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Facial nerve palsy as an unusual presentation of orbital apex syndrome

Porażenie nerwu twarzowego jako nietypowa postać zespołu szczytu oczodołu

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Abstract

Clinical cases of orbital apex syndrome are rare and most commonly manifested as a complication of fungal sinusitis, mainly in immunocompromised and poorly controlled diabetic patients. Rhino-orbital mucormycosis is a rare opportunistic, aggressive and fatal infection caused by mucor. The complex presentation of orbital apex syndrome not only poses a diagnostic challenge but also demands a multidisciplinary approach in patient management. Facial nerve palsy is an unusual presentation in orbital apex syndrome. We report the case of a 64-year-old diabetic patient presenting with ophthalmoplegia and visual loss associated with facial nerve palsy. Prompt ophthalmologic and otolaryngologic intervention with imaging and histologic confirmation, followed by early initiation of antifungal and antimicrobial therapy, were integral to preventing further complications, and reducing morbidity and mortality.

Keywords: facial nerve palsy, orbital apex syndrome, diabetes mellitus, mucormycosis

Streszczenie

Przypadki kliniczne zespołu szczytu oczodołu występują rzadko, a choroba najczęściej rozwija się jako powikłanie grzybiczego zapalenia zatok przynosowych, głównie u pacjentów z obniżoną odpornością i nieprawidłowo kontrolowaną cukrzycą. Mukormykoza nosowo-oczodołowa jest rzadkim, oportunistycznym zakażeniem o agresywnym przebiegu, które może mieć skutek śmiertelny. Schorzenie wywoływane jest przez grzyby z rzędu *Mucorales*. Złożony obraz kliniczny zespołu wierzchołka oczodołu nie tylko stanowi wyzwanie diagnostyczne, ale także wymaga wielodyscyplinarnego podejścia do leczenia. Porażenie nerwu twarzowego jest nietypową postacią zespołu szczytu oczodołu. Przedstawiamy przypadek 64-letniego chorego na cukrzycę, u którego wystąpiły oftalmoplegia i uszkodzenie wzroku związane z porażeniem nerwu twarzowego. Szybka interwencja okulistyczna i otolaryngologiczna z potwierdzeniem rozpoznania badaniami obrazowymi i histologicznymi, a następnie wczesne rozpoczęcie leczenia przeciwgrzybiczego i przeciwdrobnoustrojowego stanowiły integralne elementy zapobiegania kolejnym powikłaniom i ograniczania dalszej zachorowalności i śmiertelności.

Słowa kluczowe: porażenie nerwu twarzowego, zespół szczytu oczodołu, cukrzyca, mukormykoza

INTRODUCTION

Orbital apex syndrome (OAS) is characterised by the involvement of 2nd, 3rd, 4th and 6th cranial nerves along with the ophthalmic division of the 5th cranial nerve[1,2]. Sino-orbital fungal infection leading to OAS with involvement of multiple cranial nerves signifies severe infection, aggressive in nature, with poor prognosis. The common pathogens are fungi including mucormycosis and aspergillosis, bacteria, viruses, and spirochetes[3,4]. Paranasal sinus mucormycosis is a life-threatening rare opportunistic mycotic infection[5] which can spread to the retro-orbital region through the orbital canal, superior orbital fissure and cavernous sinus as well as the anatomically-related petrous apex. However, the involvement of the 7th cranial nerve in association with rhino-orbital mucormycosis and OAS is extremely rare. Some case reports have revealed that granuloma due to mycotic infection was misdiagnosed as malignant tumour and caused multiple nerve palsy[6,7]. Complex and vague presentation is a great hindrance to early diagnosis and initiation of appropriate management, which is critical in such situations.

CASE REPORT

A 64-year-old man with diabetes mellitus type 2 and hypertension presented with right-sided nasal discharge and blocked nose with right facial pain persisting for eight months. It was associated with drooping of the right eyelid, blurred and double vision as well as vomiting and significant weight loss. On examination, the right maxillary facial region was erythematous. Nasoendoscopy revealed right nasal mucosal congestion, middle turbinate hypertrophy, and blood-stained pus occupying the middle meatus. The examination of the left nasal cavity was unremarkable.

Fig. 1. Right facial nerve palsy grade V

Cranial nerve examination revealed involvement of the 3rd, 4th, 5th and 6th cranial nerves with sparing of taste sensation. In addition, other significant clinical features observed included drooping of the corner of the mouth, drooling of saliva, and absence of wrinkles in the right half of the forehead suggestive of lower motor neuron lesion (Fig. 1). There was hypoesthesia along the distribution of the 1st and 2nd divisions of the right trigeminal nerve. Ophthalmic examination revealed a visual acuity of counting finger at 20 cm and 3/60 in the right and left eyes, respectively. The intraocular pressure (IOP) was 13 mm Hg in the left, and 26 mm Hg in the right eye. Further examination showed severe chemosis and positive relative afferent pupillary defect in the right eye. Fundoscopy revealed proliferative diabetic retinopathy in the right eye.

