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RESEARCH ARTICLE **OPEN ACCESS**

HEREDITARY WOAKES SYNDROME; OUR EXPERIENCE

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ABSTRACT

Chronic rhinosinusitis (CRS) with or without nasal polyp is one of the most prevalent chronic disease worldwide, causing a significant impact on quality of life. CRS is broadly divided into CRS with nasal polyps (CRSwNP) and without nasal polyps (CRSsNP) (1).CRSwNP represents a more severe medical condition. Woakes described a rare form of nasal polyposis which, arises in childhood and produces severe nasal deformity and necrosis of ethmoidal cells. in 1885.B. Kellerhals and De Uthemann defined Woakes syndrome as severe recurrent infantile nasal polyposis with broadening of the nasal pyramid causing pseudohypertelorism, frontal sinus aplasia, bronchiectasis and dyscrinia.Herewith we are presenting three siblings who came with nasal polyposis and facial deformity with nasal obstruction. We did FESS and Rhinoplastysurgery and successfully treated them without any complications. **Case Report**: 17 year old male came with complaints of nasal obstruction and progressive deformity with broadening of nasal bridge, recurrent rhinitis, since 3 years & hyposmia and hyponasal voice since 1year. Second patient was15 year old female, sister of case -1, came with similar complaints with severe broadening of nasal bridge, recurrent rhinitis, hyposmia and hyponasal voice since 1year. Third patient was 13 year old female sibling came with complaints bilateral nasal obstruction, recurrent rhinitis, slight nasal deformity, hyposmia and hyponasal voice for 6 months. Compared to her siblings she only had minimal nasal deformity (Figure.1). on anterior rhinoscopy polypoidal mass seen in both the nostrils (Figure.2).CT scan of paranasal sinuses and nose (PNS) revealed nasal polyposis occupying bilateral nasal cavity with resultant maxillary, frontal, sphenoid and ethmoid sinusitis (Figure. 3) High resolution CT thorax showed bilateral symmetrical central bronchiectasis with upper lobe predominance (Figure.4). All three of them were operated. FESS and Rhinoplasty surgery were done. Intraoperative findings were showing thick tenacious secretions and fungal concretions (Figure.5). In all three cases bilateral ethmoidal, spehoid , frontal and Maxillary sinuses were full of polypoidal mass . Complete clearance of all sinuses was done (Figure .6) & good frontal glow was achieved Figure.7). Postoperatively they were given oral steroids for15 days along with nasal douching and nasal steroidal spray was given for three months. Initially azelastine & fluticazone steroidal spray was given for one month. And then only fluticazone steroidal nasal spray was given for next 2 months. They were followed at interval of onw week, one month, 3 months and 6 months. Every follow up check nasal endoscopy was done. They all had good outcome without any complications. **Conclusion:** Woakes syndrome is a clinical condition characterised by a tetrad of recurrent nasal polyposis, nasal pyramid deformity, frontal aplasia or hypoplasia and bronchiectasis. Etiopathogenesis of this syndrome still remains unclear due to its rarity and sparse documentation. Non-inherited unknown genetic factor may be a possible etiology, and needs further detailed genetic studies. As these cases are usually refractory to medical management, meticulous surgery in the form of Functional endoscopic sinus surgery forms the mainstay of the treatment with regular post-operative care for prolonged duration carrying equal importance. Rhinoplasty surgery helps the patient to give cosmetic results and better face. Furthermore, detailed study over a prolonged period is required in such patients to comment upon specific duration of follow-up required.

