Pneumocystis Jiroveci

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Introduction

Pneumocystis jiroveci (previously *Pneumocystis carinii*) is an opportunistic infection typically limited to the lungs. The organism is a fungus of low virulence that is likely spread through the air. A healthy immune system is able to control and prevent significant disease. Patients who are immunosuppressed (secondary to Human Immunodeficiency Virus, malignancy, chemotherapy, and iatrogenic secondary to steroids and other immunosuppressive agents) can develop a severe and potentially lethal pneumonia (the mortality rate is between 5–40 % even with treatment). Extrapulmonary manifestations of *Pneumocystis* are rare but can involve the liver, bone marrow, lymph nodes, and eyes. The ocular manifestations may be discovered incidentally and typically manifest as subretinal/choroidal yellow to white plaque like lesions.

Epidemiology

It has been estimated that in the pre-highly active anti-retroviral therapy (HAART) era, the incidence of *Pneumocystis* associated pneumonia occurred in 70–80 % of patients with HIV, and *Pneumocystis* associated choroiditis was diagnosed in approximately 1 % of HIV patients with CD4 counts less than 200 [1]. The choroidal involvement was more commonly diagnosed in those who were on prophylactic therapy with aerosolized pentamidine (presumably inhaled prophylaxis was inadequate to prevent disseminated disease). In one case series by Shami et al., 76 % of the cases were bilateral [2]. Since the advent of HAART therapy and routine prophylaxis with Trimethoprim/Sulfamethoxazole (TMP/SMX), the rates of *Pneumocystis* and associated choroiditis have plummeted.

Clinical Manifestations

The typical exam findings of ocular pneumocystosis are unifocal or multifocal (ranging from 2 to 50) yellow-white flat choroidal lesions predominantly involving the posterior pole. Few patients have visual symptoms despite having extensive choroidal lesions [3]. Fluorescein angiography of the lesions demonstrates early hypofluorescence and homogenous late staining

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[2]. There is generally no inflammation of the anterior chamber or vitreous.

Diagnosis

The diagnosis is often presumptive by identifying the characteristic fundus findings (yellow-white flat choroidal lesions) in patients with CD4 counts less than 200 or other high-risk criteria (chronic immunosuppression, malignancy, etc.). Although rarely performed due to risk of retinal complications, the diagnosis can be confirmed via chorioretinal biopsy demonstrating the characteristic cysts on toluidine blue or silver staining. More recently, quantitative polymerase chain reaction (qPCR) testing on bronchoalveolar lavage (BAL) specimens has been used to detect *Pneumocystis* DNA [4]. If validated, this type of testing may similarly be used on vitreous specimens.

Treatment

Pneumocystis choroidopathy requires systemic treatment as the ocular findings are a manifestation of disseminated infection. While officially classified as a fungal organism, *P. jiroveci* does not respond to antifungal therapy. The treatment of choice is TMP/SMX (dosing is TMP 15 mg/kg/day given for 21 days). Secondary agents include Pentamidine, Dapsone, and Atovaquone.

Conclusion

P. jiroveci is a rare disseminated opportunistic fungal infection that can manifest as choroidal infiltrates. The advent of HAART and prophylactic use of TMP/SMX has markedly reduced the incidence of this condition.

References

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