He was diagnosed with acute maxillary sinusitis with ophthalmoplegia, periorbital oedema, loss of visual acuity, and orbital cellulitis with the 7th cranial nerve palsy grade V. Laboratory tests revealed leukocyte count of 16,980/mL with neutrophil predominance, ESR of 37 mm/hr, and high C-reactive protein (CRP) level of 5.27 mg/dL. Contrasted computed tomography (CT) scan of the orbit and paranasal sinuses (PNS) revealed mucosal thickening and a soft tissue mass in the right maxillary sinus, eroding the medial wall, and a bony defect in the anteromedial wall of the left sphenoid sinus (Fig. 2). This finding raised the suspicion of paranasal sinus tumour causing OAS.

In view of persistent chemosis and high IOP in the right eye, endoscopic sinus surgery was planned for debridement, tissue biopsy, and culture. A histopathological specimen from the maxillary sinus tissues revealed fungal hyphae with sparse septation surrounded by chronic granulomatous inflammation, suggestive of mucormycosis. Intravenous insulin infusion and amphotericin B at 0.3 mg/kg/day were commenced, and the latter was gradually increased to 1 mg/kg/day, with monitoring of the renal function. There was a significant improvement in sinusitis, vision, ptosis, and ophthalmoplegia. The swelling and redness in the right eye improved gradually with oral amphotericin taken twice a day for a month. Nonetheless, facial nerve paralysis remained unchanged after a period of follow-up until one year.

DISCUSSION

The most common causes of OAS from PNS are sino-nasal tumour and fungal sinusitis. A retrospective review of 50 OAS patients found that fungal infection was the least common cause, while neoplasia was the most common

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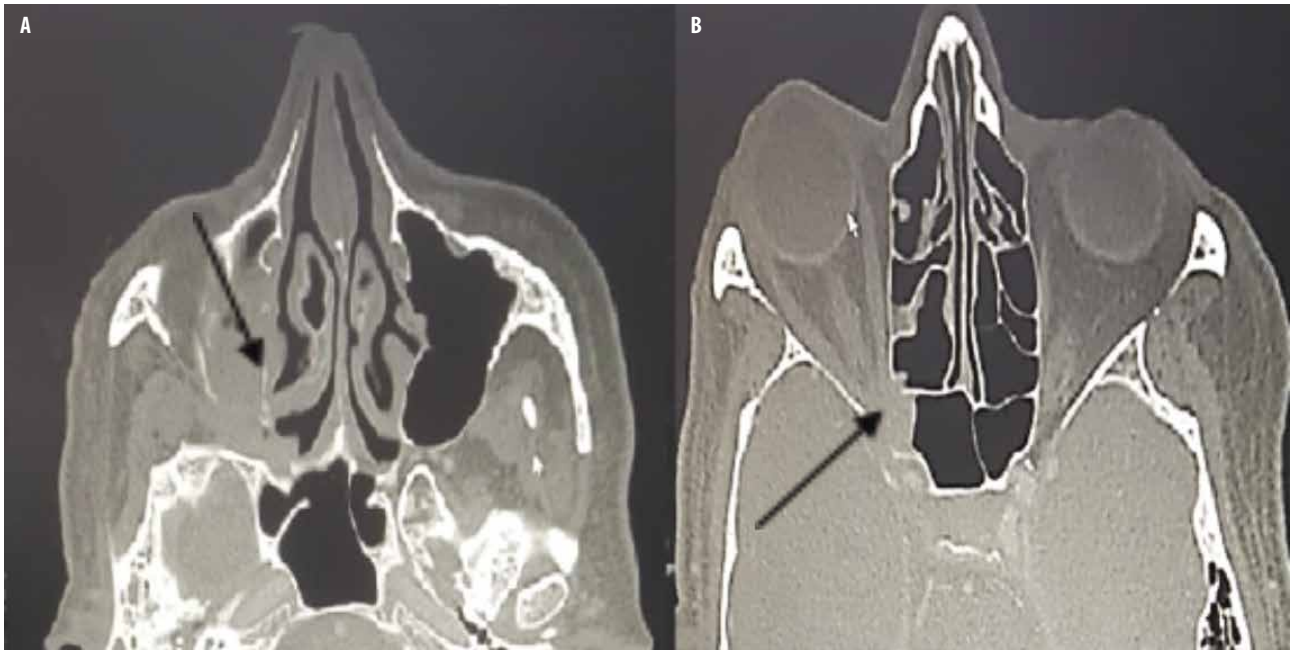

*Fig. 2. Axial CT of PNS showing mucosal thickening and granuloma in maxillary sinus with medial wall erosion (***A***) and mass extending to orbital apex (***B***)*

aetiology[8,9]. In our case, invasive granulomatous fungal sinusitis was shown to cause ocular symptoms by invading the orbital apex, a potentially fatal effect infrequently associated with OAS.

