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Case Report

Presentation of Invasive Fungal Rhinosinusitis in Sudanese Children: A Report of Four Cases

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Abstract

Fungal rhinosinusitis (FS) is considered as a disease spectrum that ranges from allergic fungal sinusitis (AFS) and chronic fungal rhinosinusitis (CFS) to invasive fungal rhinosinusitis (IFS) invading the orbit, Dura, and intra-cranium. Fungal rhinosinusitis is a common disease in Sudan. **Objective:** To present four rare cases of Invasive Fungal Rhinosinusitis in Sudanese children presented with orbital extension. **Patients and Method:** The authors have reported four cases of children in the age range of 9–11 years, two girls and two boys; they were presented with invasive fungal rhinosinusitis in Africa ENT hospital (Sudan) during the period from September 2015 to August 2017. Patients' diagnosis was made by endoscopic examination, CT, and MRI, and it was confirmed by tissue biopsy.**Results:** Patients' age range was 9–11 years, two girls and two boys. The unilateral disease was the commonest type. An orbital extension was detected in all cases on CT and MRI. No intracranial extension was detected. *Aspergillus flavus* was the most responsible agent detected in all the four cases.**Conclusion:** Fungal rhinosinusitis (FS) is a common disease in adult Sudanese patients but rarely affects children. It is mainly caused by *Aspergillus* species. The disease is extensive and can be associated with orbital and intracranial complications.

Keywords: fungal rhinosinusitis, children, Sudan

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1. Introduction

The etiology of chronic rhinosinusitis (CRS) is unclear. It has been recently suggested that a Fungus-mediated process is the primary cause of CRS with and without polyps [1].

Fungal infections of the nose and sinuses appear to be increasingly common and invade eye structures [2, 3].

As it can be difficult to confirm the presence of fungi, both histologically and by culture, the radiological features may be of paramount importance in initiating a careful search for fungal elements [3].

Classification of fungal rhinosinusitis (FS) into invasive and non-invasive depends mainly on the presence or absence of fungal hyphae within the mucosa, and not on the presence of bone erosion, which can be found in non-invasive forms [4, 5].

In Sudan, the condition was first noticed by Sanderson et al. when they reported a granuloma resembling Aspergillosis granuloma in biopsy material from three patients with proptosis [6].

Bella et al. in 1973 reported 46 cases of primary paranasal Aspergillosis granuloma seen in Sudan [7, 8].

This disease mainly affects age groups between 11 and 50 years, and it comes from different localities in Sudan. The symptoms were mainly nasal obstruction, headache, rhinorrhea, external ethmoid swelling or cheek swelling and proptosis [6, 9].

Recently, a chronic destructive form of paranasal sinus mycoses characterized by sinus expansion and bony erosion was described [9].

Paranasal Aspergillosis seems to be a rare disease in the world in general, but is relatively common in Sudan [9, 10].

Mahgoub declared that mycetoma mycoses in Sudan are significant health problems and pointed out the importance of paranasal *Aspergillus* granuloma [11, 12].

2. Patients and Methods

During the period from September 2015 to August 2017, a total of four patients with suspected invasive fungal rhinosinusitis (IFS)—two girls and two boys—were selected. The prospective analysis was done in Africa ENT hospital in Khartoum Sudan, where functional endoscopic sinus surgery (FESS) was performed on all kids.

Fungal cultures, serology, and microscopy to detect fungal elements in the nasal cavity were carried out for all patients. Patients were considered fungus-positive if at least one of these methods could demonstrate fungal elements. Specimens for histopathology and cultures were taken postoperatively, and serology was performed for all patients.

3. Results

All four patients were diagnosed as FS, and were presented with sinonasal polyposis and neuro-orbital complications. CT and MRI were done for all the four patients.

Case 1

Female, from Port-Sudan, 9 years old, weight 23 kg, had no history of asthma. Investigations include:

CBC, Urine analysis was normal;

Serology for aspergillosis was strongly positive with *Aspergillus flavus* and in;

Culture/sensitivity direct = fungal hyphae seen, culture = *Aspergillus flavus*;

Rt eye proptosis/Rt nasal polyps/lt nasal septal deviation.

CT, MRI findings: Metallic sign in the sinuses. FESS was done. The patient started itraconazole caps 100mg for one year with regular follow-up with blood investigation and nasal endoscopy.

