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SINONASAL MUCORMYCOSIS IN AN INFANT- A CASE REPORT

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ABSTRACT

Paediatric invasive fungal rhinosinusitis is a very rare entity. The diagnosis is usually delayed due to its nonspecific clinical presentation in children. ***Case Report-*** We present a case of 6-month-old female child who presented with an acute history of swelling of the nose and nasolabial folds. The patient was admitted to inpatient care. Haematological investigation and radiology were done. The patient underwent nasal endoscopy and biopsy of an intranasal mass under general anaesthesia. Histopathology confirmed the diagnosis of sinonasal mucormycosis. ***Discussion-*** Mucormycosis in the paediatric age group is itself a rare phenomenon. In this case, the child had anaemia and no other immunocompromising factors that predispose to invasive fungal infection. The presentation is also unique as there was an acute history of swelling and nasal mass with no other complaints. Clinically, this presentation posed a diagnostic challenge. ***Conclusion-*** The clinical presentation of sinonasal mucormycosis is highly variable in the paediatric age group. Moreover, the possibility of this infection cannot be ruled out in an immunocompetent host. A high index of clinical suspicion is mandatory for diagnosis. Early diagnosis and aggressive management have been advocated for better results.

KEYWORDS

mucormycosis, infant, case report, fungal, rhinosinusitis, pediatric

INTRODUCTION

Sinonasal mucormycosis is a disease which usually affects immunocompromised patients with impaired neutrophilic response. The risk factors include haematological malignancies, uncontrolled diabetes mellitus, organ transplantation, malnutrition, chemotherapy, desferoxamine therapy [1]. It rarely affects the immunocompetent host. Mucormycosis is a rare infection in the paediatric age group with <1% incidence. Overall mortality related to mucormycosis is high (50-85%), especially in immunocompromised patients. High mortality is attributed to untreated disease, disseminated infection at age <1 year [2]. High clinical suspicion and aggressive chemotherapy with surgical debridement of devitalised tissue are key to improved survival rate.

We present a rare case of paediatric acute invasive fungal rhinosinusitis in a child (6-month) with iron deficiency anaemia. The atypical clinical findings in this case posed a diagnostic dilemma. In this article, we have discussed in detail the clinical presentation, diagnosis, and treatment challenges.

CASE STUDY

A 6-month-old female child presented to ENT emergency with a history of swelling of the right nasal dorsum for 3 days (Figure1). The swelling was associated with nasal discharge and watering from the right eye for one day. There was no history of fever, nasal bleed, trauma, feeding, or breathing difficulties. The child was born of non-consanguineous marriage by full-term normal vaginal delivery. Developmental milestones achieved and immunised for age. In the past, family and medication history were not significant. The patient had no allergies as mentioned by parents. On physical examination there was diffuse swelling of the dorsum of the nose and involving the right nasolabial fold. It rapidly progressed to involve the right cheek and upper lip. The overlying skin was erythematous and scaly (Figure 2). A pinkish mass visualised in the right nostril seemed to be arising from the lateral nasal wall. No other intranasal structure could be seen because the nasal mass was obscuring the view. On palpation, the swelling was tender and firm with ill-defined margins. No intraoral bulge was seen.

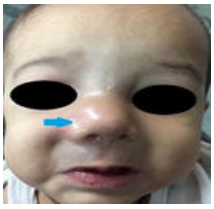

Figure 1-The blue arrow shows swelling and inflammation of right nasolabial fold and dorsum of nose.

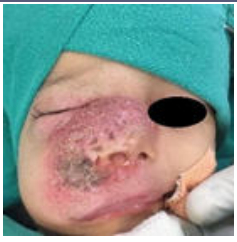

Figure 2- Clinical image showing progression of swelling and skin involvement.

The child was admitted to inpatient care. Ophthalmology consult was taken. Routine Hematological investigations (Hemoglobin, peripheral blood smear, WBC, LFT, KFT, Serum electrolytes, blood group, blood glucose) and Computed tomography of paranasal sinuses (contrast enhanced) were ordered. Blood investigation revealed iron deficiency anemia with Hb- 7.3g/dl. Blood transfusion was given under the guidance of the pediatric team to correct the anemia and later replaced by iron supplementation. CT scan (figure- 3) showed a heterogeneously enhancing mass in the right nasal cavity extending till the subcutaneous plane anteriorly and choana posteriorly and laterally eroding and filling the maxillary sinus and inferomedial wall of orbit. The infection was rapidly progressive and there was no response to I/V antibiotics. The patient underwent endoscopic nasal biopsy under general anesthesia and histopathology showed aseptate branching hyphae suggesting mucormycosis.

