

Proptosis in Children Caused by Sinonasal Pathology: A Retrospective Study

Original Article

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ABSTRACT

Background: One of the most frequent clinical signs of orbital pathology is proptosis. Because of their close anatomical relationships, para nasal sinus or nasal pathologies can exhibit proptosis.

Purpose: To report the 10-years experience of a tertiary medical center (Kasr Elainy hospital-Cairo University) with children presenting with proptosis due to nasal or sinus lesion.

Patients and Methods: Patients were identified by file review. Data were collected on demographics, findings on ophthalmologic and imaging evaluations, etiology, treatment, and outcome.

Results: The study included 209 patients suffering from proptosis due to nasal or sinus pathology. Age of the patients ranged from 40 days to 16 years. Complicated acute sinusitis with orbital cellulitis, subperiosteal or orbital abscess and allergic fungal sinusitis constituted 93.3% of the presenting cases. Allergic fungal sinusitis and complicated sinusitis with subperiosteal abscess showed statistically-significant association with necessity of surgical intervention; while complicated acute sinusitis with orbital cellulitis showed statistically-significant higher proportion of complete resolution on medical treatment. Both “acute invasive fungal sinusitis” and “Lymphoma” showed statistically- significant association with vision non-improvement. While complicated acute sinusitis with orbital abscess showed statistically- significant higher proportion of vision improvement.

Conclusion: Many sinonasal lesions can present with proptosis in children. Inflammatory causes were the most common causes of proptosis in children in our study. Proptosis needs to be evaluated completely for proper management and to limit the morbidity associated with it.

Key Words: Children, nasal lesions, proptosis, sinus lesions.

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INTRODUCTION

Proptosis is the protrusion and anterior displacement of one or both orbital globes. Chemosis, periocular edoema, nystagmus, discomfort, and visual loss may accompany proptosis, and it can be acute or chronic^[1].

Proptosis is the result of a disease encroaching on the orbit and increasing the orbital volume. Axial globe displacement is caused by any increase in orbital volume, which is typically caused upon with mass lesions, vascular anomalies, inflammatory processes, or endocrine lesions.

Proptosis direction is significant because it reveals the location of the lesion. Because proptosis is an early and occasionally the only sign of paranasal sinus or nasal pathology, it must be carefully evaluated by otolaryngologist. By looking at the proptosis direction; we can identify the lesion site^[2].

Because of its near proximity to the nose and paranasal sinuses, the orbit is frequently involved in sinonasal

disorders. Acute rhinosinusitis can cause orbital problems, which are well-known and documented in textbooks. There are numerous other diseases that affect the nose and paranasal sinuses. Some of them can cause visual loss and possibly be life threatening. In patients with sinonasal pathology, orbital symptoms may predominate. The only complaint of such patients is rarely visual disturbance or proptosis^[3].

Only a few cases of unilateral or bilateral proptosis are sent to the E.N.T. department for assessment and treatment. The foramen, fissures, vascular, erosion, and compression of the anatomical barriers have all been implicated in the transmission of sinus or nasal pathology to the orbit^[4].

Proptosis in children has a broad differential diagnosis and warrants a thorough and systematic workup. The common etiologies, presentation, diagnostic steps, evaluation, and treatment of proptosis are different between adults and children^[5].

The role of radiological investigations in the diagnosis of cases of proptosis is extremely important. Proptosis can usually be corrected with early diagnosis and therapy^[6].

The main objective of the present study was to enlist diagnoses of all the children with sinonasal disease, in which orbit was also involved unilaterally or bilaterally. Diagnosis of the aetiology was based on histopathological confirmation supported by clinical, laboratory, and radiological evidence. Management strategy and final outcome in all the cases was also analysed.

PATIENTS AND METHODS:

A retrospective cohort study design was used. The databases of the otolaryngology clinic of ENT department in the faculty of medicine, Cairo university were searched for all patients less than 18 years old who presented with proptosis due to nasal or sinus lesion from January 2010 till December 2020 and were followed for at least 1 year and the study was approved by the ethics committee of the faculty of medicine, Cairo university. A written consent obtained from participants of the study.

Exclusion criteria:

Patients with proptosis due to trauma, hemorrhage, thyroid disease and orbital space occupying lesions were excluded.

Data on demographic characteristics, etiology, and eye evaluation at diagnosis and end of follow-up, imaging studies, treatment, and outcome were collected from the medical files.

Data management and analysis

The data was presented as parameters, tables, and graphs that were analyzed after computerization by using a statistical package for social science (SPSS). All the collected data were revised for completeness and logical consistency. Pre-coded data was introduced in the computer using Microsoft Office Excel Software Program 2019. Then transferred to the Statistical Package of Social Science Software program, version 26 (SPSS) to be statistically analyzed.

