The Journal of Laryngology & Otology
August 2005, Vol. **119,** pp. 643–645

Pott's puffy tumour caused by mucormycosis

K G Effat, FRCS (Ed), FRCS (I), DLO, M Karam, MSc, A El-Kabani, MSc

Abstract
A case of Pott's puffy tumour in a diabetic patient with renal failure is reported. The patient did not respond to intravenous antibiotics and further investigation revealed that the patient had mucormycosis. As far as we are aware, this is the first case of Pott's puffy tumour due to mucormycosis to be reported in the literature.

Key words: Mucorales; Paranasal Sinuses; Fistula; Diabetes Mellitus; Kidney Failure; Therapy

Introduction

Mucormycosis is an opportunistic fungal infection caused by fungi of the order Mucorales.[1] The most common form of the disease is rhinocerebral mucormycosis.[2] The disease runs a spectrum of clinical presentations ranging from a rapidly fulminant process to a more protracted indolent course, depending on the immune status of the host.[3] Rhinocerebral mucormycosis usually originates in the nasal mucosa from where it spreads to the sinuses, orbit and cranial cavity.[4] The fungus has a remarkable affinity for blood vessel walls, causing thrombosis and infarction of tissues.[5]

Pott's puffy tumour is a doughy swelling over the forehead, with oedema of the upper eyelid,[6] which most commonly signifies an osteomyelitis of the frontal bone as a complication of sinusitis. This condition has become rare since the introduction of antibiotics.[7] In the past the common organisms isolated were *S. aureus,* non-enterococcal streptococci and oral anaerobes.[8]

A literature search did not find any reported case of Pott's puffy tumour caused by mucormycosis and, as far as we are aware, we are therefore reporting the first case with such an association.

Case report

The patient was a 62-year-old house-wife who presented with severe headache, fever and a forehead swelling which had been progressively enlarging over the last two days. There had been discharge of a purulent material from the region of the left inner canthus for one week and swelling of the upper eyelid. There was a history of insulin-dependent diabetes and hypertension and she also had chronic renal failure, for which she was undergoing haemodialysis twice a week.

Examination showed a diffuse swelling on the forehead about 6 cm in diameter with a swelling of the left upper eyelid. Also noted on inspection was a fistula discharging pus in the region of the left inner canthus (Figure 1). The patient was pyrexial with a temperature of 38°C. Rigid endoscopy of the nose showed a pale left middle turbinate but there were no polyps. Haematological investigations revealed a leucocytosis and severe anaemia, with a haemoglobin concentration of 6.2 gm/dl. The random blood sugar was high at 320 mg/dl and the urea and creatinine levels were also elevated.

A presumable diagnosis of Pott's puffy tumour and left orbital cellulitis secondary to acute sinusitis was made and an urgent CT scan was carried out. The scan revealed opaque left maxillary and frontal sinuses and the left anterior and middle ethmoid cells were occupied by a

Fig. 1
Photograph of patient on admission, showing forehead and left upper eyelid swellings and fistula (after permission of patient).

From the Department of Otolaryngology, Head and Neck Surgery, El-Sahel Teaching Hospital, Cairo, Egypt.
Accepted for publication: 15 March 2005.

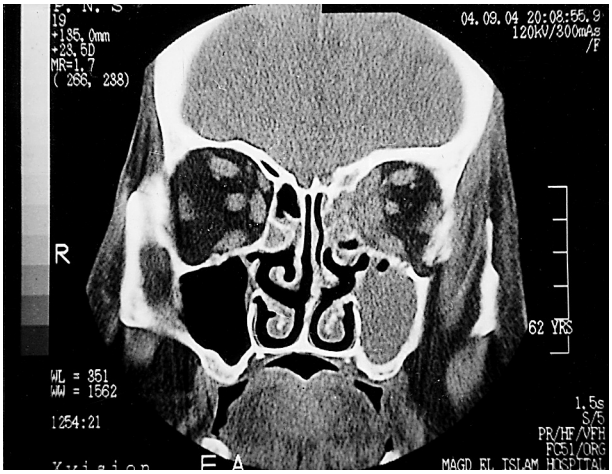

FIG. 2

Coronal CT scan of patient on admission showing opacity in left sinuses and subperiosteal abscess.

heterogeneous material with erosion of the lamina papyracea and a subperiosteal orbital abscess (Figure 2). The patient was admitted and received three units of blood.

