CASE REPORT

A Rare Case of Chromoblastomycosis in a 12-year-old boy

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Abstract

Chromoblastomycosis is a chronic fungal infection of the subcutaneous tissue. The infection usually results from a traumatic injury and inoculation of the microorganism by a specific group of dematiaceous fungi, resulting in the formation of verrucous plaques. The fungi produce sclerotic or medlar bodies (also called muriform bodies or sclerotic cells) seen on direct microscopic examination of skin smears. The disease is often found in adults due to trauma. We report a case of chromoblastomycosis in a 12-year-old child in whom the infection started when he was only 4 years old with secondary involvement of bones, cartilage, tongue and palatine tonsils. The child was not immunosuppressed.

Keywords: Chromoblastomycosis, dematiaceous fungi, verrucous lesions, medlar bodies, culture.

Introduction

Chromoblastomycosis is a subcutaneous mycosis caused by dematiaceous species of fungi, most common of them being Fonsecaea pedrosoi, Phialophora verrucosa, Fonsecaea compacta and Cladophialophora carrionii[1] and less common being Exophiala spinifera, Aureobasidium pollulans and Chaetomium funicola.[2] The infection starts after traumatic inoculation of the fungus usually on the exposed surfaces of the body such as feet, legs, hands, arms and rarely oral mucosa.[2,3] Most of these cases are reported among young agriculture workers and individuals walking bare feet, commonly in tropical and subtropical areas.[4] The initial lesion is in the form of a tiny papule on the site of inoculation and slowly progresses to form verrucous, plaque-like, nodular or cicatricle lesions.[5] Diagnosis is based on direct microscopic examination of skin smears, culture and histopathology. Despite a variety of treatment modalities, which include long courses of antifungals, surgical excision and destructive physical therapies, the disease is difficult to eradicate once progressed.

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Case

A 12-year-old boy from the northern area of Pakistan presented to Department of Dermatology at Khyber Teaching Hospital (MTI-KTH) Peshawar in April of 2018 with a 7 to 8-year history of pruritic, dark, verrucous plaques extensively involving all parts of the body. He had a vague history of exposure to a thorn-prick on the right cheek which grew from a small papular lesion into a plaque and spread to involve other parts of the body over the course of years. He was taken to multiple doctors with no satisfactory results. In addition, he also complained of a sore throat, low-grade fever, joint tenderness and swelling on his current presentation.

On examination, there were several hyperpigmented, raised, verrucous plaques on almost all parts of the body with crusting and phagedenic ulcerations on lesions involving the face, arms, legs, fourth toe of the right foot and ring finger of the left hand [Figure 1]. There was oozing from lesions on the face and extremities. On general physical examination, the inguinal and cervical lymph nodes were enlarged and tender. He was vitally stable except for low grade fever. There was swelling of the distal interphalangeal joint (DIP) of the right little finger which was tender on palpation and had mild restriction in movement. The fifth metatarsophalangeal joint of the right foot was also swollen and tender [Figure 2]. Furthermore, there was tenderness in the Achilles tendons bilaterally. Verrucous lesions involved both the ear pinnae

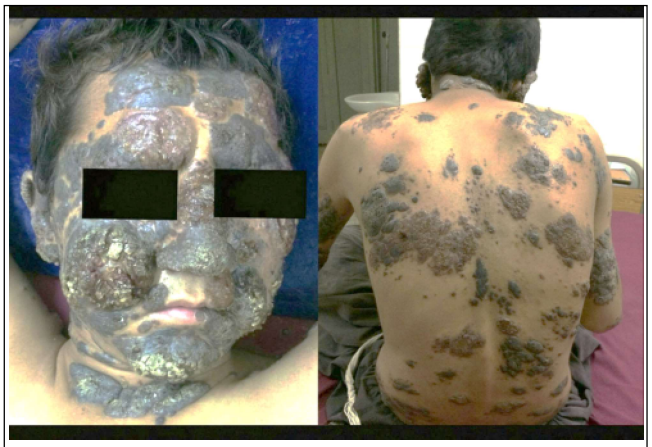

Figure-1: The face and trunk showing dark, verrucous, hyperkeratotic plaques.