- For quantitative variables were described as mean, standard deviation, median, IQR and compared using the Mann Whitney U test, where the p-value is significant if less than 0.05.

- For qualitative variables, they were described as frequency and percentage and compared using Chi-square test or Fisher exact test accordingly where the p-value is significant if less than 0.05.

RESULTS:

3.1. Descriptive characteristics of the patients and their medical condition:

The study included 209 patients suffering from proptosis due to nasal or sinus pathology. Age of the patients ranged from 40 days to 16 years, with median of 6 years and inter-quartile range (3 to 10) years.

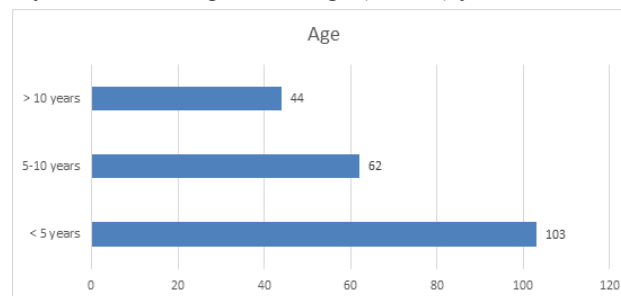


Fig. 1: Age distribution of patients.

Age distribution of patients: the bar graph shows the number of patients in 5-year age groups. All patients greater than 10 years old at the year of reporting are aggregated.

Table 1: Characteristics of study participants (n=209):

		Frequency	Percent
Gender	Male	126	60.3
	Female	83	39.7
Laterality	Right eye	116	55.5
	Left eye	93	44.5
Imaging done	CT	199	95.2
	CT and MRI	10	4.8

Almost 2/3 of the patients were males and in more than 1/2 of patients the right eye was affected. Computed tomography (CT) was the most common radiological investigations needed in our patients as it was done for all the patients and in 4.8% of our patients magnetic resonance imaging (MRI) was ordered in addition to CT.

Table 2: Frequency of pathological variations (n=209)

Pathology	Frequency	Percent
Complicated acute sinusitis with orbital cellulitis	86	41.1
Complicated sinusitis with sub periosteal or orbital abscess	76	36.4
Allergic fungal sinusitis	33	15.8
Angiofibroma	4	1.9
Acute invasive fungal sinusitis	3	1.4
Complicated acute sinusitis with CST*	3	1.4
Lymphoma	2	1.0
Intraorbital hydatid cyst	1	0.5
Rhabdomyosarcoma	1	0.5

*CST= cavernous sinus thrombosis.

Complicated acute sinusitis with orbital cellulitis, complicated sinusitis with subperiosteal or orbital abscess and allergic fungal sinusitis constituted 93.3% of the presenting cases. The other less common causes include angiofibroma, acute invasive fungal sinusitis, and complicated sinusitis with cavernous sinus thrombosis (CST), lymphoma, hydatid cyst and rhabdomyosarcoma as shown in (Table 2).

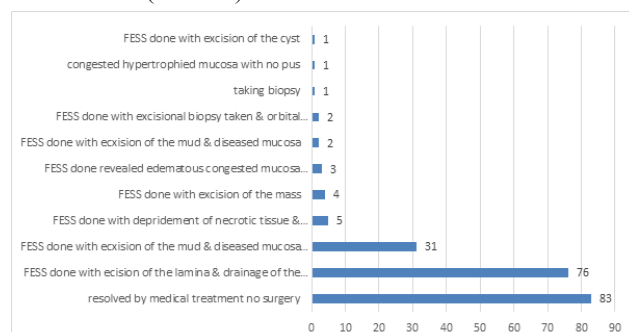


Fig. 2: Bar graph showing the intervention conducted among patients.

About 40% of cases showed resolution on medical treatment without surgical intervention as shown in (Figure 2).

