

MEDIASTINAL AND PULMONARY ENTOMOPHTHOROMYCOSIS WITH SUPERIOR VENA CAVA SYNDROME: CASE REPORT

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SUMMARY

The first case of mediastinal and pulmonary entomophthoromycosis with superior vena cava syndrome is reported. The patient presented with a history of edema of the face, neck and upper limbs as well as collateral circulation in the anterior wall of the chest. Histological examination of tissue from mediastinum revealed a granulomatous reaction with microabscesses surrounded by eosinophilic amorphous material and with broad hyphae in the center. Culture was not performed because a preliminary diagnosis of non Hodgkin's malignant lymphoma was made. Surgical correction of the obstructed area was performed and the patient was successfully treated with potassium iodide. The authors propose that mediastinal entomophthoromycosis must be considered in the differential diagnosis of diseases causing superior vena cava syndrome in tropical and sub-tropical regions. This case enlarges the spectrum of clinical manifestations of the zigomycosis caused by **Entomophthoraceae**.

KEY WORDS: Mediastinal and pulmonary entomophthoromycosis; Superior vena cava syndrome.

INTRODUCTION

Zygomycosis is a collective designation of the mycotic infections caused by several species of **Zygomycetes**, orders **Mucorales** and **Entomophthorales**^{3, 6, 9}. Zygomycosis is characterized by the presence of broad (5-20 μ m in width), occasionally septate, and aberrantly ramified hyphae in the host's tissue^{1, 2, 4}. It is possible to distinguish the hyphae of **Mucorales** from those of the **Entomophthorales** in tissue sections stained by HE: the hyphae of **Mucorales** are clearly seen

because they have a well stained cell wall; those of **Entomophthorales** are sleeved by eosinophilic material (Splendore-Hoeppli phenomenon)^{3, 6, 9}.

Zygomycosis caused by species of **Entomophthorales** has a chronic course and affects apparently healthy hosts in tropical or subtropical regions. Its subcutaneous form^{7, 9} caused by **Basidiobolus ranarum** (= haptosporus) or

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Conidiobolus coronatus is more frequent. However cases of disseminated or primary localized visceral involvement have also been reported^{8, 10}. Exclusive thoracic involvement was described only once¹⁰. The purpose of this paper is to report a case of zygomycosis caused by an *Entomophthoraceae* involving the lung and the mediastinum and causing a superior vena cava syndrome (SVCS).

CASE REPORT

A 29 years-old woman from Salvador, Bahia, Brazil, was admitted in July 1984 with a recent history of edema of the face, neck and upper limbs as well as collateral circulation in the anterior wall of the chest. Urinalysis, hemogram, blood sugar, urea and nitrogen were within normal limits. Electrocardiogram showed no abnormalities. An X-ray of the chest and a computerized tomography revealed a small nodule on the upper lobe of the right lung and an enlargement of the middle and superior mediastinum, especially near the azigos vein (Fig. 1). The cavography revealed invasion and obstruction of the superior vena cava with evidence of collateral circulation (Fig. 2). A clinical diagnosis of non-Hodgkin's malignant lymphoma with SVCS was made. Exploratory thoracotomy showed a granular and friable tissue involving the wall of the superior vena cava and other mediastinal veins. A biopsy was obtained and surgical correction of the obstructed area performed.



Fig. 1 — Computerized tomography showing small nodule in the right hemithorax.

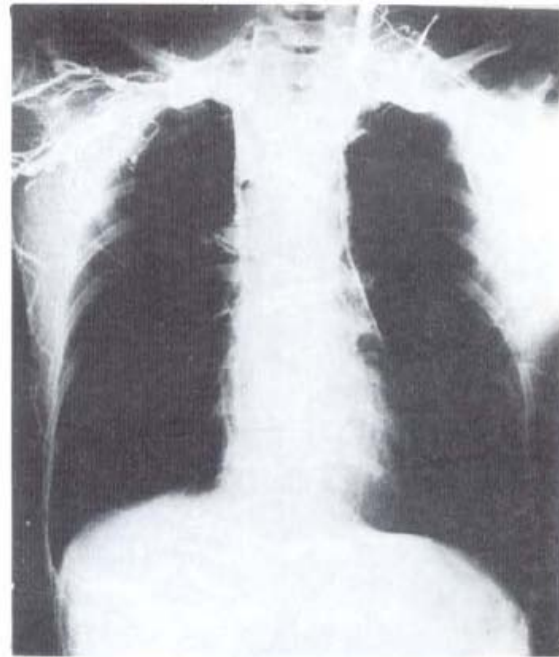


Fig. 2 — Cavography with obstruction of the superior vena cava and showing collateral circulation.