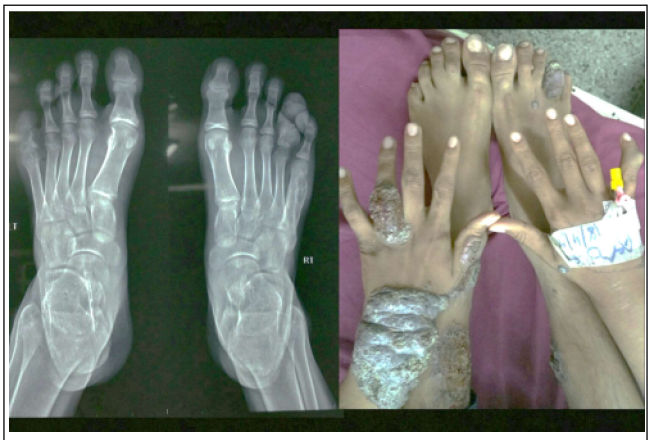

Figure-2: Image and X-ray showing the skeletal involvement in the patient. Notice the periosteal reaction and lytic resorption of phalanges of right fourth toe and left fifth toe, and right fifth metatarsal. Similar process was taking place in the fifth digit of the right hand.

as well but the left pinna was relatively deformed by the destructive nature of the lesions. He had history of a recent maggot infestation of the left external ear that was treated surgically. On examination of oral cavity the tip of the tongue showed a 2x2 cm flat irregular lesion on the ventral surface which was blue to purple in colour with a brown center. The right palatine tonsil had an irregular bluish-purple outgrowth as well and the throat was congested. He only complained of mild difficulty in swallowing.

His baseline investigations were within normal ranges but the Erythrocyte Sedimentation Rate (ESR) was raised. Urine and stool examinations were also insignificant. Viral profile for HBsAg, Anti-HCV and Anti-HIV was negative. Smear for LD bodies and skin PPD test were negative. X-ray of the hands and feet showed expansile, multiloculated lytic lesions and periosteal reaction involving the terminal phalanx of the right little finger and the right fifth metatarsal. There was narrowing of the joint spaces of the digits involved. There was partial resolution of the phalanges of the left fifth toe and complete resolution of the phalanges of the right fourth toe [Figure 2]. However, no sign of shortening of the toes and no restriction of movement was present physically. Chest X-ray was unremarkable. Computed tomography of the head and Echocardiography both were normal. Ultrasound of the abdomen showed no organomegaly or lymphadenopathy.

Serum uric acid level was slightly below the normal range. Pus culture and sensitivity from oozing lesions showed growth of MRSA on two occasions. Culture and sensitivity of the joint aspirate was planned but was not performed upon refusal by the patient and his parents.

KOH mount smear of the skin scrapings showed clusters of pathognomonic round brown cells or medlar bodies (also called muriform bodies or copper penny bodies) in the background of densely packed long slender septate hyphae suggestive of chromoblastomycosis.

Fungal culture on sabouraud dextrose agar showed growth of pigmented velvety dark colony with lighter centre and a darker periphery. Microscopic examination of the colony revealed cylindrical hyphae with septations and terminal conidiophores suggestive of Fonsecaea pedrosoi species.

Histopathology of the skin lesions showed pseudoepitheliomatous hyperplasia, dermal oedema, and several well-formed neutrophilic granulomas. PAS stain of the section showed numerous sclerotic cells.

