CASE REPORT

CHROMOBLASTOMYCOSIS: A RARE PRESENTATION WITH POLYMORPHIC CUTANEOUS LESIONS AND BONE INVOLVEMENT, CAUSED BY *EXOPHIALA JANSELMEI*

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Chromoblastomycosis, a chronic fungal infection of skin and subcutaneous tissue arises as a result of traumatic inoculation of exposed areas of the body. We present a unique case of chromoblastomycosis caused by *Exophiala janselmei* in a female farmer who presented with multiple smooth non-tender nodules on trunk and limbs for 5 years and pigmented indurated plaques on her face for 2 years along with deformities of her hands. Imaging investigations revealed multiple lytic lesions in the bones of the upper and lower limbs. Histopathological findings showed characteristic sclerotic bodies, consistent with the diagnosis of chromoblastomycosis. She was started on a combination of oral antifungals with a good response. This case highlights the importance of high suspicion and early diagnosis of deep fungal infections in order to avoid disfigurement and comorbidities.

Keywords: Chromoblastomycosis; Cutaneous and bone involvement; *Exophiala janselmei*; Combination antifungals

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INTRODUCTION

Chromoblastomycosis is a chronic cutaneous and subcutaneous infection, caused by dematiaceous fungi, commonly being *Fonsecea Pedrosi, Cladophialophora Carrionii* and *Phialophora Verrucosa*. Less commonly *Exophialophora Janselmei* and *Exophialophora Spinofera* may also form sclerotic bodies although they are mostly implicated in pheohypomycosis. Lesions arise as small papules on exposed parts of the body and usually progress to form verrucous plaques or nodules. Less frequently it may spread to involve distant cutaneous sites and rarely other organs including lymph nodes, lungs, brain or bones through a lymphatic or haematogenous route. Infection is difficult to treat with frequent relapses. Combination therapy with oral antifungals has been reported to be well tolerated with good response. We describe a case of chromoblastomycosis in a female patient with no apparent comorbidities, who presented with extensive cutaneous and skeletal involvement.

CASE REPORT

A 35-years-old female farmer, a resident of KPK Pakistan, presented with multiple indolent asymptomatic nodular swellings on the trunk and limbs for 5 years and raised lesions on the face for 3 years. Started initially as small pea-sized swellings on forearms, gradually progressed in size and involved other areas of the body. Two years later she noticed raised annular pigmented lesions developing on cheeks, forehead and chin. On further inquiry, she complained of deformity of her hands due to swellings. However, there was no history of morning stiffness, pain, tenderness or restriction of movement at these joints. She also denied a history of photosensitivity, oral ulcers or hair loss. There was no fever, weight loss, cough or night sweats. Family history was insignificant regarding her skin lesions with no history of TB or any other chronic illness in the family. Her past medical and surgical history was unremarkable.

On examination, she was vitally stable with mild pallor and no lymphadenopathy. On cutaneous examination, the patient had polymorphic lesions. Multiple hyperpigmented indurated scaly plaques with central atrophy present on the face predominantly involving the nose, cheeks and chin. Multiple skin-coloured swellings of variable sizes were distributed over the trunk and extremities. Swellings were soft, non-tender, and mobile with smooth surfaces no temperature changes and no discharge. The overlying skin was normal. There was fixed flexion deformity of the right index finger and bulbous swellings were present on the proximal end of the index, middle and forefinger of the left hand. However, there was no tenderness, erythema or limitation of movement appreciated at these joints.

Her blood complete picture showed Hb 10.2 mg/dl and MCV 79.1 fl. ESR 44mm/hour. Her liver function tests, renal function tests and coagulation profile were within normal limits. HBsAg,

AntiHCVAb and HIV serology were non reactive and Mantoux test was negative. Her ACE levels were 15 nmol/ml/min.

In imaging investigations her chest X-ray was normal. X-rays of upper and lower limbs showed multiple well-defined osteolytic lesions along with multiple soft tissue densities seen in hands, forearms, arms and legs. Skeletal bone scintigraphy showed multifocal bone pathology involving bilateral radius, bilateral ulna, bilateral tibia and small bones of hands. A skin biopsy was done on a nodular lesion on the left forearm. H&E stained sections showed atrophic epidermis. Underlying dermis showed extensive areas of necrosis, sheets of foamy histiocytes along with numerous multinucleated giant cells with engulfed pigmented fungal sclerotic bodies consistent with chromoblastomycosis. Fungal cultures showed growth of *Exophiala jeanselmei.* Tissue sections from the pigmented plaque on the chin also came suggestive of Chromoblastomycosis with a large number of multinucleated giant cells engulfing pigmented fungal sclerotic bodies in the dermis.

As the patient had extensive cutaneous lesions with bone involvement, she was started on combination treatment with Itraconzole 200 mg twice daily and Terbinafine 250 mg once daily. Her lesions started improving by the second month and on a follow-up visit at 6 months she had marked improvement and visible clearance of her cutaneous lesions. In order to avoid excessive load on the liver Terbinafine was stopped at six months and Itraconazole was continued, with no side effects reported till now.

