RHINO-FACIAL CONIDIHOLOMYCOSIS IN YOUNG VIETNAMESE MEN

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SUMMARY

Conidiobolomycosis is a rare fungal infection caused by Conidiobolus coronatus. We provide the first case of rhino-facial conidiobolomycosis in the North of Vietnam. A 32-year-old rural male patient presented with some diffuse subcutaneous nodules affecting centrofacial area. Histopathology showed granulomas with hyphae known as Splendore-Hoeppli phenomenon. The patient was successfully treated with combination therapy including saturated solution of potassium iodide and itraconazole.

1. INTRODUCTION

Conidiobolomycosis (also known as Rhinoentomophthoramycosis) is a rare, chronic, localized, deep fungal infection. It affects skin and soft tissue characterized by the granulomatous infection arising from a lesion that originates in the inferior nostril [1]. The infection spreads through the ostia, the foramina to involve facial, subcutaneous tissues and the paranasal sinuses. The first identification of the fungus from a human case was made in 1965 in Jamaica [2]. It is an uncommon disease, with few cases being reported from the tropical and subtropical zones of Africa, Asia and Americas [3]. Here, we present a rare case in Vietnam of Conidiobolomycosis.

2. CASE REPORT

A 32-year-old man from rural Vietnam with a nasal mass having been evaluated since 2 years. Nasal polyp was given as pathology and treated by excision. Four months after surgical intervention, the right nasal wing began to swell and stiffen (Fig. 1a). The mass was light red, painless. The patient was operated the second time at National Otorhinolaryngology Hospital of Vietnam. However, the mass continued spreading to the forehead, cheeks (Fig. 1b,c). Improvement was not noted after 4 interventions, as well as antibiotic treatment. Then, a course of methylprednisolone 16mg per day was prescribed within 1 month. His facial swelling initially improved, but relapsed when the dose of methylprednisolone was reduced to 8mg per day.

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Fig. 1: Nasal mass distorted the dorsum of the nose 1 year ago (a), then on the forehead 8 months ago (b) and on the right wing of nose 6 months ago (c).

Histopathology was realized many times. The patient was referred to the National hospital of Dermatology and Venereology (Fig. 2) and histopathology was checked again. At the end, conidiobolomycosis was identified.

Fig. 2: Two nontender, firm subcutaneous nodules in the right cheek

History of insect bites or nasal trauma was not noted. He did not have any immunodeficiency disorders or autoimmune diseases. Physical examination revealed two nontender, firm subcutaneous nodules affecting right cheek. Surgical scarring of the cheek, nose and forehead. No cervical lymphadenopathy was found and the immunological system appeared normal. Biological investigation was normal.
The ultrasound revealed the image of hypoecho, well-difined mass, 40 x 26 x 12mm and 17 x 8mm. CT scan revealed a diffuse thickening of the soft tissue (Fig. 3a). KOH preparation of nasal biopsy tissue revealed some broad, aseptate, thin-walled ribbon-like hyphae with wide-angle branching (Fig. 3b), but the culture on Sabouraud’s agar was negative.

The histopathology showed florid granulomatous inflammation along with multifocally striking deposition of crystalline-like eosinophilic material, know as the Splendore-Hoeppli phenomenon (Fig. 3c,d).

Fig. 3: (a) A diffuse thickening of the soft tissue with heterogenous enhancement in the right maxillary sinus and subcutaneous (arrows) on CT scan. (b) Broad, thin-walled, hyaline fungal filaments on KOH 10% preparation (x40). (c) Aseptate hyphae along with granulomatous infiltrate consisting of eosinophils and multinucleated giant cells are seen. (d) Hyphae of cross section with a thick eosinophilic sheath (Splendore–Hoeppli phenomenon) within giant cells stained with PAS.
Patient was treated by Saturated Solution of Potassium Iodide 1g/ml (SSKI). Initially SSKI was given 5 drops/dose three times per day, then gradually increased up to 20 drops/dose three times per day. After 1 month, the lesion stopped developing and had a mild improvement. Oral itraconazole 200mg/day was added to the regimen. The combination treatment led to improvement and the lesion gradually disappeared after 2 months (Fig. 4). We have planned to treat this patient for at least 6 months with antifungals and recheck with CT scans.