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INTRODUCTION

Chronic rhinosinusitis (CRS) with or without nasal polyp is one of the most prevalent chronic disease worldwide, causing a significant impact on quality of life. CRS is broadly divided into CRS with nasal polyps (CRSwNP) and without nasal polyps (CRSsNP) (1).CRSwNP represents a more severe medical condition. Nasal polyps are severe eosinophilic inflammation of the upper airways characterised by response to medical management and frequent recurrences (2).With advancements and availability of endoscopic evaluation and computerized tomography (CT) scans, an early diagnosis of nasal polyp can be made easily. Most of them are treated medically with antibiotics and topical and/or systemic corticosteroids. Some refractory cases requiresurgical management in the form of functional endoscopic sinus surgery (FESS). Due to the complex nature of the etiology and pathophysiology of CRSwNP in children, they usually pose an enigma to otorhinolaryngologists. Nasal polyposis in paediatric population demands a meticulous search for etiology. Some of the common etiologies linked to nasal polyposis in children areallergy, cystic fibrosis or Kartagener's syndrome (3). Woakes described a rare form of nasal polyposis which, arises in childhood and produces severe nasal deformity and necrosis of ethmoidal cells in 1885.B. Kellerhals and De Uthemann defined Woakes syndrome as severe recurrent infantile nasal polyposis with broadening of the nasal pyramid causing pseudohypertelorism, frontal sinus aplasia, bronchiectasis and dyscrinia. It is usuallyfound to have a hereditary origin (4). Till date only few such cases have been documented in the literature. Here we describe a case series of Woakes syndrome in three siblings which has never been previously reported.

Fig 1. showing the clinical picture with broadening of Nose

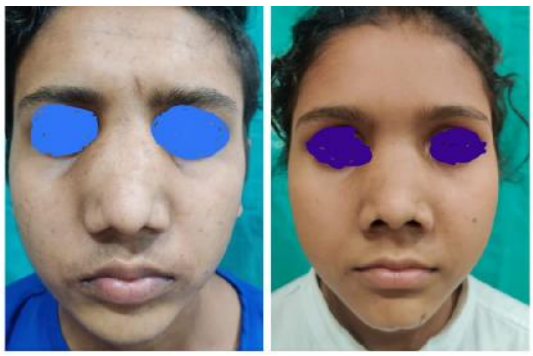

Figure 2. Showing the polypoidal nasal mass on endoscopy

Case report: Three siblings hailing from Central India belonging to middle class family were encountered in outpatient clinic of Department of ENT at Seth G S medical college and King Edward Memorial hospital with similar complaints of progressive nasal blockage and nasal deformity of varying duration.

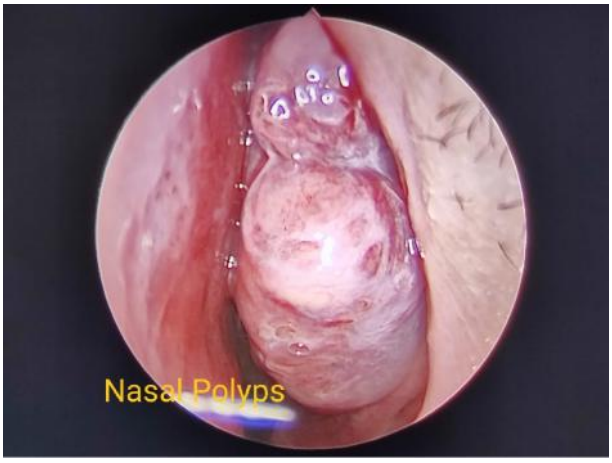

Figure 3. Showing Ctscan images of paranasal sinuses with extensive polyposis

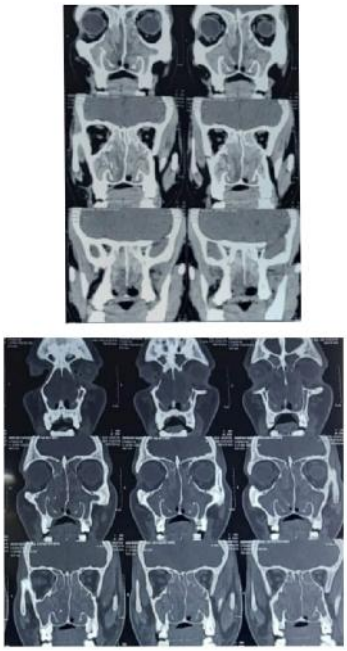

Figure 4. Showing High resolution CT scan images of Thorax showing bronchiectesis

