

CONTINUING MEDICAL EDUCATION ΣΥΝΕΧΙΖΟΜΕΝΗ ΙΑΤΡΙΚΗ ΕΚΠΑΙΔΕΥΣΗ

Hematology Quiz – Case 6

A 62-year-old man with acute myeloblastic leukemia (M2), who had completed his first cycle of consolidation chemotherapy for his disease, was admitted to the hospital due to fever up to 38.6°C and a left nasal congestion which was started 6 days before. The patient had been examined by an external otolaryngologist when the symptoms started and was treated with mometasone nasal spray, oral loratadine, and amoxicillin/clavulinate without any improvement in his symptoms.

His family history was unremarkable. The diagnosis of M2 AML was established 4 months before and the patient was treated with 2 courses of DAT chemotherapy (daunorubicin 50 mg/m² for D1, 3, 5; cytarabine 100 mg/m², bd, for 10 days & thioguanine 100 mg/m² bd for 10 days) followed by one cycle of high dose cytarabine. A complete remission was achieved after the first cycle induction therapy and was continued after the second cycle of high dose cytarabine. After the first consolidation therapy the patient experienced a remarkable neutropenia which lasted for one month despite the use of sc G-CSF. The symptoms started 42 days after the first day of consolidation therapy.

On physical examination the patient had a temperature of 38.2°C, a pulse rate of 110/min and a blood pressure of 125/90 mmHg. There was also an extended red & brownish area in the skin of the left nasal region along with left nasal congestion. A complete blood count revealed mild leukopenia ($3.1 \times 10^9/L$) with severe lymphopenia ($0.5 \times 10^9/L$), anemia (Hb 10.9 g/dl, Ht 34%) and normal platelet counts. The basic blood chemistry was within normal limits, with the exception of increased liver enzymes (AST 52 U/L, ALT 64 U/L and γ GT 73 U/L). Blood cultures were negative, while chest X-rays revealed no abnormalities. Due to the nasal symptoms a maxillofacial CT scan was performed and showed extensive left maxillary sinus disease. Soft tissue density was noted in the left maxillary sinus with higher density areas within. A widening of the left osteomeatal complex was also observed. The other paranasal sinuses were normal.

The patient was given a combination antibiotic therapy (ceftazidime, gentamicin and teicoplanine) with no effect on fever and then on day 4 of admission amphotericin B was added to therapy. The fever was stopped 5 days after amphotericin B therapy but the red/brownish area of the left nasal area was enlarged. One week after, the patient underwent sinus surgery

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ΑΡΧΕΙΑ ΕΛΛΗΝΙΚΗΣ ΙΑΤΡΙΚΗΣ 2008, 25(3):403-404

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Figure 1

where a left antrostomy/uncinectomy and left maxillary sinus evacuation were performed. The contents in the maxillary sinus consisted of yellowish to greenish, inspissated mucoïd material. Tissue specimens revealed inflammation with some tissue

necrosis, but also the presence of fungal hyphae collections. Fungal cultures revealed *Mucor rhizopus* infection of the area. The patients continued on antifungal therapy with the addition of voriconazole for one month. The patient continued to have intermittent nasal symptoms during the 4-month period of follow-up but a new maxillofacial CT scan showed no soft tissue in the left maxillary sinus.

Comment

Sinus mycetomas are considered a non-invasive form of fungal sinusitis. They are usually not associated with immunosuppressed hosts. The most common causes are Aspergillus fumigatus and dematiaceous fungi and are usually not bilateral. Although mucor has been identified in fungal sinusitis it is almost exclusively seen in immunosuppressed patients and it produces a rapid invasive, often fatal, infection.

The class zygomycetes is divided into two orders, Mucorales and Entomophthorales. These two orders produce dramatically different infections. Mucormycosis is an angioinvasive infection caused by the fungus Mucorales (Rhizopus, Mucor, Rhizomucor, Absidia, Apophysomyces, Cunninghamella and Saksenaea). Mucormycosis presents with rhinoorbital, cerebral, pulmonary, disseminated, cutaneous or gastrointestinal involvement. Our patient

had immunosuppression due to leukemia and the chemotherapy which had been given. Biopsy from the necrotic area was received in the department of Microbiology and processed according to the standard procedures. Mucor grew on SDA and was identified after seeing LCB mount. Treatment consists of a combination of surgical debridement of devitalized tissue and prompt medical therapy with amphotericin B. In general, therapeutic doses range from 1–1.5 mg/kg/day. Patient was immediately put on amphotericin B when the possibility of a fungal infection of the area was arisen. After 10 days of antifungal therapy, good clinical and microbiological response was seen. Amphotericin B was given for a period of 8 weeks (a total of 3 g), while voriconazole was also given for one month. In such conditions, mortality rates are very high and can approach almost 100% depending on the patient's underlying condition and form of Mucormycosis. Early diagnosis, along with the treatment of the underlying medical condition, surgery and an amphotericin product are needed for a successful outcome.

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