION CAUSED BY SAKSENAEA VASIFORMIS IN AN IMMUNOCOMPETENT INDIVIDUAL: FIRST CASE REPORT FROM INDIA

A 56-year-old female patient presented with history of nasal blockage, nasal bleeding, headache, since one month. On admission the patient developed severe headache, decreased vision of eyes and blindness. Clinical diagnosis of rhino-orbital involvement was confirmed by computerized tomography of the paranasal sinuses. The diagnosis of fungal infection was confirmed by KOH examination, haematoxylin and eosin examination and Gomori’s methanamine silver stain of the biopsy material. Diagnosis was confirmed by culture on Sabouraud Dextrose agar, slide culture on Czapek Dox medium and the isolate was identified as Saksenaea vasiformis. The patient was started on intravenous amphotericin B (received only one dose before succumbing), but she did not respond to the treatment, developed hemiparesis, slurred speech, diminished reflexes and ultimately died. The involvement of the brain was confirmed by computerised tomographic scan.

We believe this case to be the first case of rhino-orbito-cerebral infection due to Saksenaea vasiformis and that of second case in an immunocompetent patient in the world.

Key words: Immunocompetent host, rhino-orbito-cerebral mucormycosis, Saksenaea vasiformis

Saksenaea vasiformis is a soil saprophytic fungus.[1] There is only one species in the genus Saksenaea i.e., Saksenaea vasiformis. As the fungus produces flask shaped or “vase shaped” sporangium, the name vasiformis is given to the species.[1] The vast majority of cases of S. vasiformis resulted from trauma, with introduction of spore-containing dirt into open lesions.[1,2-7] The other probable modes of infection in human beings are either inhalation of spores into sinuses or direct inoculation into facial wounds or sinuses by contaminated water.[1-7]

Various predisposing factors for this fungal infection described are trauma, needle stick injury, patients on antibiotics, steroids, diabetes mellitus and neutropenia associated with lymphoblastic leukemia.[1-9] Most cases of S. vasiformis are subcutaneous or disseminated.[1-7] Rhinocerebral disease with this fungus is uncommon, only one case described so far in the world.[1] Here we report the rare case of fatal rhino-orbito-cerebral infection in an immunocompetent host. This is the first case report from India.

Case Report

A 56-year-old female patient was admitted to the ward on 11th February 2007 with a history of nasal blockage and nasal bleeding and headache since one month. After admission the patient developed decreased vision of the left eye, ptosis and finally blindness of the left eye. The patient also complained of severe headache.

The laboratory investigations showed a haemoglobin of 10 gm/dL, total leukocyte count of 9000 /mm³, with a differential count of 80% neutrophils and 20% lymphocytes, raised ESR (40 mm at the end of one hour). The liver function and kidney function tests were within normal limits. The patient was non-diabetic and was HIV seronegative. The patient was immunocompetent with no other predisposing
factors.

The treating physician sent the biopsy samples for histopathology, which showed aseptate hyphae in the haematoxylin and eosin (H & E) stain.

CT scan of paranasal sinuses and the brain showed gross inflammatory soft tissue in bilateral ethmoidal sinuses and sphenoid sinus extending to the nasal cavity. Mucosal thickening was observed in both the sinuses. Diffuse ischemic changes in bilateral periventricular white matter were observed. There was ill defined hypodense soft tissue seen abutting right rectus muscle with oedema of adjacent retro-orbital fat. Soft tissue was also seen in left retro-orbital fat adjacent to the optic nerve (Fig. 1a). Repeat biopsy from nasal mucosa was taken which showed distorted aseptate hyphae in 10% KOH preparation (Fig.1b). Histopathology was also repeated and stained with H and E (Fig. 1c) and Gomori’s methenamine silver staining (Fig. 1d), which showed findings suggestive of mucormycosis. Based on the microscopic findings, partial surgical debridement of the mass was done and the patient was started on intravenous amphotericin B. There was a delay of seven days in starting the antifungal treatment from the appearance of initial symptoms i.e., decreased vision of the left eye, ptosis, blindness and severe headache.