Case 2

Female, aged 10 years, weight 24 kg, from Port-Sudan and had no history of asthma.

The patient was presented with nasal obstruction for one year and Lt Eye proptosis, endoscopy showed; Extensive nasal polyposis.

CT, MRI findings: Sinuses were full of fungi.

FESS Findings: lt polyposis, muddy fungal material from maxillary, ethmoidal sinuses and sphenoid were cleaned. Pulsatile, exposed posterior skull base dura, was preserved, and the orbital cavity was cleaned from the fungal muddy material Lt eye was decompressed.

Cases 3 and 4

Both were male, aged 8 and 9 years, respectively, weighing 20 and 24 kg, respectively, having no history of asthma. Investigations include:

CBC, Urine analysis, and CXR were normal;

Serology for aspergillosis was positive with *Aspergillus flavus* and in;

Culture media, fungal hyphae were seen.

CT, MRI findings: Sinonasal masses and the metallic sign were seen in the sinuses. FESS was done for both kids, and they started itraconazole caps 100mg for 1 year with regular follow-up.

No.	Sex	Age	C/S	Histopathology	S/S	Diagnosis	Drugs
1	Male	8	*A. flavus*	Mucosal invasion	–Nasal obstruction – Proptosis –Headache	CT MRI	Itraconazole caps + Isotonic solution spray
2	Female	9	*A. flavus*	Septate fungal hyphae was seen	–Nasal obstruction – Proptosis –Headache	CT MRI	Itraconazole caps + Nasal spray
3	Female	10	*A. flavus*	Mucosal invasion	–Nasal obstruction – Proptosis –Headache	MRI CT	Itraconazole caps + Rhinocort spray
4	Male	9	*A. flavus*	Mucosal invasion	–Nasal obstruction – Proptosis –Headache	MRI CT Serology	Itraconazole caps + Flixonase spray

Table 1: The result of age, sex, culture/sensitivity (C/S), histopathology, symptoms /signs (S/S), images and drugs options.

Symptoms/Signs	No.	%
Nasal Obstruction	4	100
Nasal Mass	4	100
Anosmia	3	75
Postnasal Drip	3	75
Cacosmia	3	75
Proptosis	4	100
Headache	4	100
Nasal Discharge (Greenish Secretions)	4	100

Table 2: Clinical presentation.

4. Discussion

The diagnostic criteria for AFRS vary among authors, but the most widely accepted are the five criteria described by Bent and Kuhn. To diagnose AFRS, the presence of allergic mucin in histopathology specimens is important in addition to the demonstration of fungal elements [13].

Aspergillus species were found to be the common species of fungi causing rhinosinusitis in Sudan, and this is in agreement with previously published Sudanese series [9, 12, 14].

No gender was predominant in childhood, but in the adult, female was found to be predominate; this is in a harmony with the majority of the national and international studies [1, 2, 6–10, 15].

Complications included orbital, and intracranial extensions were found in two patients (50%), which agrees with a group of authors [3, 5, 6, 9, 16].

In this study, two kids started their disease as a non-invasive type and changed to an invasive type during the study period; this also goes with Thacker et al. who recommended that fungal sinusitis should be considered a potentially progressive continuum, where the non-invasive disease may coexist with an invasive form [17].

Aspergillus fumigatus was considered the primary etiologic agent of AFRS cases and was found as a predominant etiologic agent in Western literature [18, 19]. However, in contrast, *A. flavus* is the predominant etiologic agent causing chronic fungal rhinosinusitis (CFS) in Sudan. In the international literature, mucormycosis is the most implicated fungus for invasive fungal sinusitis (IFS) [9, 12, 14–16].

5. Conclusion

Fungal rhinosinusitis (FS, CFS, AFS, and IFS) is a common disease in Sudan that mostly affects females of young age groups and is rare among kids. The usual presentation is a nasal mass or polyp and can be associated with orbital and intracranial invasion. The main causative agents are *Aspergillus* species with *A. flavus* being the most commonly encountered.

Recommendation

Fungal rhinosinusitis (FS, CFS, AFS, and IFS) is a common disease in Sudan but rarely affects kids. More studies and efforts must be done to look for best methods of management.

Conflict of Interests

The authors declare no conflict of interests.

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