Acute Fungal Rhinosinusitis

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5.1 Case Presentation

A 39-year-old male patient with a medical history of autologous bone marrow transplantation 2 months ago due to relapse of a B-cell Hodgkin's Lymphoma, was admitted to Haematology Department with symptoms of worsening nasal congestion, purulent nasal discharge, persistent high fever, midfacial pain, and hard palate numbness. An urgent ENT assessment was requested. Nasal endoscopy revealed oedematous nasal mucosa, purulent nasal discharge, and black crust formation inside the right nostril. Multiple swabs, along with tissue samples, were collected for microbiological cultures and histopathological examination. Intraoral examination revealed a black ulcer with irregular margins over the right side of the hard palate, approximately 2×2 cm^2 in dimensions. Cranial nerve function was unremarkable. An urgent contrast-enhanced CT scan of the head and paranasal sinuses was performed, showing right maxillary sinus opacification with bone erosion (Fig. 5.1). Laboratory studies revealed elevated ESR and neutropenia. Blood cultures were also obtained. An urgent sinus MRI (Figs. 5.2 and 5.3) was performed, and the patient was taken to theatre for surgical debriment of the necrotic tissue, middle meatal antrostomies and biopsies under general anesthesia. A high dose of Amphotericin B was administered intravenously. The suspected diagnosis of mucormycosis, a subtype of Acute Invasive Fungal Sinusitis was confirmed.

5.2 Background Knowledge

Acute Invasive Fungal Rhinosinusitis is a rare and extremely aggressive disease with high morbidity and mortality (50–80%). It is related in the vast majority of cases with immunosuppression and especially malignancy, chemotherapy, uncontrolled diabetes, autoimmune disorders, and organ transplantation. There are several saprophytic fungi associated with acute invasive fungal rhinosinusitis. These fungi are inhaled and deposited in the airway, causing local or generalized inflammation to the immunocompromised patients. The most common are Mucor, Rhizopus, Rhizomucor, Absidia, Mortierella, Apophysomyces species and Aspergillus fumigatus, which is also related to the chronic form of invasive fungal sinusitis. Histopathological studies demonstrate mucosal invasion, vasculitis, arterial and venous thrombosis, and eventually, tissue necrosis. As it represents a potentially fatal and rapidly evolving disease, early diagnosis is of great importance.

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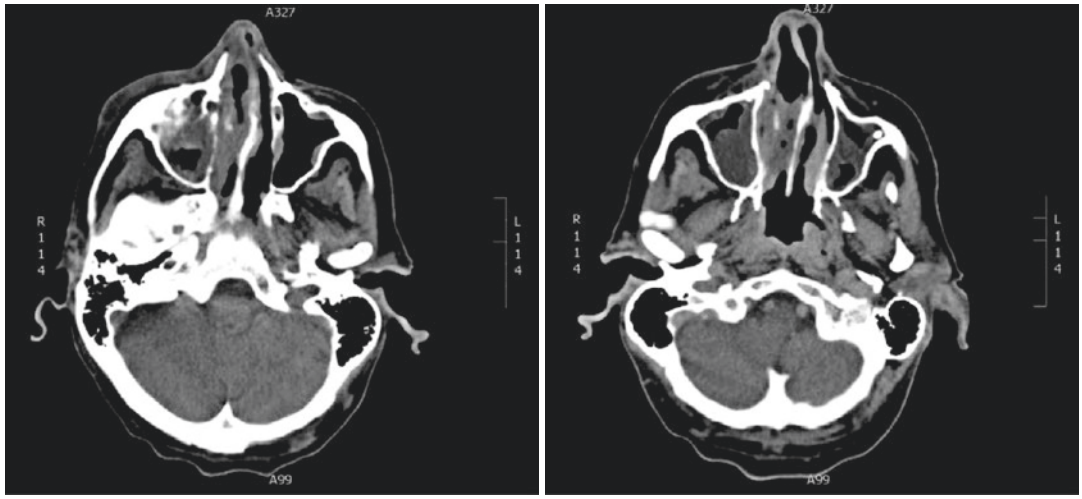