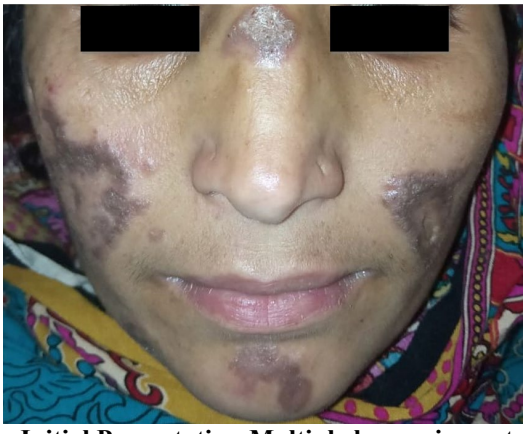
DISCUSSION

Chromoblastomycosis usually arises in farmers in tropical and subtropical areas of the world. Clinically it has five distinct forms including verrucous, nodular, tumoral, plaque and cicatricial types. In severe cases different morphological lesions may arise either close together or distantly by autoinoculation, lymphatic or hematogenous spread. Extracutaneous spread is infrequent although rarely it may involve lymph nodes, lungs, brain or bones.[1] Uptil now there are very few cases reported of chromoblastomycosis with skeletal involvement. In bones it usually presents as osteolytic lesions secondary to contiguous spread.[2] Javed *et al* reported case of a 20 years old female who presented with hyperpigmented verrucous plaques involving both sides of face with similar scattered lesions on ears, arms and legs along with lytic lesions in x-rays of hands and feet.[3] Another similar case was reported by Khan *et al,*12 years old boy presented with extensive hyperpigmented verrucous plaques on body along with x-rays showing multiloculated lytic lesions in right fifth toe and adjoining metatarsal.[4] In our patient widespread cutaneous involvement resulted in multiple lytic lesions in the adjacent bones of the upper and lower limbs.

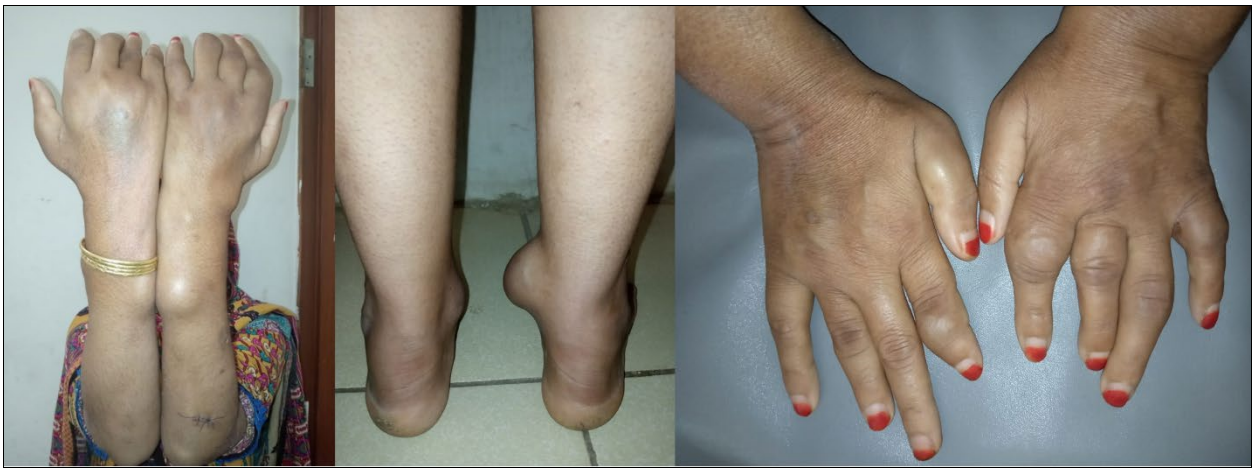
Cultures came positive for *Exophila Janselmei*, which is a rare causative agent of Chromoblastomycosis, commonly known to cause Pheohypomycosis. In a review article of 169 cases of chromoblastomycosis from India, cultures were positive for Fonsecaea species (66.1%) followed by Cladophialophora species (25.1%) and only in 2 cases were positive for Exophiala species.[2] To the best of our knowledge six cases of chromoblastomycosis caused by *Exophiala janselmei* are reported to date.

At present, there are no standard guidelines regarding the treatment of Chromoblastomycosis. Due to its resistant nature, particularly in severe cases, combination therapy with oral antifungals is recommended. Good response has been reported with a combination of Itraconazole and Terbinafine because of their synergistic effects[5]. Gupta *et al* reported four patients, initially with poor response to monotherapy treated with either combination or alternate week of Itraconazole and Terbinafine. Significant improvement was noted with no abnormality in lab parameters.[6] Our patient also showed excellent response with combination therapy of Itraconazole and Terbinafine.

