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Gilchris Disease of the Central Nervous System Mimicking Malignant Brain Neoplasm: A Rare and Fatal Complication of Blastomyces Dermatitidis

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Abstract- Blastomycosis or "Gilchrist disease" is a deep mycosis due to a dimorphic fungus: "Blastomyces Dermatitidis." It is a telluric fungus of the wooded areas of the Eastern North American continent, causing chronic granulomatous diseases, responsible for pulmonary manifestations due to the inhalation of its spores. Dissemination can lead to skin, bone, visceral, and even neurological signs that can be fatal.

We report the case of a 50-year-old African man who has been complaining for five days before admission of headaches and behavioral disorders. On examination, he had a frontal syndrome with idiopathic slowdown without motor or sensory deficit. The rest of his check up was normal. Cerebral MRI showed basal left with intense enhancement. The post-operative evolution was marked by the absence of awakening and the patient's death. The pathological examination has concluded to cerebral blastomycosis.

Keywords: blastomycosis; abscess; central nervous system.

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Gilchris Disease of the Central Nervous System Mimicking Malignant Brain Neoplasm: A Rare and Fatal Complication of Blastomyces Dermatitidis

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Abstract- Blastomycosis or "Gilchrist disease" is a deep mycosis due to a dimorphic fungus: "Blastomyces Dermatitidis." It is a telluric fungus of the wooded areas of the Eastern North American continent, causing chronic granulomatous diseases, responsible for pulmonary manifestations due to the inhalation of its spores. Dissemination can lead to skin, bone, visceral, and even neurological signs that can be fatal.

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Extra-pulmonary blastomycosis remains a rare entity. The diagnosis is mainly based on examination of the CSF or the specimen after special staining. The mild or moderate forms are treated with itraconazole. In the case of severe, life-threatening infection, amphotericin B is required. The prognosis is unfortunately reserved for the price of several complications.

Keywords: blastomycosis; abscess; central nervous system.

I. INTRODUCTION

Blastomycosis or "Gilchrist disease" is a deep mycosis due to a dimorphic fungus: "Blastomyces Dermatitidis." It is a telluric fungus of the wooded areas of the Eastern North American continent, causing chronic granulomatous diseases, responsible for pulmonary manifestations due to the inhalation of its spores. Dissemination can lead to skin, bone, visceral, and even neurological signs that can be fatal.

II. PATIENT AND OBSERVATION

We report the case of a 50-year-old African man with no medical history working as shepherd, who has been complaining for five days before hospitalization of intensives headaches, behavioral disorders, and

dizziness on standing. The patient did not smoke, drink, or use recreational drugs. On examination, he had a frontal syndrome with idiopathic ideomotor slowdown without motor or sensory deficit. The rest of his check up was normal.

Cerebral MRI (Figure 1) showed a left retro-orbital non-hemorrhagic front-basal lesion in iso signal T1, and hypo signal on T2 weighted image taking contrast in an intense and homogeneous way with a hypo diffusion signal and marked surrounding vasogenic edema on T2 Flair weighted image.

The patient developed unfortunately rapidly a consciousness deterioration and underwent quick resection of his lesion, which was hard, rubbery, and pedunculated on macroscopic examination. Microscopic analysis of the specimen showed granulomatous inflammation with yeast-forming fungi, consistent with «B.D». The postoperative evolution was marked by the absence of awakening and his death.

Pathological examination (Figure 2) showed, after coloration with Grocott's methenamine silver method, large broad-base and unipolar budding yeast-like cells in favor of blastomycosis.

III. DISCUSSION

Blastomycosis is a fungal infection of humans and other animals, notably dogs and occasionally cats, caused by the organism «Blastomyces dermatitidis (BD)». Endemic to portions of North America, blastomycosis can cause clinical symptoms similar to histoplasmosis [1].

The optimal habitat for «Blastomyces» colonization is wetland enriched with animal droppings and decaying vegetation. Inhalation is the most common route of transmission, followed by cutaneous inoculation via direct penetration which is much less common. Our patient used to have this type of contact with animals as he was a shepherd. «BD» is generally not transmitted from person to person, and therefore, is not considered contagious [2].

Clinical presentation is uncommon and may vary from subclinical infection to rapidly progressive

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dissemination and death. Constitutional symptoms such as discomfort, weight loss, and fatigue can be obviously seen [2]. Our patient was presented just for neurological disorders without others abnormalities.

The likelihood of infection with «*BD*» is equal among immunocompetent and immunosuppressed individuals unlike other opportunistic infections [2]. However, in people with compromised cellular immunity, infection is not more aggressive [2]. Lymphatic and hematogenous spread may ensue to potentially any organ [3]. After the lungs, other sites of disease involvement include skin, bone, genitourinary system, and the CNS, in order of decreasing frequency [2].

The preferred imaging modality in patients with suspected CNS infection is contrast-enhanced MRI. On MRI, pyogenic bacterial parenchymal abscesses typically will have a robustly enhancing peripheral rim of tissue that correlates with the host response and capsule formation. Centrally, pyogenic abscesses will display reduced diffusion correlating with purulent material, which is typically composed of cellular debris and inflammatory cells such as polymorphonuclear cells of the immune system [4]. However, the case presented here indicates a total and homogeneous lesion enhancement without central reduced diffusion.

In fact, the imaging characteristics of solitary parenchymal CNS blastomycosis infection can be hard to differentiate from other fungal entities, and the differential should include histoplasmosis and coccidioidomycosis.

CSF analysis will usually show pleocytosis with a neutrophilic or lymphocytic predominance [2]. If there is a diagnostic concern for blastomycosis, the detection of the «*B.D*» antigen rather than culture is useful.

On histopathology, tissue specimens show the host's response to the infection as an inflammatory reaction of polymorphonuclear leukocytes in a cluster of granulomas, usually of the non-caseating type [2]. Budding yeast cells with capsules will be seen, staining positive with Grocott-methenamine silver (GMS). Acid-fast stains can help distinguish «*B.D*» from *C. immitis*, as it will usually be negative in the latter and weakly positive in the former [2].

Orally given itraconazole is the best choice for most forms of the disease. Ketoconazole can also be used. Cure rates are high, and the taken charge over months is usually well tolerated. Amphotericin B is considerably more toxic, and often reserved for immunocompromised patients who are seriously sick and those with central nervous system disease. Voriconazole has shown promise and continues to be used due to its ability to reach high concentrations within the CSF and brain tissue [5]. Most recently, a combination of surgical resection with antifungal therapy is considered the optimal management of solitary fungal brain abscesses [1,2].

IV. CONCLUSION

Extra-pulmonary blastomycosis remains a rare entity. The diagnosis is mainly based on examination of the CSF or the specimen after special staining. The mild or moderate forms are treated with itraconazole. In the case of severe, life-threatening infection, amphotericin B is required. The prognosis is unfortunately reserved for the price of several complications.

V. CONFLICTS OF INTEREST

The authors declare no competing interest. There were any non-financial competing interests.

Figures

Figure 1: Brain MRI showing a left retro-orbital non-hemorrhagic front-basal lesion in iso signal T1 and hypo signal on T2 weighted image taking contrast in an intense and homogeneous way with a hypo diffusion signal and marked surrounding vasogenic edema on T2 Flair weighted image.

Figure 2: Pathological examination showing, after coloration with Grocott's methenamine silver method, granulomatous inflammation and large broad-base and unipolar budding yeast-like cells.

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