Fig. 5.1 CT scan of the paranasal sinuses

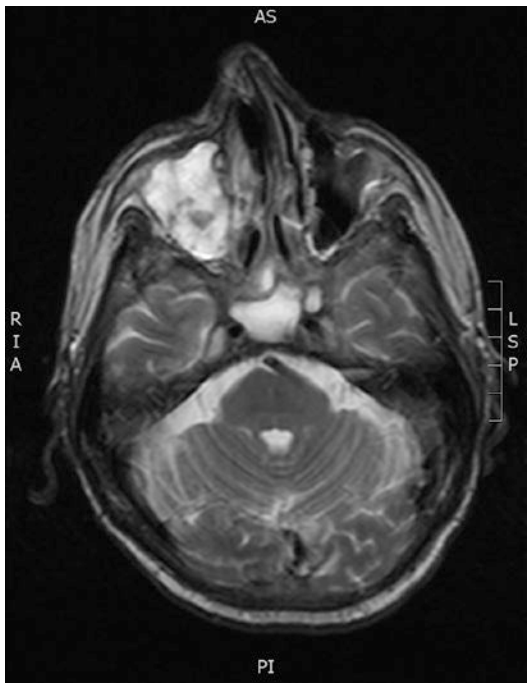

Fig. 5.2 T2-weighted MRI

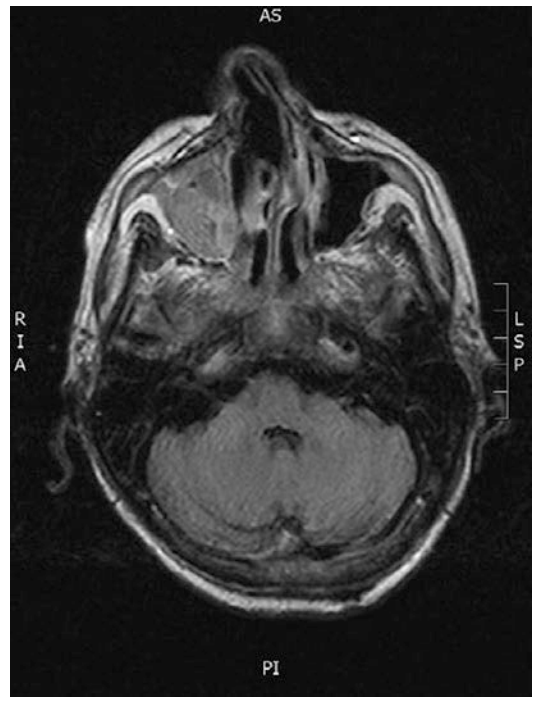

Fig. 5.3 T1-weighted MRI

5.3 Clinical Approach

5.3.1 Diagnosis

The clinician should be aware of the disease and its clinical manifestations in cases of immunosuppressed or immunocompromised patients, presenting with persistent high fever, severe headache and symptoms of rhinosinusitis. The clinical approach involves a thorough evaluation of the nasal cavities and paranasal sinuses, oral and pharyngeal mucosa, including endoscopy. Dark ulcers with eschar formation and swollen mucosa on the septum, turbinates and

palate, accompanied by nasal discharge and severe headache, are pathognomonic of the disease. Microbiology samples for bacteria and fungi should be taken. Tissue samples for culture and histopathologic examination should also be collected with great care because of the risk of bleeding, especially in patients with low platelet count. Cranial nerve function should be assessed because of the risk of intracranial invasion. Signs of ophthalmoplegia and/or exophthalmos with decreased pupillary responses indicate extension beyond paranasal sinuses to the orbit. Signs of cavernous sinus thrombosis are indicative of extensive disease. Urgent brain—sinus contrast CT scan should be performed. Bone erosions, soft tissue edema, mucosa thickening and vessel invasion may be noticed. MRI is more useful when intracranial, intraorbital or extension to the adjacent tissues is suspected.

5.3.2 Treatment

Once a rapidly evolving and possibly fatal disease, treatment should also be aggressive and effective. Urgent surgical resection with tissue debriment and complete disease removal, with simultaneous treatment of the underlying causes such as uncontrolled diabetes, neutropenia and immune system deficiency. In cases of early diagnosis with the disease isolated to the nasal cavity, without adjacent tissue invasions, surgical approach is restricted to extended endoscopic sinus surgery procedures (ethmoidectomy, medial maxillectomy, etc.). Sometimes extensive surgical procedures are required, such as transoral maxillectomy and orbital exenteration. Preoperative platelet transfusion should be considered in patients with low platelet count because of the risk of bleeding. High doses of Amphotericin B or Lipid Formulation of Amphotericin B should be administered intravenously as soon as the diagnosis of invasive fungal sinusitis is suspected.

