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Case report

Orbital Actinomycetoma with cranial extension: A rare case report

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ABSTRACT

Introduction: Mycetoma is a neglected tropical disease that commonly affects the lower extremity. The disease is attributed to subcutaneous granulomatous inflammation leading to distinct clinical features of gradual painless swelling accompanied by nodules and drains. Orbital mycetoma is an extremely rare entity of the disease. We reported the clinical presentation, diagnosis, and surgical outcomes in a case of orbital mycetoma with cranial extension.

Case presentation: A 25-years-old male complained of left eye protrusion for 8 years, followed by complete loss of vision on the left eye for 7 years and eventually left eye pain for the last year. The left eye was displaced anteriorly and inferiorly with normal oculomotor, abducent, and trochlear examination. Brain CT scan showed an increase in orbital bone thickness with extension to the anterior cranial base, middle cranial base, and the orbital process of the zygomatic bone. MRI revealed a large lesion involving the left frontotemporal region with extension to left orbit, left posterior ethmoid air cells, and left temporal suprasellar region. The lesion was homogenously enhanced with contrast. The patient underwent a left orbitozygomatic cranitomy for resection of the lesion. However, total resection was inapplicable due to the extension of the bony lesion up to the petrous bone. Cranioplasty was performed by titanium mesh.

Conclusion: Mycetoma is a chronic inflammatory disease affecting subcutaneous tissues commonly in the lower limbs. The disease can be caused by fungi (Eumycetoma) or bacteria (Actinomycetoma). Orbital mycetoma is an extremely rare entity of the disease. However, it is commonly associated with the cranial extension. Early diagnosis and prompt surgical and medical treatment are the keys to good outcomes.

1. Introduction

Mycetoma is a neglected tropical disease characterized by chronic escalating granulomatous inflammation that usually occurs in subcutaneous tissues and continues to invade skin and bones eventually [1,2]. The disease presents with painless slow-growing swelling commonly affecting mainly lower limbs after transmission of the causative organism through traumatic inoculation [1]. In addition, the swelling contains sinuses with purulent discharge and grains [3]. Mycetoma is classified into two categories according to the causative organism. Actinomycetoma (bacterial) is commonly caused by *Streptomyces somaliensis*, however, Eumycetoma (fungal) is commonly caused by *Madurella mycetomatis* [4].

Cranial mycetoma is a rare entity of the disease. One study revealed that the incidence of head and neck mycetoma was merely 0.76% of the total cases of mycetoma over a period of 23 years reported at the mycetoma research center in Sudan [5]. In the same study, orbital involvement of mycetoma was confirmed only in 2 patients out of 49 patients with head and neck mycetoma [5]. We reported a case of orbital Actinomycetoma treated surgically and medically by a multidisciplinary team of neurosurgeons and maxillofacial surgeons and physicians. This case has been reported in line with SCARE guidelines [6].

2. Case presentation

A 22-year-old male, a shepherd by profession, and originally from Gadarif state in the eastern part of Sudan, came to the outpatient clinic complaining of left eye protrusion for 8 years, followed by complete loss of vision on the left eye for 7 years and eventually left eye pain for 1 year. The patient is not known to have diabetes, hypertension, or other chronic illness and without any previous history of surgical interventions.

At the time of admission, the patient was confused with a GCS of 15 (M = 6, V = 5, E = 4), his vital signs were stable, like the following, BP:

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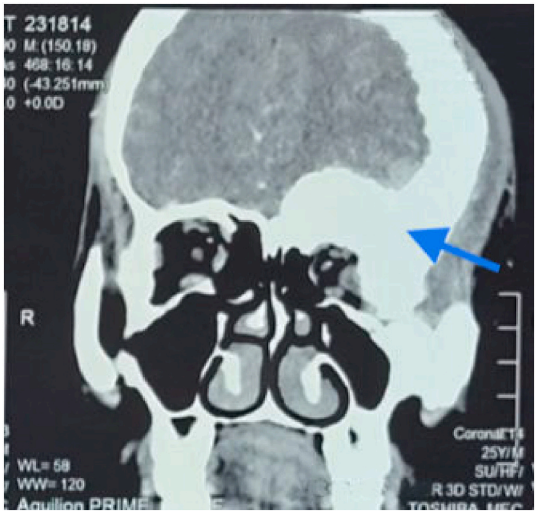

Fig. 1. CT brain shows supraorbital bony thickness.

