Case Report

First case of extensive spinal cord infection with Aspergillus nidulans in a child with chronic granulomatous disease

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Abstract

Chronic granulomatous disease (CGD) is characterized by a defect in phagocytic cells that lead to recurrent bacterial and fungal infections. The etiology of most common fungal infections in CGD are Aspergillus species. Aspergillus nidulans is one of several species of Aspergillus with low pathogenicity. However, it was reported to cause fatal invasive Aspergillosis in patients with CGD. Here we report the first cured invasive Aspergillus nidulans infection with extensive involvement of the spinal cord in a five-year-old child with CGD.

Keywords: CGD, chronic granulomatous disease, Aspergillus nidulans, spinal cord


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Introduction

Patients with chronic granulomatous disease (CGD) are at risk for fungal infections such as invasive Aspergillosis [1]. The mortality of invasive Aspergillosis is high despite specific antifungal treatment, particularly for cases that involve the spinal cord in CGD [2]. Only a few cases have been reported for A. nidulans invasive infections, almost exclusively in patients with CGD [3]. Here we add a case of A. nidulans infection that caused extensive spinal cord involvement in a five-year-old boy with CGD. At the best of our knowledge, this represents the first cured case of extensive A. nidulans infection of the spinal cord in a child with primary immunodeficiency.

Case Report

A five-year-old Saudi boy with a known case of chronic granulomatous disease was referred to our medical center with a history of progressive torticollis, upper back swelling, and weight loss with no fever for one month’s duration.

Physical examination revealed that the patient looked ill; he was afebrile; and he had swelling at the mid region of the upper part of his back. The swelling measured 5 x 8 cm with fluctuation without redness or hotness of the overlying skin. The rest of the physical examination including the neurological examination was normal. In order to reveal the underlying cause of this swelling, magnetic resonance imaging (MRI) of the brain and spine were requested and it showed extensive para-vertebral, epidural and subcutaneous abscess with marked spinal cord compression involving the dorsal spine (Figure 1).

Because of these findings in the MRI and after discussion with the neurosurgeon, the patient underwent extensive surgical debridement of the paraspinal and subcutaneous abscess. Histopathology studies showed fungal hyphae with evidence of chronic inflammatory changes; culture of the debrided material grew A. nidulans (Figure 2).

The patient was treated with antifungal drug (Voriconazole 11 mg/kg/dose) twice daily for eighteen months, and serial follow-up MRI showed complete resolution of the inflammatory process (Figure 3). After one and a half years of antifungal treatment, bone marrow transplantation (BMT) was performed with successful results.

Discussion

Chronic granulomatous disease (CGD) is a rare inherited disorder of the NADPH oxidase complex in which phagocytes are defective in generating reactive oxidants. As a result, patients with CGD suffer from recurrent bacterial and fungal infections [1]. In CGD,
Fig. 1. Case MRI of the spine.

MRI of the spine shows; extensive inflammatory process within the cervico-dorsal spine with high signal intensity in T1 (A) and low in T2 (B), with paravertebral and epidural abscess formation causing along segment of spinal stenosis, extensive syringomyelia. There is significant inflammatory process involving dorsal vertebrae and subcutaneous area along with abscess formation.

Fig. 2. Case microscopy.

(A); hisopathology study with GMS show fungal hyphae. (B); *Aspergillus nidulans* colonies on Czapekdox agar after 10 days of incubation. (C); conidial head of *Aspergillus nidulans* on Lactophenol Cotton Blue Stain
Fungal infections are caused mainly by Aspergillus species[2]. Aspergillus fumigates considered to be a common pathogen in CGD compared to other Aspergillus species; however, A. nidulans was found to be more virulent and can cause disseminated disease. It is, therefore, more likely to result in death [3,4]. In addition, A. nidulans is generally refractory to intensive antifungal therapy [4,5].

Central nervous system infections caused by Aspergillus species are uncommon, and infection involving the spinal cord is considered to be a rare event [5].

There were few reported cases of A. nidulans infections in patients with CGD worldwide. Among those few reports the infection mainly involved lung and bone, but none was reported with spinal cord involvement [6]. Kim M et al. report a fatal case of A. nidulans in a six-year-old boy with CGD in which A. nidulans was isolated from culture of the paraspinal abscess. The patient succumbed despite prolonged treatment with a high dose of amphotericin B, Itraconazole and interferon-gamma.

Our patient had invasive A. nidulans infection that extended from the lung to the vertebrae and caused subcutaneous, paraspinal and epidural abscess with spinal cord compression and extensive syringomyelia. The patient received voriconazole 11 mg/kg/dose twice daily, and unlike the few reported cases elsewhere, he was completely cured and able to undergo bone marrow transplantation successfully.

References

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