Ophthalmoplegia and vision loss are the hallmark features of OAS, as it involves the 2nd, 3rd, 4th and 5th cranial nerves (ophthalmic division) in the orbital apex. The underlying aetiologies can be classified into vascular, inflammatory, neoplastic, and infective[10]. Any infection arising in the maxillary sinus, and extending to the orbit and apex, can produce a significant effect on the movements of the extraocular muscles and vision. The orbital apex is formed by the optic canal with the optic nerve and very close to the superior orbital fissure which contains numerous neurovascular structures of the cranium[11]. It can spread through these channels to the intracranial compartment, leading to meningitis, intracranial abscess, and venous sinus thrombosis, and can be fatal if the infection is not treated effectively[12].

PNS mucormycosis can cause complications as seen in rhino-orbito-cerebral mucormycosis, including headache, fever, sinusitis, facial swelling, and unilateral OAS. Intracranial involvement presents with convulsions, changes in consciousness, coma, and stroke[11]. Direct orbital extension from the maxillary and sphenoid sinus eroding the orbital apex may involve the optic nerve with blindness, either of central retinal artery occlusion. The orbital findings in our case included painful eye, vision loss and periorbital oedema in the right eye, leading to closure of the eye and proptosis. Other signs of significant neurological involvement were drooping of the corner of the mouth, drooling of saliva, and absence of wrinkles in the right half of the forehead, which were suggestive of right lower motor neuron facial nerve palsy in our case.

However, facial nerve involvement in association with rhino-orbital mucormycosis and OAS is an unusual presentation. Our patient had right facial nerve palsy. It is important to note the close relationship of the orbital apex, petrous apex and pterygopalatine fossa which is considered to be a reservoir of mucor, from where it spreads to the retro-orbital space and infratemporal fossa[13]. The incidence of facial nerve paralysis in conjunction with rhino-orbital-cerebral mucormycosis was 11%[10]. Although the pathophysiology of facial nerve paralysis is not known, in most diabetic patients pathological changes in the wall of the arteries causing ischaemia and compression effect and facial nerve palsy sparing the taste sensation from the chorda tympani nerve, as seen in our patient. Although whole facial nerve involvement is more commonly associated with viral or other aetiologies, it is not certain that only the distal part of facial nerve is affected in diabetics.

In terms of age, neoplastic aetiology is more commonly seen in the younger age group, while infectious and inflammatory causes together with immunosuppression associated with diabetes mellitus, renal failure or long-term use of immunosuppressive agents, are more common in older patients[14,15]. Similarly, our 64-year-old patient is a known diabetic who presented with PNS infection extending to the orbital apex, superior orbital fissure and cavernous sinus, resulting in OAS.

Radiological investigations are mandatory in identifying sinus lesions with intracranial as well as infratemporal

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extent of the disease and cavernous sinus thrombosis. Contrasted CT scan of the orbit and PNS showed mucosal thickening and a soft-tissue mass in the right maxillary sinus, eroding the medial wall, and a bony defect in the anteromedial wall of the left sphenoid sinus. Nevertheless, tissue culture and histopathology is the gold standard investigation for the diagnosis of mucormycosis and differentiation between benign or malignant lesions[16]. In our patient, the diagnosis was confirmed histologically by detecting sparsely septate hyphae with angled branching, which is pathognomonic of mucormycosis in the maxillary sinus.

Since OAS secondary to invasive fungal sinusitis is associated with a high mortality rate of almost 30–70%, prompt and appropriate management, ideally within 5 days, following early detection and diagnosis, is quintessential for a better prognosis[17]. OAS secondary to fungal infection, being a rare condition, has no consensus in terms of algorithm for patient management[11,13]. Therefore, most patients reviewed had neurological sequelae and fatal outcomes. Nonetheless, combined medical and surgical treatment increases the survival rate from 57.5% to 78%, compared to the medical management alone[18,19]. The standard medical therapy for sino-orbital mucormycosis is amphotericin B at 1–1.5 mg/kg/day for several weeks, based on the clinical response and degree of nephrotoxicity[20]. Our patient was initially scheduled for endoscopic sinus surgery and orbital decompression. However, his symptoms improved upon receiving amphotericin B at 0.5 mg/kg over 6 hours intravenously for the first three days, followed by 1 mg/kg/day for four weeks. In refractory patients and infections that are resistant to amphotericin B, posaconazole is an alternative drug of choice primarily as a cytochrome P-450 3A4 inhibitor[21].

CONCLUSION

Facial nerve palsy in uncontrolled diabetic patients with mucormycosis and OAS is a rare but significant and disfiguring manifestation. Therefore, it is of utmost importance to optimise the blood sugar level prior to surgical debridement and initiate appropriate antifungal therapy for better outcome.

Conflict of interest

The authors do not declare any financial or personal links with other persons or organisations that might adversely affect the content of the publication or claim any right to the publication.

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