Table 3: Clinical findings during Follow-up visit (n=209)

	Frequency	Percent
Complete resolution/no evidence of recurrence till now	183	87.6
Recurrence happened with no proptosis	17	8.1

Table 5: Pathological types associated with presence of complications (n=209)

Pathology	complications		Total	p-value
	No (n=196) N(%)	Yes (n=13) (%)		
Acute invasive fungal sinusitis	0	3 (23.1)	3 (1.4)	< 0.001*
Allergic fungal sinusitis	31(15.8)	2 (15.4)	33 (15.8)	1*
Angiofibroma	0	4 (30.8)	4 (1.9)	<0.001
Complicated acute sinusitis with CST	3 (1.5)	0	3 (1.4)	1*
Complicated acute sinusitis with orbital cellulitis	86 (43.9)	0	86 (41.1)	0.005
Complicated sinusitis with sub periosteal orbital abscess	72 (36.7)	4 (30.8)	76 (36.4)	0.77*
Intraorbital hydatid cyst	1 (0.5)	0	1 (0.5)	1*
Lymphoma	2 (1)	0	2 (1)	1*
Rhabdomyosarcoma	1 (0.5)	0	1 (0.5)	1*

P-value <0.001 denoting statistically-significant relation between pathology and complications, Cramer's V = 0.731 (large effect)

Acute invasive fungal sinusitis and angiofibroma showed statistically-significant higher probability of complications.

Complicated acute sinusitis with orbital cellulitis showed statistically-significant least probability of complications.

Patient referred to clinical oncology	3	1.4
Patient died	2	1.0
Residual tumour around the carotid	1	0.5
Residual tumour needed revision surgery	1	0.5
Complete removal	1	0.5
Patient is blind	1	0.5

During follow-up complete resolution of proptosis with no recurrence of the disease was achieved in 183 patients (87.6%) and recurrence happened but without proptosis in 17 patients (8.1%) as shown in (Table 3).

Only 13 (6.2%) patients suffered from complications as shown in (Table 4).

Table 4: complications encountered by patients (n=209)

	Frequency	Percent
No complications	196	93.8
Bleeding need blood transfusion	4	1.9
Medial rectus injury	4	1.9
Blindness	3	1.5
CSF LEAK	2	1.0

3.2. Factors affecting complications:

There is statistically significant association between allergic fungal sinusitis and recurrence of the disease 51.5% (however no proptosis occurred) upon follow-up visit, *p-value* < 0.001, Cramer's V = 0.689 (large effect)

Intraorbital hydatid cyst showed complete resolution.

Patients with lymphoma and rhabdomyosarcoma were referred to clinical oncology.

3.3. Factors promoting complete medical resolution (without surgical intervention):

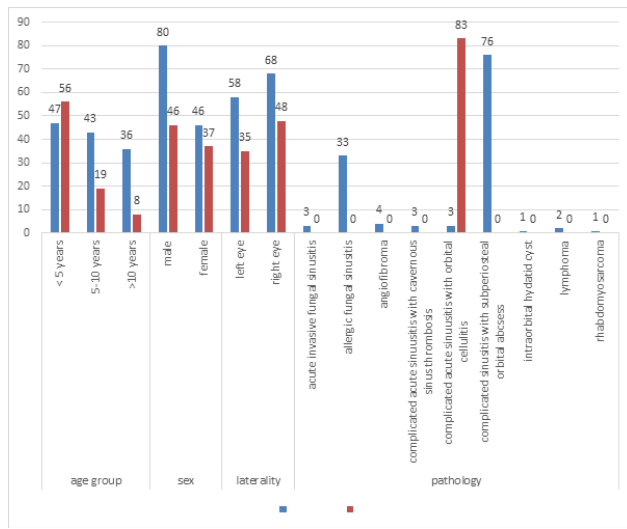


Fig. 3: Bar graph illustrates the relation between different factors and resolution on medical treatment (blue columns refer to pt need surgery and red columns refer to pt treated medically).

The odds of needing surgical intervention among “5-10 years” and “>10 years” age groups are respectively 2.7 and 5.3 times the odds of younger age group “<5 years” as shown in (Figure 4).

Allergic fungal sinusitis and complicated sinusitis with subperiosteal abscess showed statistically-significant

association with necessity of surgical intervention; while complicated acute sinusitis with orbital cellulitis showed statistically-significant higher proportion of complete resolution on medical treatment as shown in (Figure 3).

3.4. Improvement in visual acuity:

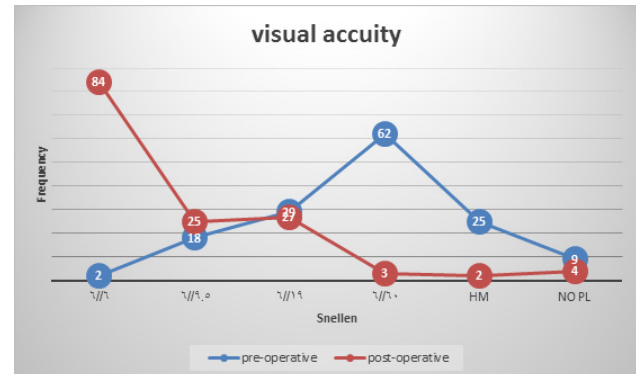


Fig. 4: Line graph illustrate the visual acuity among patients pre- and post- operative (n=145).