The patient had been on multiple treatment regimens including oral Itraconazole, Voriconazole, Terbinafine, intravenous Amphotericin and antibacterials such as Amoxicillin, Ceftriaxone, Linezolid and topical Fusidic acid on previous visits and admissions. On current admission he was given oral Itraconazole 200 mg BD, oral Ciprofloxacin 500 mg BD, IV Amoxicillin Clavulanate 1.2 g BD, Nystatin oral drops TDS and topical Fusidic acid. Oral analgesics were given for his joint and tendon pains. Cryotherapy was performed on some lesions. Within initial days of his treatment the oozing in the lesions stopped, but recurred after 3 to 4 days and was then put on oral Linezolid 600 mg BD on discharge. On follow up the patient showed little signs of improvement of his condition with the same oozing lesions that were present on initial presentation.

Discussion

From our case study we report that Chromoblastomycosis is a subcutaneous fungal infection having the potential to infiltrate into deeper tissues. It is caused by an infection with dematiaceous species of fungi by traumatic inoculation on the exposed areas of the body, found mostly among agriculture workers and often among children and adults who walk barefoot and live in the tropical and subtropical areas of the country.[6] Our young patient's infection started when he was only 4 years old. Chromoblastomycosis usually starts with a small papule at the point of inoculation and progresses to verrucous, hypertrophic, warty plaques with crusting, scaling, scarring and ulcerations as was evident in our case. Secondary bacterial infections are common and

most probably due to scratching of the lesions. He had inguinal and cervical lymphadenopathy and a lesion on the tongue and right palatine tonsil that needed further workup.

Few cases of chromoblastomycosis with skeletal and oral cavity involvement have been reported. Javaid et al report unique skeletal features of a case of chromoblastomycosis. Physical findings include swelling and shortening of single or multiple digits of involved hands and feet. Radiographic findings include bone lesions undergoing lytic process resulting in partial or complete resorption and telescoping of the phalanges involved.[2] The skeletal involvement in our case showed tender swelling of the middle phalanx and DIP joint of the right little finger without an overlying skin lesion. A similar process but with an overlying skin lesion obscuring it was noted on the left ring finger and the fifth toe of the right foot. Plain radiographs of these lesions showed expansile, multiloculated lytic lesions and periosteal reaction of the bones and joints involved. There was narrowing joint space and partial and/or complete resolution of multiple tarsal and metatarsal bones and phalanges of the feet and hands involved. Pinnae of both the ears were disfigured by the lesions as well. The bone, enthesis and cartilage involvement is unique and depicts the ability of the lesions to involve deeper structures. Due to the destructive nature of the involvement, debridement and amputation of the affected bones and joints was suggested. Further workup on the bone and joint involvement was not done as it was refused by the patient.

Oral involvement in chromoblastomycosis has been reported to be either secondary to cutaneous lesions or isolated oral lesions in the presence or absence of immunosuppressive state. Characteristics of the oral lesions range from large thick whitish plaques to small pink warty lesions on the lips, gingivae, palate or tongue.[4,8] Our case had large bluish to purple plaque with brownish centre on ventral surface of the tongue and a bluish warty outgrowth on the right palatine tonsil, both of which were noted later in the course of the disease and showed minimal response to treatment similar to the skin lesions.

Diagnosis of Chromoblastomycosis is made by observing muriform cells called sclerotic bodies, copper penny or Medlar bodies on biopsy and KOH mount smear, and identification of the fungal agent in culture medium.[4,5]

Treatment of Chromoblastomycosis is associated with low cure rates and high rate of relapse.[5] Our case had been on combination regimens ever since his diagnosis, including Itraconazole, Voriconazole, Terbinafine and Amphotericin with less fruitful results. The patient was resistant to all kinds of treatment and radiotherapy was being discussed as a last yet experimental option. Symptomatic treatment appears to be the only management in such resistant cases. Due to its chronic nature, the lesions of Chromoblastomycosis may undergo neoplastic change into skin cancer later in the course of the disease.[7]

Conclusion

Chromoblastomycosis is one of the most difficult fungal infections to eradicate. It could be due to the presence of therapy-refractive medlar bodies and differential susceptibilities between taxonomically closed related groups. Early intervention is necessary for response to treatment. Other treatment modalities should be studied for cases at an advance stage of the infection and showing resistance to conventional therapy.

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