Fig. 4. After 2 months of treatment by SSKI

3. DISCUSSION

Conidiobolomycosis is a deep mycosis usually caused by *Conidiobolus coronatus*, a mould belonging to the order Entomophthorales of the class Zygomycetes, a saprophyte that lives in humus and on decomposing plant materials in moist and warm climates. It has been described in South East Asia, Africa, South and Central America. Infection is acquired through inhalation of spores or through their introduction into the nasal mucosa by soiled hands. Men aged 20-60 years with outdoor occupations such as agriculture are most likely to be affected. In a study involving 199 patients of Choon et al, age ranged from 3 months to 72 years with an average age of 34. Our patient was a worker from Thai Binh province, in the countryside of Vietnam. Until this moment, we have not found any other clinical cases of Conidiobolomycosis reported in Vietnam.

Infection of nasal mucosa usually begins unilaterally, but later spreads bilaterally. Symptoms include nasal obstruction, discharge and sinus pain. Subcutaneous nodules may
develop in the nasal and perinasal regions, with progressive facial swelling that can involve the upper lip. Dysphagia and laryngeal obstruction have been developed with sub-mucosal pharyngeal involvement.

The culture of the causative fungus is confirmatory, although culture attempts are often unsuccessful [4]. It is also suggested to use MGYP agar medium to culture this fungus.

Histopathologically, aseptate hyphae along with granulomatous infiltrate consisting of eosinophils and multinucleated giant cells are seen. The star like eosinophilic projections has been described as Splendore - Hoepli phenomenon. Splendore - Hoepli phenomenon is described with other fungal and parasitic diseases, but presence of aseptate hyphae along with this phenomenon is usually enough to make a diagnosis of entomophthoramycosis. Although, aseptate hyphae is also seen with mucormycosis, but Splendore-Hoepli phenomenon is rarely described in mucormycosis.

Diagnosis of conidiobolomycosis is usually based on both clinical presentation and histopathology. Fungal cultures are usually negative, with a positive rate of about 15%, although microscopy can be positive.

There is no consensus for the antifungal drug of choice for the management of conidiobolomycosis. Treatment is difficult because the diagnosis is usually delayed. The literature provides several therapies such as monotherapy with systemic antifungals, combination therapy with multiple antifungals and surgical debridement.

We observed that SSKI followed by itraconazole were the most commonly used drug either alone or in combination. Gupta et al studied on ten patients with conidiobolomycosis treated with SSKI and itraconazole (200mg twice daily) and followed up more than 1 year after stopping treatment [5]. Sevens patients responded to the combination treatment. However, the response was minimal (< 25% regression of the swelling) in 2 patients and one patient did not have any improvement after 6 months of treatment.

Prognosis depends on several factors, but in general, the cure/improvement rate is 83% with antifungals and surgical excisions according to the meta-analysis of Choon et al on 199 cases [6]. Late disease with delay in antifungal therapy, involvement of orbit or the brain, nonstereotypical presentation, immunocompromised patients, absence of Splendore-Hoepli Phenomenon, female gender are the factors that have a poor prognosis.

This is the first case that we confirmed the pathology. Four years ago, there was a similar case with the same clinical manifestation. However, the fungal test was not realized. We had been treated with many drugs without improvement. At the end, he had been treated with itraconazole. However, we had no information from him.

4. CONCLUSIONS

Conidiobolomycosis remains a diagnostic challenge in tropical regions. It should be brought into mind as differential diagnosis of subcutaneous swelling in the rhino-facial region especially in male agricultural workers. Treatment should be started early to avoid complications such as facial elephantiasis and obstruction and to decrease the cost of treatment as well as the morbidity.
REFERENCES