5.3.3 Follow-up

As already mentioned, acute invasive fungal rhinosinusitis is related to high morbidity and mortality rates. Antifungal medication should be continued for an extended period after surgery. Those who remain disease-free require close monitoring due to the possibility of recurrence of the disease. Several specialists may be involved in the follow-up, such as ENT and Oral&Maxillofacial surgeons due to possible facial deformities after extensive surgeries, Hematologists in cases of hematologic malignancies, Clinical Oncologists, Immunologists, Endocrinologists in cases of diabetes etc.

Summary and Author's Comments

1. Acute Invasive Fungal Rhinosinusitis is a rare, extremely aggressive disease with high morbidity and mortality rates (50–80%).
2. Clinicians should be aware of the disease in cases of immunocompromised patients, presenting with persistent high fever, rhinosinusitis symptoms with nasal discharge, severe headache, black crust—eschar formation in the nasal cavity.
3. Intraorbital extension, opthalmoplegia, cavernous sinus thrombosis and cranial nerve involvement are signs of extensive disease.
4. Urgent wide surgical resection combined with high doses of antifungal medication and treatment of underlying conditions is the treatment of choice.

Bibliography

1. Roden MM, Zaoutis TE, Buchanan WL, Knudsen TA, Sarkisova TA, Schaufele RL, et al. Epidemiology and outcome of zygomycosis: a review of 929 reported cases. Clin Infect Dis. 2005;41(5):634–53.
2. Drakos PE, Nagler A, Or R, Naparstek E, Kapelushnik J, Engelhard D, et al. Invasive fungal sinusitis in

patients undergoing bone marrow transplantation. Bone Marrow Transplant. 1993;12(3):203–8.
3. Kwon-Chung KJ. Taxonomy of fungi causing mucormycosis and entomophthoramycosis (zygomycosis) and nomenclature of the disease: molecular mycologic perspectives. Clin Infect Dis. 2012;54(Suppl 1):S8–S15.
4. Montone KT. Pathology of fungal rhinosinusitis: a review. Head Neck Pathol. 2016;10(1):40–6.
5. Deshazo RD. Syndromes of invasive fungal sinusitis. Med Mycol. 2009;47(Suppl 1):S309–14.
6. Fung M, Babik J, Humphreys IM, Davis GE. Diagnosis and Treatment of acute invasive fungal sinusitis in cancer and transplant patients. Curr Infect Dis Rep. 2019;21(12):53.
7. Ferguson BJ. Mucormycosis of the nose and paranasal sinuses. Otolaryngol Clin N Am. 2000;33(2):349–65.
8. Fernandez IJ, Crocetta FM, Dematte M, Farneti P, Stanzani M, Lewis RE, et al. Acute invasive fungal rhinosinusitis in immunocompromised patients: role of an early diagnosis. Otolaryngol Neck Surg. 2018;159(2):386–93.
9. Kasapoglu F, Coskun H, Ozmen OA, Akalin H, Ener B. Acute invasive fungal rhinosinusitis: evaluation of 26 patients treated with endonasal or open surgical procedures. Otolaryngol Neck Surg. 2010;143(5):614–20.
10. Zuniga MG, Turner JH. Treatment outcomes in acute invasive fungal rhinosinusitis. Curr Opin Otolaryngol Head Neck Surg. 2014;22(3):242–8.
11. Goldstein EJC, Spellberg B, Walsh TJ, Kontoyiannis DP, Edwards J Jr, Ibrahim AS. Recent advances in the management of mucormycosis: from bench to bedside. Clin Infect Dis. 2009;48(12):1743–51.
12. Turner JH, Soudry E, Nayak JV, Hwang PH. Survival outcomes in acute invasive fungal sinusitis: a systematic review and quantitative synthesis of published evidence. Laryngoscope. 2013;123(5):1112–8.
13. Stavrakas M, Karkos PD, Dova S, Tzorakoeleftheraki SE. Unilateral fungal sphenoiditis presenting with diplopia and ptosis. Indian J Otolaryngol Head Neck Surg. 2017;69(3):428–9. https://doi.org/10.1007/s12070-017-1138-x.