Primary Cutaneous Coccidioidomycosis: Clinical Case Report and Discussion
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Citation

Abstract
Coccidiomycosis is a rare fungal infection which usually affects only the lungs. Involvement of the skin by the causative fungus is very rare. Depending on the organ of primary infection, cutaneous involvement can be sub grouped into two types a) primary and b) secondary. Primary cutaneous infection is extremely rare and usually occurs when Coccidioides inoculates traumatized skin. In this article we report a case of the primary cutaneous coccidiomycosis of the nose. To our knowledge this is only second reported case of the primary infection of the skin of the nose by Coccidioides and first reported case of primary cutaneous coccidiomycosis of the nasal alae. (1)

CASE REPORT
A 36 year old male with no significant past medical or surgical history, resident of San Joaquin County, presented with a non healing slowly enlarging growth on the skin of the nose for two months. On asking further questions, we found that the patient had a trauma when he was stuck by a paint ball on the face followed by the appearance of the lesion five days later. Examination showed a 2x2 cm plaque with central crustation on the skin of the left nostril. (Fig1)

FIGURE 1
Figure 1: Lesion of Primary Cutaneous Coccidioidomycosis on the left nostril.

Biopsy of the lesion showed Coccidioides spherules. The diagnosis of Primary Cutaneous Coccidioidomycosis was made and patient was treated with Fluconazole for three years. The treatment resulted in complete resolution of the lesion (Fig 2).

DISCUSSION
Coccidiomycosis has been recognized as pathogenic to humans for several decades. Its causative agent, Coccidioides immitis, is endemic in areas with a more avid climate. Since deserts of southwestern United States, Mexico, South and Central America are well known for warm weather, they are the areas of its high prevalence. Usual mode of transmission is by inhalation during which infectious particles (arthroconidia) are transferred from the soil to the human's lungs. Thus, lungs are usually the primary suffering organs. (2)
The sign and symptoms of Coccidioides infection are usually mild and in two thirds of immuno-competent humans the disease usually subsides without even coming to the attention of the physicians. Some patients present as upper respiratory infection which is usually self limited. In these individuals, positive skin test is the only means to diagnose past sub-clinical infection.

Clinically, symptomatic infection of Coccidioides mimics bacterial pneumonia. The usual presentation include (1) high grade fever, chills, chest pain, productive cough rarely accompanied with blood, sweating, anorexia, arthralgias, fatigue and weakness. Examination usually reveals nonspecific wheezes, rubs and decreased breath sounds. Immunological response of the skin including erythema nodusum, erythema multiforme and toxic erythema can accompany such presentation. If appropriately treated, most of these cases usually resolve in few months without any complications.

One percent of primary pulmonary coccidiomycosis ends up in dissemination. Such involvement of other organs is known as secondary infection. This complication is more common in infants, men, pregnant women, immuno-compromised patients (with thymectomy, corticosteroid therapy, hepatitis C patients) (4) and surprisingly in patients of non Caucasian ethnicity. The common sites of metastasis include skin, meninges, bones and joints. Disseminated infection involving skin usually presents as nodules, verrucous plaques, plaques with central atrophy, papules and pustules. Rarely involvement can mimic keloids (5) or lesions mimicking mycosis fungoides. (6)

Primary cutaneous infection of the Coccidioides is rare and to our knowledge less than 25 cases have been reported since first case that was published in 1920s. None of these include involvement of the nasal ala, although one case involving the tip of the nose has been reported.

The diagnostic criteria (7) for primary cutaneous Coccidiomycosis described by Wilson et al include the following:

1. No preceding pulmonary infection
2. history of break in the skin
3. a short incubation period
4. lesion resembling a chancre
5. positive precipitin reaction to the coccidioidin with slow decline
6. positive skin test for coccidioidin
7. presence of regional lymphadenopathy and lymphangitis only in the region of drainage of the skin(can be completely absent
8. The compliment fixation test reaction is initially negative and low titers may be measured for several weeks.
9. Spontaneous healing

Grossly the primary cutaneous lesion of Coccidioidomycosis mimics the primary cutaneous lesion of other granulomatous disease such as syphilis and tuberculosis. But other lesions such as non healing ulcers and verrucous nodules are also not uncommon. The resolution of these lesions is usually followed by life-long immunity in almost all the individuals. (7)

Diagnosis of the primary cutaneous variety is usually made by combination of above mentioned diagnostic criteria, microscopic demonstration of Coccidioides immitis spherules and negative chest xray. Presence of only one of the above mentioned criteria is not sufficient to confirm diagnosis of Primary Cutaneous Coccidiomycosis.

Treatment options for Coccidiomycosis include Amphotericin B and azoles, with later far less toxic than former. Data suggests that the azoles therapy should be continued for at least six months after the disease has become inactive. Due to rare number of patients with primary cutaneous infection, the data on the efficacious treatment is scarce but continuation of azoles therapy at least for six months after the disease has become inactive is usually recommended. We treated our patient with fluconazole for three years followed by complete resolution of the nose lesion (Fig. 2)

In conclusion, (9) although Primary Cutaneous Coccidiomycosis is a rare entity it should be considered in immuno-competent patients with long standing skin lesions appearing secondary to skin trauma. Threshold to biopsy these lesions should be kept very low especially in regions of high prevalence.

References
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