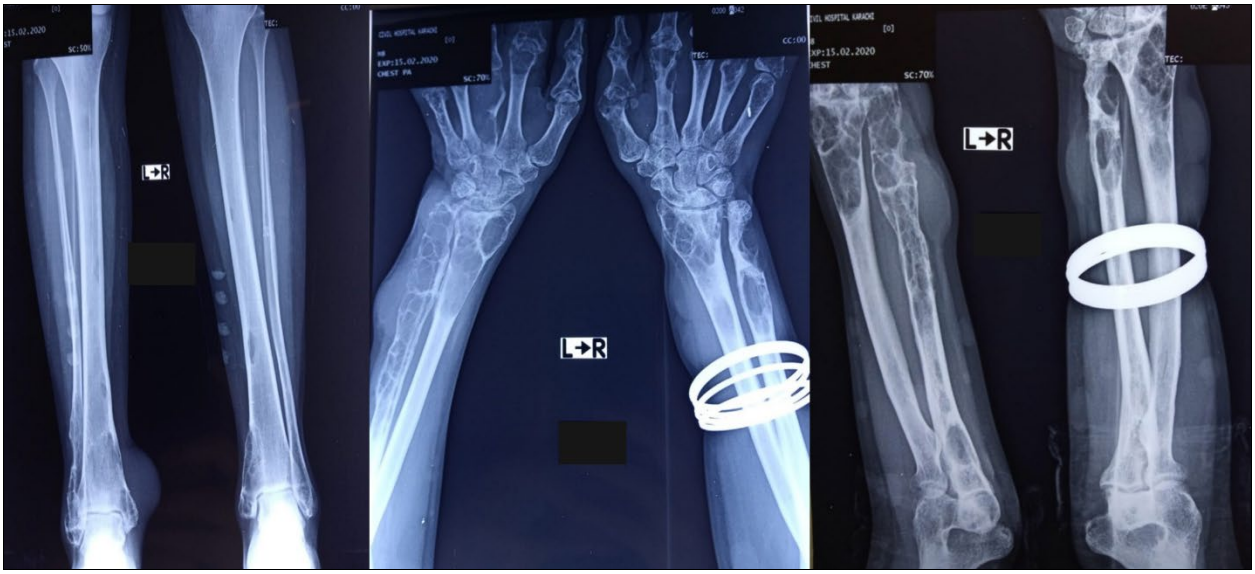
This case highlights an unusual presentation of Chromoblastomycosis caused by *Exophiala spp*., as well as good treatment response with combination therapy. We emphasize having a low threshold of suspicion of deep fungal infections where susceptible individuals present with chronic skin lesions enabling early diagnosis, prompt treatment and avoidance of complications as well as disfigurement.



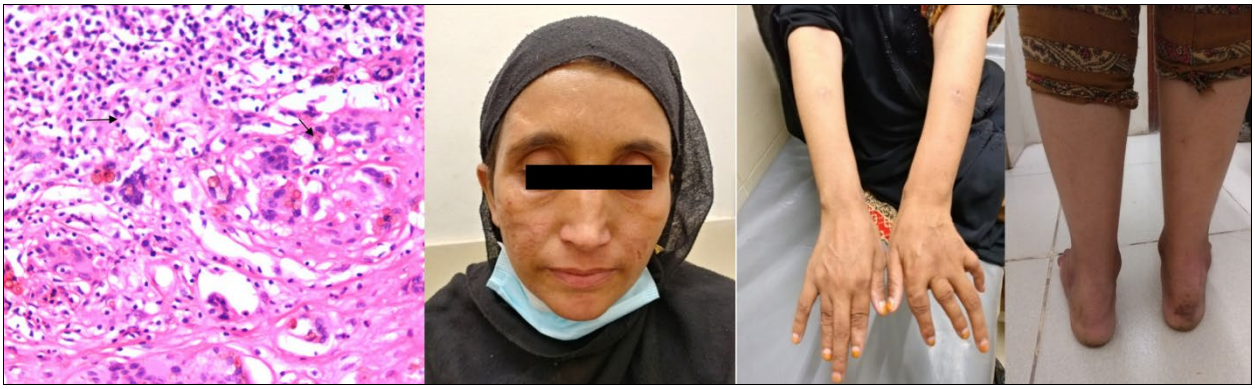
A: Initial Presentation-Multiple hyperpigmented indurated scaly plaques with central atrophy involving nose, cheeks and chin.



B: Initial Presentation - Bulbous swellings and gross deformity noted in both hands along with multiple skin-coloured swellings of variable sizes distributed over trunk and extremities.



C) Multiple well defined osteolytic lesions along with soft tissue densities are seen in small and long bones of upper and lower limbs.



D) Histopathology showing sheets of foamy histiocytes along with numerous multinucleated giant cells with engulfed pigmented fungal sclerotic bodies. E) At 6 months follow-up, there was marked improvement and visible clearance of her skin lesions.

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