PATHOLOGIC FINDINGS

Histological sections stained by HE revealed a chronic granulomatous reaction. Small abscesses with neutrophils and eosinophils surrounded by histiocytes and multinucleated giant cells were also observed. Broad hyphae (10 μ m in diameter) were seen inside the microabscesses. The hyphae, occasionally septate were sleeved by and eosinophilic material (Splendore-Hoeppli phenomenon) (Fig. 4). This reaction was also well observed in P. A. S. stained sections. The hyphae were well impregnated by silver (Grocott's method) (Fig. 5).

Following histopathological diagnosis immunological evaluation was made. Intradermal skin tests with PPD, Candidin and Trichophitin and the determination of population and sub populations of lymphocytes showed no abnormalities (Lymphocytes T = 69%; OKTA = 50%; OKT8 = 19%; lymphocytes B = 15%; mixed lymphocytes = 16%). The patient was successfully treated with potassium iodide (20 mg/kg/day during 6 months). No relapse was observed in a 20 months follow-up.



Fig. 3 — Hyphae coated with eosinophilic material (Splendore-Hoeppli phenomenon) HE (X 400).



Fig. 4 — Hyphae impregnated by silver (Grocott's method) HE (X 400).

DISCUSSION

This case represents the second of primary intrathoracic Entomophthoromycosis and the first manifested as SVCS. The first was described by ECKERT¹⁰ and was caused by *C. incongruus*. In the case of BUSAPAKUM et al⁸, there were lesions in the lungs, liver, digestive tract and skin,

but there is doubt if the lungs were the primary site of disease. Another case of Entomophthoromycosis with pulmonary involvement was secondary to a skin lesion in the chest⁴. Although, the etiological agent was not isolated in the present case the histopathological aspects permit the diagnosis of Entomophthoromycosis, due to the presence of a granulomatous process with an eosinophilic infiltrate and the Hoeppli-Splendore phenomenon. Mucormycosis is also a zygomycosis that causes visceral lesions but it manifests itself with a different histopathological pattern^{5, 12}; moreover, it occurs only in immunosuppressed patients¹².

The SVCS results usually from the involvement of the mediastinum by malignant pulmonary tumors, lymphomas and, less frequently, by inflammatory processes like tuberculosis and histoplasmosis¹¹. In the case reported the mycotic origin of the syndrome was strongly suggested by its disappearance with the specific treatment. Certainly, the pulmonary lesion was primary, with posterior propagation to the mediastinum.

We propose that mediastinal Entomophthoromycosis must be considered in the differential diagnosis of diseases causing SVCS in tropical and subtropical regions. On the other hand, this case enlarges the spectrum of clinical manifestation of the zygomycosis caused by *Entomophthoraceae*.

RESUMO

Entomofitose mediastinal e pulmonar com síndrome de veia cava superior: registro de caso.

O primeiro caso de entomofitose mediastinal e pulmonar com síndrome de veia cava superior, é descrito. A paciente apresentou-se com história de edema facial, cervical e de membros superiores, bem como circulação colateral na parede anterior do tórax. O estudo histopatológico do tecido do mediastino, próximo à veia cava, revelou reação granulomatosa com microabscessos, circundados por material amorfo, eosinofílico e com hifas largas no centro. Cultura não foi realizada porque o diagnóstico clínico foi de doença de Hodgkin ou de um linfoma não-

Hodgkin. Correção cirúrgica da área obstruída foi realizada e a paciente tratada com iodeto de potássio, nas doses preconizadas, obtendo-se sucesso. Os autores propõem que a entomofotoromicose mediastinal deva ser considerada no diagnóstico diferencial de doença causando síndrome de veia cava superior em regiões tropicais e sub-tropicais. Este caso aumenta o espectrum de manifestações clínicas das zigomicoses causadas por *Entomophthoraceae*.

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