An ophthalmologic consultation revealed normal ocular mobility and no deterioration in her vision. She was started on intravenous cefoperazone, gentamicin and metronidazole while the results of blood culture and cultures from the pus were awaited. However, the following day the patient became toxic and the forehead swelling enlarged and became fluctuant. A cruciate incision was undertaken under local anaesthesia and yielded yellowish brown purulent material which was sent for bacteriological study. The results of all cultures were negative. Over the next three days she was haemodynamically stable but there was persistent drainage from the forehead incision and the fistula. Due to her immunocompromised status a fungal infection was suspected, and a sample of necrotic tissue from the fistula was sent for histopathology. This revealed broad fungal hyphae, which were rarely septate, together with necrosis and inflammation of the tissues (Figure 3). The diagnosis of invasive mucormycosis was made.

The patient was started on amphotericin B (Fungizone) infusions together with daily debridement of necrotic material from the fistula and frequent dressings. Her condition progressively improved with cessation of drainage and resolution of the forehead and eyelid swellings. She was discharged from hospital three weeks after admission and at the last follow up was in complete remission.

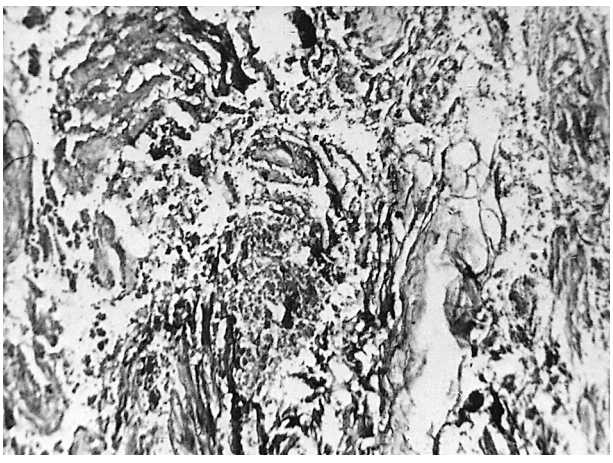

FIG. 3

Histopathology revealing fungal hyphae (H&E; original ×40).

Discussion

The anatomy of the orbit predisposes to the spread of infection from the sinuses, both extracranially and intracranially. The lamina papyracea is exceptionally thin and fragile. The veins around and within the orbit, including the superior and inferior ophthalmic veins, form a diffuse network of interconnected valveless branches, and the direction of blood flow depends on local pressure gradients. Thus communication between the nose, sinuses, face, orbit and cavernous sinus exists.[9]

Osteomyelitis of the frontal bone may follow an acute or chronic course. In the acute form there is headache, oedema of the upper eyelids and a doughy swelling of the skin over the frontal sinuses (Pott's puffy tumour) and this may be followed by pericranial abscess formation and intracranial spread if there is perforation of the inner table of the cranium. In the chronic form the onset is insidious with a low-grade fever, local pain, a doughy forehead swelling and malaise. The diagnosis of osteomyelitis is made on the basis of the history, physical findings and CT scans and involvement of the brain by MRI scanning.[6]

A Medline computer search over the last 10 years did not reveal any case of Pott's puffy tumour caused by mucormycosis. One microbiological study of 103 trephination procedures in patients with acute frontal sinusitis did not reveal any fungal agents[8] and another study of 46 patients also failed to reveal fungi.[10] However, fungal diseases have emerged as major challenges for physicians and microbiologists and the prevalence of mycotic infections and the number and diversity of pathogenic fungi have increased dramatically in recent years.[11] The organisms causing rhinocerebral mucormycosis have a propensity to colonize the nasal cavity and sinuses and then spread by direct extension or through blood vessels into the brain, orbit and other head and neck structures. Patients with diabetic ketoacidosis are the most likely patients to develop rhinocerebral mucormycosis[12] and our patient was a poorly controlled diabetic. The diagnosis can be confirmed by biopsy, as histopathological examination is the most definite and rapid means of establishing the diagnosis. Positive microscopy demonstrates with certainty that a fungus is present in the specimen, and it is also possible to determine whether the fungus has invaded host tissue and to characterize the tissue response. Moreover, it is possible to study the fungal morphology.[13] The causative aetiology in our patient was only obtained by histopathology, which revealed invasive mucormycosis.

The management of rhinocerebral mucormycosis consists of control of the original precipitating condition, heparinization, systemic amphotericin B and local drainage and debridement.[2] The status of the host is most important, as the phagocytic response, rather than antibody production, provides the primary host defence against invasive fungi. Immunologic stimulation has been attempted in this disease by the use of granulocyte-macrophage colony-stimulating factor (GM-CSF) and granulocyte colony-stimulating factor (G-CSF) in an attempt to improve the phagocytic response.[14] Our patient had been heparinized during her stay in hospital three times a week for the haemodialysis sessions. This may have contributed to a more favourable outcome by minimizing the vascular thrombosis and infarction. Lipid formulations

https://doi.org/10.1258/0022215054516304 Published online by Cambridge University Press

of amphotericin (liposomal amphotericin B) are now available, and are much more potent and less toxic to the kidneys than conventional amphotericin B.[15] It has also been suggested that patients should have surgery, as the removal of all the devascularized tissue permits the penetration of the antifungal agent via the blood stream.[13] However, our patient did not require extensive surgery as there was a prompt response to amphotericin B.