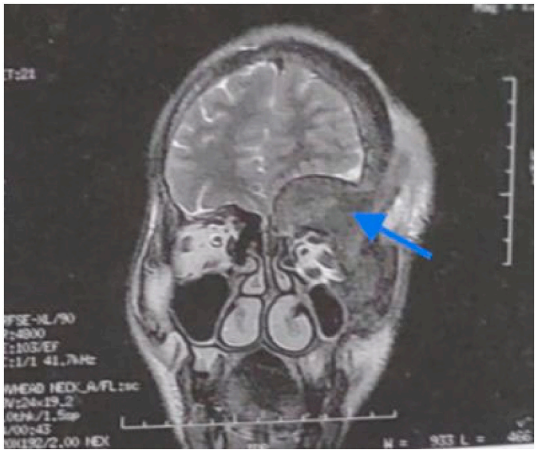

Fig. 2. Brain MRI shows large hypodense frontotemporal lesion.

120/80, PR: 86/min, Spo2: 98% on room air, RR: 16/min, temperature: 37.4. The neurological examination showed normal power and tone in both upper and lower limbs. However, the optic nerve examination revealed total loss of vision on the left eye with normal oculomotor, abducent, and trochlear examination. The systematic review was insignificant with normal chest, abdominal, and pelvic examination.

The condition started in 2013 when the patient got hit by a cow hoof in his left eye which led to a small wound in his left eyelid, he had no pre-accident events (loss of consciousness, convulsions, or amnesia). Then he gradually started to have protrusion of his eye which led to a purulent discharge in 2014. This purulent discharge leads to complete loss of vision on the left eye. After that event, the patient sought medical advice from an ophthalmologist, prescribed some medications protrusion improved. However, no improvement of vision was noticed. After protrusion improvement, the patient lost the follow-up till 2020 when protrusion started to increase again gradually and was associated with pain in the left eye. Then the patient sought medical advice again, this time the ophthalmologist requested CT brain which revealed a large bony lesion on the left supraorbital region. The patient was referred to our hospital for surgical intervention.

Coronal CT scan of the brain revealed supraorbital bone thickness with extension to the anterior cranial base and the orbital process of the zygomatic bone. Therefore, the lesion displaces the orbital globe inferiorly (Fig. 1). MRI images of the brain were obtained and revealed a large hypodense lesion (7.2 × 6.3 cm), involving the left frontotemporal

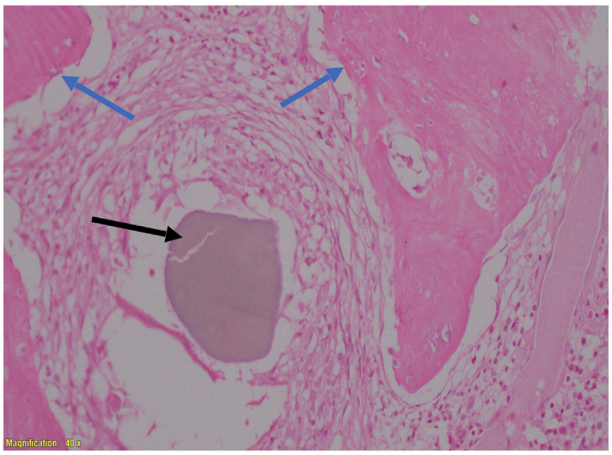

Fig. 3. *Streptomyces somaliensis* grain (black arrow) in marrow space between bone trabeculae (blue arrow). H&E stain ×10HPF.

region with extension to left orbit, left posterior ethmoid air cells, and left temporal suprasellar region (Fig. 2). This lesion compresses and deviates the left eye globe anteriorly and inferiorly. Following the administration of contrast, the lesion showed homogeneous enhancement. Both optic nerves appear normal in course, morphology, and thickness.

Histopathology of the specimen showed multiple pink cracked grains of *Streptomyces somaliensis* surrounded by marked histiocytic and giant cell granulomatous reaction (Fig. 3). The stroma shows prominent fibrous septa. These features are consistent with Actinomycetoma attributed to *Streptomyces somaliensis*.

Under possible aseptic conditions, the patient was put in a supine position and a left orbitozygomatic question mark-shaped incision was performed. After dissecting the temporalis muscle the operator removed a firm not suckable mass from the periosteal surface of the muscle. Then 4 burr holes were performed. The thickness of the bone was 8 cm. After that orbitozygomatic craniotomy was performed. The dura was opened after removal of a thin, non-bloody epidural mass without subdural invasion. Bone was drilled to remove the thickened part. However, the thickness extended up to petrous bone and it was difficult for total removal. Titanium mesh was used for cranioplasty and dura was closed on watertight. Intraoperatively, the patient received 2 units of blood, phenytoin, and mannitol.

