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CASE REPORT

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KEYWORDS

Paracoccidioidomycosis; Paracoccidioides brasiliensis; Pulmonary fibrosis; High-resolution computed tomography **Abstract** Paracoccidioidomycosis is a systemic mycosis which is endemic in rural areas of Latin America, an important European source of immigrants and a growing European touristic destination as well, with most cases occurring in Brazil, Argentina, Venezuela and Colombia. The authors report a case of a 43-year-old man who previously worked in Venezuela and is living in Portugal for 8 years, presenting with a single cutaneous lesion. Despite the absence of valuable respiratory complaints, severe lung damage was found with high-resolution computed tomography (HRCT). Biopsy of the cutaneous lesion and mycologic sputum examination were performed revealing *Paracoccidioides brasiliensis* infection.

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PALAVRAS-CHAVE

Paracoccidioidomicose; Paracoccidioides brasiliensis; Fibrose pulmonar; Tomografia computorizada de alta resolução

Paracoccidioidomicose pulmonar: relato de caso clínico com alterações na tomografia computorizada de alta resolução

Resumo A Paracoccidioidomicose é uma micose sistémica endémica nas áreas rurais da América Latina, uma fonte importante de imigrantes e destino de emigração e turismo europeu, a maioria dos casos ocorrendo no Brasil, Argentina, Venezuela e Colômbia. Os autores descrevem o caso clínico de um paciente com 43 anos, anteriormente emigrado na Venezuela e residente em Portugal há 8 anos, que se apresenta com lesão cutânea isolada. Embora sem queixas relevantes do foro respiratório, apresentava extensas lesões do parênquima pulmonar caracterizadas por tomografia computorizada de alta resolução (TCAR). Foi realizada biópsia da lesão cutânea e exame micológico da expetoração que revelaram infeção por *Paracoccidioides brasiliensis* (PB).

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Introduction

Paracoccidioidomycosis (PCM) is the most common endemic systemic mycosis in Latin America. It occurs in about 10% of the population in the subtropical regions of Brazil,¹ and mostly affects farm workers; the highest incidence happens between the ages of 25 and 60.² The causing agent, Paracoccidioides brasiliensis (PB), is a dimorphic fungus that can remain viable in the host for long periods, maintaining the potential for disease reactivation for several years after the initial infection. The portal of entry is the respiratory tract, the lung being the organ which is most frequently affected.¹ Chronic infection with severe lung damage and progression to end-stage fibrosis occurs even where there are no obvious symptoms. Spread from a primary pulmonary lesion may affect other organs, not only the skin and mucous membranes most often, but also the adrenal glands, kidneys, gastrointestinal tract, liver, spleen and central nervous system.^{3,4} The most common sequelae include fibrosis with respiratory failure, cor pulmonale and also Addison's disease and intestinal malabsorption.² The authors present a case of PCM with pulmonary and cutaneous involvement.

Case report

A 43-year-old Portuguese male, former farm worker in Venezuela until 2001, presented with a single back ulcerated lesion, with a 2-month evolution (Fig. 1). He had no other complaints and his clinical background was unremarkable, except for a previous ulcerated lesion of the nasal mucosa in 1996, for which he had had an unspecified 6-month treatment. His general condition was normal, without fever, palpable lymphadenopathy or hepatosplenomegaly. There were no laboratory changes, results for HIV 1 and 2 were negative. Bilateral diffuse changes in pulmonary auscultation were noticed and a chest radiograph was performed, showing a bilateral reticulo-nodular pattern predominantly in the



Figure 1 Back ulcerated lesion, with a 2-month evolution.

middle and upper lung zones and a segmental consolidation in the right lung base.

Pulmonary HRCT revealed exuberant bilateral parenchymal lesions with fibrotic changes and areas of active inflammatory disease mostly affecting the middle and upper lobes, despite the absence of respiratory symptoms. Multiple irregular spiculated nodular lesions ranging from 8 to 25 mm in size were identified, some with central cavitation and confluence; these were most conspicuous in the upper lobes. Other findings included a central area of consolidation with air bronchogram in the middle lobe, architectural distortion, scattered foci of 'ground glass' attenuation, septal thickening, parenchymal bands, diffuse pleural speculated thickening, apical emphysematous bubbles and traction bronchiectasis. Hilar adenopathy and parietal irregularity of a dilated trachea were also present (Fig. 2).

Biopsy of skin lesions and mycological examination of sputum revealed infection by *Paracoccidioides brasiliensis* (PB).

Therapy with itraconazole (200 mg/day for 2 months, followed by 100 mg/day for 8 months) was performed, with favorable clinical course.

Discussion

PCM was initially described by a Brazilian doctor, Lutz, in 1908.² It is the most common endemic mycosis in Latin America. Brazil has the highest number of cases; it is most prevalent in subtropical regions where it affects about 10% of the population.^{1,2}

It is a systemic granulomatous disease, predominantly pulmonary and mucocutaneous. The lungs are involved in 50-100% of cases.¹ The main route of infection is by inhalation of *PB* spores which are present in the soil.^{1,2,4,7,8} The host's initial contact with the fungus usually progresses to a subclinical infection, with formation of a pulmonary granuloma, which may resemble the primary complex of tuberculosis.^{1,2}

In most individuals the natural defense mechanisms provide a balance between the host and the agent, with the fungus remaining viable in a latent form.^{1,3,4,6} Spread to other organs and tissues may occur, most often to the skin and the mucosa of the airway and the oral cavity, with formation of granulomatous hemorrhagic ulcerated lesions.⁵

Two clinical forms have been described:

- an acute/subacute or juvenile type, accounting for less than 10% of cases, affecting both sexes under the age of 25 years, with fever, weight loss and malaise. There is rapid and progressive organ involvement with diffuse superficial and deep lymphadenopathy and hepatosplenomegaly. In a small number of cases cutaneous and osteolytic lesions coexist. The small intestine is involved in about 50% of cases. There is rarely involvement of the lung and bone marrow. The most common complications are lymphatic obstruction, intestinal malabsorption or protein-losing enteropathy.^{2,4}
- a chronic form that arises primarily in adult males (male / female ratio of 10:1-25:1), 25-60 years old, most often involving the lung, followed by skin and mucous membranes. Patients may be asymptomatic or present with

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