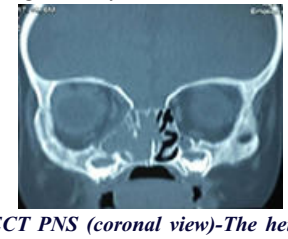

Figure 3- CECT PNS (coronal view)-The heterogenous lesion is epicentered in right nasal cavity with extension to orbit, maxillary sinus, subcutaneous tissue, nasal vestibule and nasal septum.

The child was then referred to the paediatric intensive care unit. Liposomal amphotericin (dose-3mg/kg) was started by intravenous infusion. Hepatic and renal function tests, blood counts, and plasma electrolyte (including plasma potassium and magnesium concentration) monitoring were done. The patient underwent endoscopic debridement. Endoscopic examination revealed a pinkish mass filling the entire nasal cavity and extending to the maxillary sinus. There was no fungal debris or black crust seen which are characteristic

of mucormycosis. Amphotericin was given until 32nd day of starting the drug as three consecutive biopsies were negative for mucormycosis. The skin changes were reversed at the end of treatment. The child responded well to treatment and was discharged after completion of antifungal therapy.

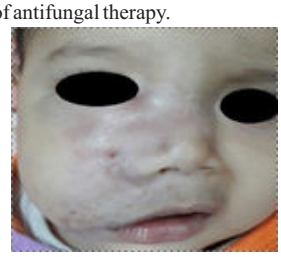

Figure 4: Improvement In Skin Changes Seen At Day 10 Of Antifungal Therapy.

DISCUSSION

Mucormycosis in children is associated with significant morbidity and mortality [1]. Mucorales are generally characterized by their tendency for rapid vascular invasion leading to vascular thrombosis and tissue necrosis [2]. In previous paediatric reviews, prematurity at birth and age <1 year are independent risk factors for mortality [3]. As there are very few cases [R] been reported, the true incidence of the disease in children is difficult to estimate.

In a systematic review, 226 cases of paediatric cases were studied. This review showed that the median age was 10 years with male predominance. The most common risk factors include haematological malignancies and neutropenia, followed by other factors. However, 10% of the patients had no risk factors. Sinus and disseminated disease were the most common manifestations [4]. On reviewing the literature studies, we found out that very few cases are reported in the neonatal and infant age groups.

The initial presentation of a fulminant type of disease resembles cellulitis. The symptoms could be facial swelling, numbness, pain, periorbital cellulitis, and fever of unknown origin. Clinical diagnosis is difficult in early stages as the symptoms are highly variable and not specific. This progresses to tissue infarction and eschar formation which is characteristic of mucormycosis [5]. Our patient presented at an early cellulitic stage with no obvious immunocompromising factors, hence posing a diagnostic challenge. The patient was diagnosed with iron deficiency anaemia based on haematological investigation. Hasan et al. concluded in an observational study that humoral nonspecific immunity (phagocytic activity and oxidative burst), and IL-6 are influenced in patients with iron deficiency anaemia [6]. In our case, iron deficiency anaemia was found to be the predisposing factor for mucormycosis.

The characteristic histological features are mycotic infiltration of blood vessels, vasculitis with thrombosis, tissue infarction, haemorrhage, and acute neutrophilic infiltrate [7]. The tissue infarction manifests as necrosis and black eschar formation. Histopathology is confirmatory for the diagnosis of acute invasive fungal rhinosinusitis.
CT is the initial radiological investigation of choice; however, findings are nonspecific and include unilateral mucosal thickening or infiltration of the periantral fat. Signs of bony dehiscence and orbito-cerebral involvement are late findings [8].

Multimodal treatment approach involving prompt institution of appropriate antifungal therapy with amphotericin B, reversal of underlying predisposing conditions, and, where possible, surgical debridement of devitalized tissue is mandatory to achieve therapeutic success. Amphotericin B in different formulations remains the drug of choice for mucormycosis. Outcomes are highly dependent upon the degree of immunosuppression, site and extent of infection, timeliness of therapy, and the type of treatment provided [9]. Our patient was managed with antifungal therapy, surgical debridement, and correction of anaemia and responded well to treatment.

CONCLUSIONS

Invasive fungal rhinosinusitis (mucormycosis) is a life-threatening condition and is rare in children. However, a high index of clinical suspicion in immunocompromised children is mandatory. Even iron deficiency anaemia could be a risk factor to predispose the host (especially children) to get infected. There are variable symptoms which must be kept in mind while making a clinical diagnosis of mucormycosis. Contrast enhanced CT of paranasal sinus and orbit has a limited value in early-stage disease; specific signs of bone erosion and orbito-cerebral involvement are late findings. Endoscopic sinonasal evaluation and histopathological examination is the mainstay for early diagnosis. The key to managing the condition is identifying and treating the predisposing factor, histopathological diagnosis, aggressive chemotherapy, and surgical debridement.

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