The patient was discharged in day 7 postoperatively with medication for 6 weeks, including the following, amoxicillin and clavulanate combination 1 g, 1 tab B.D + cotrimoxazole 480 mg, 2 tabs B.D + folic acid 0.5 mg, 1 tab per day, and referred to the mycetoma center to continue the medical treatment and follow-up.

3. Discussion

The current global burden of mycetoma is not accurately known [5]. However, the disease has an explicit geographical distribution which is commonly known as “mycetoma belt” including South India, Sudan, Senegal, and South America [5]. In 2013, a systematic review and meta-analysis estimated that the prevalence of mycetoma in Sudan was 1.8 cases per 100,000 inhabitants [7]. This prevalence is underestimated because it is based only on published cases to literature in comparison to a cohort study conducted in an endemic village in Sudan, indicating that the prevalence of mycetoma is 6.2 per 1000 inhabitants in that specific village [8].

Mycetoma commonly appears as painless subcutaneous swelling increased in size gradually with the development of secondary nodules that produce drain secreted through sinus tracts [3]. This distinct clinical presentation increases the suspension of the disease. However,

Table 1
Shows the case reports of cranial mycetoma during the past 14 years.

Study	Gender	Age of patients	Causative organism	Location of the lesion	Treatment
Beeram V et al. [13]	Male	18 years	*Madurella mycetomatis*	Lt parietal bone	Surgical + medical (itraconazole and ketoconazole)
Maheshwari S et al. [14]	Male	31 years	*Madurella mycetomatis*	Paranasal sinus + lt cavernous sinus	Surgical + medical (liposomal-amphotericin B)
Shanbhag NU et al. [15]	Male	25 years	*Nocardia brasiliensis*	Scalp + orbit + eye led	Medical (streptomycin + rifampicin + cotrimoxazole)
Gueye NN et al. [16]	Male	17 years	Not mentioned	Ethmoid bone + orbit	Surgical + medical (ketoconazole)

histopathology is required to confirm the diagnosis. Fine needle aspiration cytology is a simple and sensitive test that can be used to identify the causative organism [9]. Soft tissue invasion and bone distractions by mycetoma can be best assets through MRI [10].

Head and neck are extremely rare sites for mycetoma [11]. Because the disease is transmitted from soil to the body through traumatic inoculation [1]. Therefore, the disease generally affects the lower limb followed by hand [1]. In 1950 Beeram et al. [13], reported 3 cases of cranial mycetoma treated at Khartoum Civil Hospital. One patient presented with extensive calvarium destruction and new bone formation. As a consequence, only a biopsy was performed because surgical removal was not applicable. The other two patients presented with mycetoma involving the paranasal sinuses and orbits treated surgically with eye removal in one patient [12]. Conversely, in our case, the enucleation of the affected eye was not performed owing to the normal shape of the eye globe despite the impaired function of the eye, particularly the optic nerve.

After a narrative review of medical literature in the last 14 years, we found 4 case reports of cranial mycetoma (Table 1). *Madurella mycetomatis* was reported as a causative organism of Cranial mycetoma in 2 different case reports [13,14], while *Nocardia brasiliensis* was mentioned in one report [16]. Orbital involvement was reported in 2 cases [15,16]. 3 patients were treated by surgical removal of the lesions followed by medical treatment. However, only one patient was treated medically by intravenous streptomycin 80 mg and cotrimoxazole for 6 weeks followed by rifampicin 600 mg and cotrimoxazole tabs without surgical intervention [15].

4. Conclusion

Mycetoma is a tropical disease that presents with gradual swelling contains nodules commonly involving the lower extremity. The disease can be caused by bacteria (Actinomycetoma) or fungi (Eumycetoma) after transmission to the body through traumatic inoculation. Cranial mycetoma is an extremely rare entity of the disease. MRI is required to identify soft tissue involvement and bone distraction. Early diagnosis and prompt surgical and medical treatment are the keys to good outcomes.

Consent

Written informed consent was obtained from the patient for publication of this case report and attached images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Ethical approval

This study was excepted from ethical approval by the ethical committee of The National Ribat University, Khartoum, Sudan.

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CRediT authorship contribution statement

Tarig Fadalla: Involved in study design, data acquisition, drafting the article, revising it critically and finally approved the manuscript.
Mohammed Hamed: Involved in conception of the study design, drafting the article and finally approved the manuscript.
Nahla Elnayir: Involved in conception of the study design, drafting the article and finally approved the manuscript.
Mujahid Imam: Involved in conception of the study design, drafting the article and finally approved the manuscript.
Emad Ibrahim: Involved in the design of the study, revising it critically and finally approved the manuscript.

Declaration of competing interest

No conflict of interest reported by authors.

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