Biopsy samples were also taken for culture and inoculated on Sabouraud Dextrose agar (SDA) without antibiotics and actidione. After 48 hours of incubation, luxurious whitish cottony growth, typical of mucormycosis was observed (Fig. 1e). Lactophenol cotton blue (LPCB) preparation was performed from the growth, which showed only aseptate hyphae with no sporulation. Slide culture was performed with potato dextrose agar (PDA) and SDA, incubated at 25°C. LPCB preparation from both the slide cultures did not reveal sporulation even after 15 days of incubation. Two sets of slide cultures were performed on Czapek Dox media and incubated at room temperature. Typical flask shaped sporangia and rhizoids were observed in LPCB preparation from Czapek Dox agar within seven days of incubation, suggestive of Saksenaea vasiformis (Fig. 1f).

Meanwhile the patient developed right hemiparesis, high grade fever, slurred speech, diminished reflexes and ultimately succumbed to death which was attributed to the cerebral mucormycosis (the patient could receive only one dose of intravenous amphotericin B before her death).

Discussion

Infection with zygomycetic fungi is well recognized but most infections occur in persons with underlying disorders of various types, such as uncontrolled diabetes mellitus,
haematological malignancies, severe malnutrition, chronic renal failure, chronic hepatic diseases or immunodeficiency disorders etc. The common causative organism is *Rhizopus* spp., although others like *Mucor* and *Absidia* are also frequently seen, whereas *Saksenaea vasiformis* and *Apophysomyces elegans* are rare pathogens. Most reported cases of *S. vasiformis* are subcutaneous or cutaneous, and *Koren et al.* described primary invasive cutaneous mucormycosis in non-immunocompromised. The patient was a 72-year-old non-diabetic. He was successfully treated with debridement of the lesion and amphotericin B. Disseminated zygomycosis by *S. vasiformis* in a 14-year-old child without any predisposing factors was reported by Hey *et al.* in 1983. The child from Iraq presented with acute febrile illness accompanied by large necrotic skin lesions and bronchopneumonia from which he died. Dean *et al.* reported cranial zygomycosis caused by *S. vasiformis*. He reported that previously healthy 19 year old man who had sustained severe head trauma had received steroids and broad spectrum antibiotics and developed cranial zygomycosis. This was the first time that this zygomycete species had been implicated as disease agent. Rhinocerebral infections by *S. vasiformis* was reported in 1988 by Kaufmann *et al.* In our patient also there was no predisposing factor for rhinocerebral zygomycosis, the patient being immunocompetent but the patient died in spite of treatment with amphotericin B of which the patient could receive only one dose. There are few reports of infections caused by *S. vasiformis* from India. Padhye *et al.* reported the first case of subcutaneous zygomycotic infections in rice mill worker by *S. vasiformis* in India in 1988. The infection confined to the left foot showed multiple draining sinuses, low grade fever following a crushing injury, when a log fell on his foot. Necrosis of the affected area led to amputation of the forefoot. A whole thickness graft was affected and treatment with potassium iodide cured the patient. Other reports also showed cutaneous or subcutaneous infection.

This is the first case report from India, where *S. vasiformis* is reported to cause fatal rhino-orbito-cerebral infection in immunocompetent individual. This is also the second case of rhino-orbito-cerebral infection due to *S. vasiformis* from the world.

The diagnosis of *S. vasiformis* may be missed as it usually does not sporulate easily; but can lead to a fatal disease. Hence when a zygomycete species is isolated, it should be subcultured on nutritionally deficient media to hasten identification and to start treatment promptly. The media used to induce sporulation are agar blocks containing hyphal growth on sterile distilled water, with sterilized yeast extract solution added to it, or Czapek Dox agar and in most of the cases, sporulation was successful. In our case, the organism produced abundant flask shaped sporangia and rhizoids just beneath it only on Czapek Dox agar.

To conclude, although *S. vasiformis* causes cases of subcutaneous zygomycosis it may occasionally cause acute fulminant fungal sinusitis leading to rhino-orbital and rhino-orbito-cerebral disease. Early diagnosis and treatment may save the life of the patient, as in the present case the patient probably died due to the delayed diagnosis and due to the initial suspicion of malignancy. Therefore, a very high index of suspicion is required to diagnose such rare cases of fungal sinusitis.

References


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