In about 31% (64/209) the visual acuity couldn’t be assessed, there is improvement of visual acuity in 58% (122/209) and about 11%(23/209) the visual acuity did not improve.

Wilcoxon-signed rank test revealed significant difference between LogMAR visual acuity before and after intervention (*p-value* <0.001) with median & IQR before and after treatment 1 (0.5 & 1) & 0 (0 & 0.3) respectively.

Table 6: Factors affecting improvement of vision among patients

	Improved vision		Total	<i>p-value</i>	OR (95% CI)
	No, n=23 N(%)	Yes, n=122 N(%)			
Age group					
< 5 years	5 (21.7)	34 (27.9)	39	0.798	
5-10 years	10 (43.5)	52 (42.6)	62		
>10 years	8 (34.8)	36 (29.5)	44		
Gender					
Male	13 (56.5)	72 (59)	85	0.824	0.9 (0.37-2.2)
Female	10 (43.5)	50 (41)	60		
Laterality					
Left eye	7 (30.4)	59 (48.4)	66	0.113	0.47 (0.18-1.2)
Right eye	16 (69.6)	63 (51.6)	79		
Pathology					
Acute invasive fungal sinusitis	3 (13)	0	3	0.004*	41.8(2.1-840)
Allergic fungal sinusitis	5 (21.7)	28 (23)	33	0.899	1.07 (0.37-3.1)
Angiofibroma	0	4 (3.3)	4	1*	0.84 (0.78-0.9)
Complicated acute sinusitis with CST	0	3 (2.5)	3	1*	0.84 (0.78-0.9)
Complicated acute sinusitis with orbital cellulitis	4 (17.4)	49 (40.2)	53	0.038	0.31(0.1-0.98)

Complicated sinusitis with subperiosteal orbital abscess	9 (39.1)	36 (29.5)	45	0.36	0.65 (0.26-1.6)
Intraorbital hydatid cyst	0	1 (0.8)	1	1*	0.8 (0.78-0.9)
Lymphoma	2 (8.7)	0	2	0.024*	28.5(1.3-614.2)
Rhabdomyosarcoma	0	1 (0.8)	1	1*	0.84 (0.78-0.9)

Both “acute invasive fungal sinusitis” and “Lymphoma” showed statistically- significant association with vision non-improvement, with odds ratio 41.8 and 28.5 respectively.

While complicated acute sinusitis with orbital abscess showed statistically- significant higher proportion of vision improvement.

DISCUSSION

Many sinonasal diseases may present by orbital manifestations. Proptosis or exophthalmos can be a sign of a disastrous pathology, not only primarily of the eyes but also of the other neighbouring structures e.g., that of nose and paranasal sinuses. The protrusion of the eye ball can be regarded as a protective mechanism against a raised intra orbital pressure. Infections, inflammations and neoplasms of the nose and paranasal sinuses constitute important causes of proptosis^[3].

The present study reports interesting information that describes the cumulative 10-year experience of a large tertiary medical center with proptosis of a sinonasal cause in children.

To our knowledge and after reviewing the literature, our study was the first one that discusses and investigates causes of proptosis due to sinus or nasal lesions in children. There was only some studies were conducted on proptosis due to sinus or nasal lesions on adult patients.

Our study included 209 patients suffering from proptosis due to nasal or sinus pathology. Age of the patients ranged from 40 days to 16 years. The highest incidence of proptosis secondary to sinonasal disorders was noted in male patients, accounting for 126 cases (60.3%). In female patients, it accounted for 83 cases (39.7%). This is similar to the data presented in the study by Sinha *et al*, 2005. The male to female ratio was 2:1 in Sinha *et al*, 2005 study^[4].

In the present study, complicated acute sinusitis with orbital cellulitis, complicated sinusitis with subperiosteal or orbital abscess and allergic fungal sinusitis constituted 93.3% of the presenting cases.

A previous study Yousry Al Sayed, 1995 in his study on 28 patients found that 9 of his patients had bacterial sinusitis and 8 were diagnosed to be having allergic fungal sinusitis and this in agreement with our study as inflammatory causes were the most common aetiology of proptosis in children^[7].

Another previous study Venugopal and Sagesh, 2013 stated that sinonasal malignancies were the most common pathologies found in their study and this in contrast to our study in which sinonasal malignancies constituted 1.5% of the presenting cases. This difference may be explained by the different age group as in venugopal study most of the patients were from the age group 41-50 years. Fungal sinusitis was found in only 15.9% of the patients and this in agreement with our study^[2].