In conclusion, a case of invasive mucormycosis presenting as a Pott's puffy tumour in a diabetic patient with renal failure is reported. Early clinical recognition of this potentially fatal disease is essential before irreversible changes occur.[2] The diagnosis and treatment of this life-threatening infection are discussed.

- **This paper presents the first case of an invasive mucormycosis presenting as a Pott's puffy tumour in an immunocompromised patient**
- **The authors point out that histopathology is the most rapid and effective method of diagnosing invasive fungal rhinosinusitis**
- **They outline and discuss recent therapeutic modalities such as growth factors and liposomal amphotericin B, and emphasise timely adequate surgical intervention - all of which have improved the prognosis in this serious condition**

This paper presents the first case of an invasive mucormycosis presenting as a Pott's puffy tumour in an immunocompromised patient. The authors point out that histopathology is the most rapid and effective method of diagnosing invasive fungal rhinosinusitis. They outline and discuss recent therapeutic modalities such as growth factors and liposomal amphotericin B, and emphasize timely adequate surgical intervention - all of which have improved the prognosis in this serious condition.

References

1 Ferguson BJ. Mucormycosis of the nose and paranasal sinuses. *Otolaryngol Clin North Am* 2000;**33**:349–65
2 Weir N, Golding-Wood DG. Infective rhinitis and sinusitis. In: Kerr A, ed. *Scott-Brown's Otolaryngology,* Vol 4, 6th edn. Oxford: Butterworth-Heinemann, 1997;4/8/41
3 Ferguson BJ. Definitions of fungal rhinosinusitis. *Otolaryngol Clin North Am* 2000;**33**:227–35
4 Gillespie MB, O'Malley BW. An algorithmic approach to the diagnosis and management of invasive fungal rhinosinusitis in the immunocompromised patient. *Otolaryngol Clin North Am* 2000;**33**:323–34
5 Janecka IP. Maxillofacial infections. *Clin Plast Surg* 1979;**6**:553–73
6 Feldman BA, Feldman DE. The nose and sinuses. In: Lee KJ, ed. *Essential Otolaryngology,* 6th edn. Connecticut: Appleton & Lange, 1995;746
7 Goldberg AN, Oroszglan G, Anderson TD. Complications of frontal sinusitis and their management. *Otolaryngol Clin North Am* 2001;**34**:211–25
8 Antila J, Suonpaa J, Lehtonen O-P. Bacteriological evaluation of 194 adult patients with acute frontal sinusitis and findings of simultaneous maxillary sinusitis. *Acta Otolaryngol (Stockh) Suppl* 1997;**529**:162–4
9 Kleinman DM, Johnson DW, Braverman JM. The eye and orbit. In: Jafek BW, Stark AK, eds. *ENT Secrets.* Philadelphia: Hanley & Belfus, 1996;375
10 Ruoppi P, Seppa J, Nuutinen J. Acute frontal sinusitis: etiological factors and treatment outcome. *Acta Otolaryngol (Stockh)* 1993;**113**:201–5
11 Mitchell TG. Overview of basic medical mycology. *Otolaryngol Clin North Am* 2000;**33**:237–49
12 Maitra A, Kumar V. The lung and the upper respiratory tract. In: Kumar V, Cotran RS, Robbins SL, eds. *Robbins Basic Pathology,* 7th edn. ????: WB Saunders, 2003;494
13 Schell WA. Histopathology of fungal rhinosinusitis. *Otolaryngol Clin North Am* 2000;**33**:251–76
14 Adler SC, Isaacson G, Sasaki CT. Invasive aspergillosis of the paranasal sinuses and orbit: can you save the eye? *Am J Otolaryngol* 1997;**18**:230–4
15 Mondy KE, Haughey B, Custer PL. Rhinocerebral mucormycosis in the era of lipid-based Amphotericin B: case report and literature review. *Pharmacotherapy* 2002;**22**:519–26

Address for correspondence:
Dr Kamal G Effat,
37 Kods El-Sherif Street,
Madinet El-Mohandeseen,
Giza, Egypt

E-mail : kamaleffat@hotmail.com

Dr K G Effat takes responsibility for the integrity of the content of the paper.
Competing interests: None declared