A NEWLY DIAGNOSED WEGENER’S DISEASE AS THE UNDERLYING CAUSE FOR A DISSEMINATED COCCIDIOIDOMYCOSIS

Ide L¹, Thibo P²
¹AZ Jan Palfijn Ziekenhuis AV, Gent, Department of Microbiology, ²AZ Jan Palfijn Ziekenhuis AV, Gent, Department of Internal Medicine, Belgium

Correspondence and offprint requests to: Louis Ide, E-mail: louis.ide@janpalfijngent.be

ABSTRACT

In most cases coccidiodomycosis presents as a benign mildly severe respiratory disease with a benign course and spontaneous resolution. Rarely dissemination can lead to complications. We believe this is the first published case of a patient with a disseminated coccidiodomycosis, as shown on a urinary sample, in association with Wegener’s disease. It was a challenge to diagnose and consecutively treat the patient as therapies seem to be conflictual.

This case illustrates how migration, changing habits and attitudes, travelling, changing geo-ecological circumstances can lead to a change in medical environment.

It is therefore essential that the microbiologist becomes a clinical microbiologist who communicates intensively with his fellow clinicians.

Key words: coccidiodomycosis, Wegener, travel, environmental, disseminated

INTRODUCTION

Coccidiodomycosis is not endemic in Europe. But cases of coccidiodomycosis, also known as ‘valley fever’ occur in patients who lived or travelled in endemic areas. Medical environment changes. Usually coccidiodomycosis is a self-limiting, mild disease. Rarely Coccidioides immitis and Coccidioides posadasii cause dissemination. Preferential locations of dissemination are the central nervous system, the bones, lymph nodes and skin. Involvement of the urinary tract as in this case is rather rare. It should alert us to look for a reason behind the dissemination. What’s the patient’s immune status? Finally when coccidioidomycosis is suspected the mycology lab should be informed to prohibit accidental exposure as the athroconidia are quite contagious.

CASE REPORT

A 23-year old Caucasian male who was born and grew up in the USA resided for more than 1 year in Belgium at the time of his current illness. About 3 months before admission he went on holiday to his native country, driving a quad in the woods of Prescott, Arizona. From that time on he complained of nasal congestion, nose bleeding, xerostomia and sinusitis. Initially his general practitioner referred him to our hospital to exclude Sjögren syndrome. He returned to our hospital because of a persistent cough with haemoptysis. He had been suffering a substantial loss of weight (about 10 kilograms) and complained of diffuse arthralgias, especially in the legs. He felt feverish. On clinical examination only the presence of a cervical lymph node was noticed.

Radiography of the lungs, followed by a CT scan of the thorax showed diffuse infiltrates with ground-glass opacities (Figure 1). Blood results showed eosinophilia (16%) and impaired renal function (creatinine 2.38 mg/dL (normal range 0.66-1.25 mg/dL)), ureum 93 mg/dL (normal range 20-45 mg/dL)). Hematuria was present (12447 red blood cells (RBC)/µL (normal value: < 25 RBC/µL)) as well as proteinuria (3.5 g/L (normal range 0-0.150 g/L)). Samples of sputum and urine were taken for direct microscopic examination and culture. Tuberculosis was excluded. Direct examination of both sputum and urine revealed the presence of cystic elements. Spherules of various sizes (10 to 100 µm) with multiple endospores (2 to 5 µm) which are characteristic of coccidioidomycosis were presented in both samples (Figure 2). The walls of some of the spherules appear to be disrupted, with endospores spreading into the surrounding tissues. Culture revealed the typical barrel-shaped arthro-aleurioconidia (Figure 3).
A newly diagnosed Wegener’s disease as the underlying cause for a disseminated coccidioidomycosis

All possible causes of immunodeficiency such as HIV, drug abuse, etc. were considered as normally coccidioidomycosis is a benign, rapidly resolving disease. Eventually the history of renal impairment with hematuria and the pulmonary involvement did ring a bell: c-ANCA's were positive. PR3-ANCA was positive (62 U/mL, normal value < 7 U/mL) and MPO-ANCA was negative (0.0 U/mL, normal value < 7 U/mL) compatible with Wegener’s granulomatosis.

A renal biopsy revealed diffuse crescentic glomerulonephritis. All these findings confirmed the diagnosis of Wegener’s disease as the underlying cause for the disseminated coccidioidomycosis.

In view of the decline in renal function, the patient was initially treated with liposomal amphotericin B (AmBisome®). Corticosteroids were started in combination with ciclofosfamide as soon as the diagnosis of Wegener’s disease was established. After a few weeks, with improvement of renal function, amphotericin B was replaced by fluconazole. At present the patient is doing well under therapy with steroids and fluconazole (400 mg/day).

DISCUSSION

Coccidioidomycosis usually presents as a subclinical or mildly severe upper respiratory infection that rapidly resolves (1). About 60% of the exposed individuals are asymptomatic. Coccidioides immitis and Coccidioides posadasii are the possible etiological agents of coccidioidomycosis, with only small differences in morphology as well as in clinical course.

Rarely the disease evolves into a severe acute or chronic disseminated mycosis, which eventually can be fatal. This kind of evolution should draw our attention towards the presence of an underlying medical condition leading to immunodeficiency. We have to ask ourselves ‘what’s wrong with the patient’?

Isolation of the causative organism from urine is rare. The first description of a positive urinary sample comes from Goldman et al. (1948) in a 27-year-old negro male (2). The patient died eventually. Since then only few cases were published. Nevertheless all published case reports warn not to ignore coccidioidomycosis (3).

Chen et al. conducted a survey to assess healthcare providers’ knowledge, attitudes, and practices regarding coccidioidomycosis diagnosis and treatment in Arizona, an endemic area. They concluded that even in endemic areas there is a need for a comprehensive coccidioidomycosis education campaign for health care workers (4). On June 29th 2012 ProMED-mail published data which indicate a significant increase in cases of valley fever. GIDEON provided some epidemiology through ProMED-mail. During the last few years the number of cases in de USA is considerably growing (5). According to the records of ProMED-mail only few import cases were registered (6).

This case illustrates that environmental changes may have a huge impact on day-to-day practice of the microbiologist. Premature exclusion of diseases thought never to occur in a certain area, can lead to misdiagnosis and maltreatment. Migration, changing habits and attitudes, travelling, changing geo-ecological circumstances, all can lead to a changing medical environment. It is therefore essential that the
microbiologist becomes a clinical microbiologist who communicates intensively with his fellow clinicians (5).

CONFLICT OF INTEREST: None.

REFERENCES