Revising the literature, Samil *et al*, 2015 in their results stated that mucocele was the most common sinonasal disease that presented as proptosis in their patients^[8].

In our study, complicated sinusitis (orbital cellulitis or subperiosteal abscess) constitutes about 77.5% of sinonasal causes of proptosis in children. Orbital extension is the most common complication of ethmoid sinusitis. In order to avoid orbital or intracranial complications, the American Academy of Paediatrics advises prompt antibiotic prescription for sinusitis. If therapy is started as soon as possible, there is a strong chance that the orbital complications of ethmoiditis will improve^[9].

In our study allergic fungal sinusitis constitutes about 15% of causes of proptosis in children. Allergic fungal sinusitis is a disease most commonly affects immuno-competent patients. Sometimes it can cause disfiguring telecanthus and proptosis. Massive polyposis and allergic fungal mucin, popularly referred to by otorhinolaryngologists as "fungal debris," are present in these cases. Following clinical and radiological diagnosis, treatment is initiated. Proptosis can sometimes recover sooner, but the cosmetic deformity including telecanthus takes some months or years to resolve^[10].

The radiological diagnosis is made with a plain CT scan, which reveals opacification of the paranasal sinuses coupled with the expansile effect of the disease on the lamina papyracea. The hyperintense shadows on the soft tissue CT films that are indicative of a fungus infection. Sometimes MRI is necessary to rule out intra axial cerebral disease extension. Visual loss, if present, is caused by involvement of the optic nerve^[11].

It is noteworthy to state that acute invasive fungal sinusitis comprises 1.4% of all pathologies in our cases. Taken in to account, this type of sinusitis is an invasive life threatening disease found in immunocompromised Patients. It carries poor prognosis due to underlying

debilitated condition of the patient and the aggressive nature of the disease. This was also detected by Biswas *et al*, 2013 in their study^[12].

Early loss of vision with fixed and frozen eye is the early signs of acute invasive fungal sinusitis disease. Treatment should ideally be started based upon clinical suspicion sometimes supported by radiological evidence. Emergency surgical debridement is done in our department in all the cases of clinically diagnosed cases of mucormycosis. Endoscopic approach is used in all these cases. An important factor in the treatment is control of the underlying disease e.g. diabetes mellitus. In addition, intravenous Amphotericin B is given to all these patients.

Importantly, computed tomography (CT) evaluation is considered the best diagnostic modality to assess the disease process (Mahsud and Bano.^[10]). It is therefore CT was the most common radiological investigations needed in our patients and it was done for all the patients. In addition, 4.8% of our patients' magnetic resonance imaging (MRI) was ordered in addition to CT.

Regarding treatment, about 40% of our cases showed resolution on medical treatment without surgical intervention and in the remaining 60 % surgery was needed. The odds of needing surgical intervention among “5-10 years” and “>10 years” age groups, respectively, were 2.7 and 5.3 times the odds of younger age group “<5 years”. In our study, Allergic fungal sinusitis and complicated sinusitis with subperiosteal or orbital abscess usually treated surgically by endoscopic approach; while complicated acute sinusitis with orbital cellulitis showed good response to medical treatment. Complicated acute sinusitis with orbital abscess showed significant improvement in the visual acuity after treatment while there is no improvement in the vision in cases of acute invasive fungal sinusitis and lymphoma.

In the present work, the incidence of complications was higher in cases of acute invasive fungal sinusitis and angiofibroma and less common in cases of orbital cellulitis. The odds of suffering complications among females is 11.4 times the odds of suffering complications among males; while the odds of having complications among age groups “5-10 years old” and “>10 years old” is 5.9 and 10.5 times that of younger age group “<5 years old”.

Collectively, the present study included all patients in the defined population were from a single paediatric clinical centre where all the patients were examined by both paediatric and ENT clinicians and the relatively large number of patients included in our study. Meanwhile, the main limitations of our study were the observational nature and the retrospective aspect of data collection.

CONCLUSION

The present study concluded that many sinonasal lesions can present with proptosis in children. Sinonasal causes of proptosis in children might show different incidence from adult as inflammatory causes were the most common causes of proptosis in children in our study. Proptosis needs to be evaluated completely for proper management and to limit the morbidity associated with it.

Clearly, it is essential for an otorhinolaryngologist and ophthalmologist to thoroughly evaluate the proptosis and work as a team to diagnose and treat this disorder to prevent deterioration of vision as well as to address the cosmetic problem.

CONFLICT OF INTEREST

There are no conflicts of interest.

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