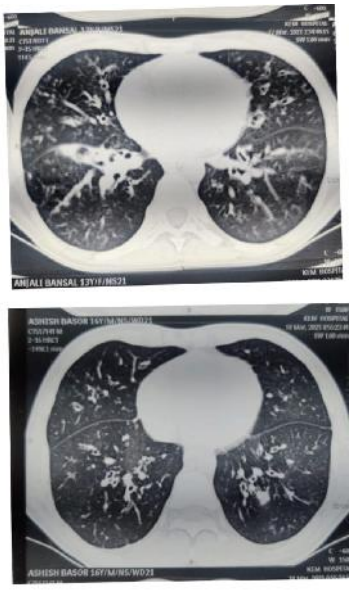

Figure 5. Showing thick tenacious nasal secretions

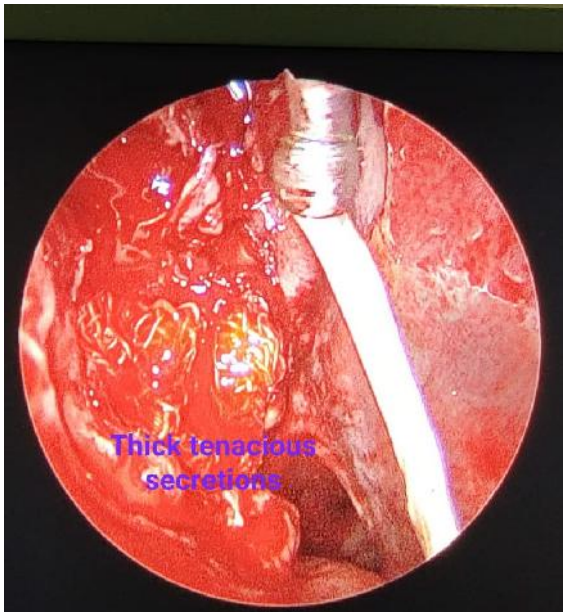

Figure 6. showing itraoperatice widely opened frontal sinus with clearanceof disease

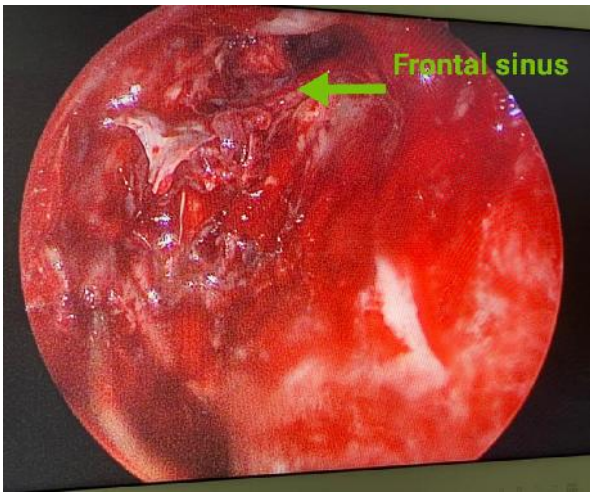

Figure 7. Showing Frontal Sinus Glow

Case 1- 17 year old male from central India, came to ENT outpatient department with complaints of nasal obstruction and progressive deformity of nose since 3 years Patient gave positive clinical history of hyposmia and hyponasal voice since 1year. There is no history of allergy or trauma. On further questioning, patient does not give history of any abdominal complaints.Patient gave history of similar complaints in the past, 5 years back, for which he received multiple course of antibiotic and topical steroid nasal spray. Healso underwent FESS surgery for the same 2 years back which led to temporary relief of symptoms for few months. On clinical examination, there was broadening and enlargement of the nasal pyramid ((Figure 1 frog nose deformity). Anterior rhinoscopy revealed multiple polypoid mass filling bilateral nasal cavity.On diagnostic nasal endoscopy bilateral polyposis with thick viscid secretion causing widening of nasal cavity seen(Figure 2).CT-PNS revealed nasal polyposis occupying bilateral nasal cavity with resultant maxillary, frontal, sphenoid and ethmoid sinusitis(Figure 3).. High resolution CT thorax showed bilateral symmetrical central bronchiectasis with upper lobe predominance. (Figure4). Patient was given a trial of medical management in the form of antibiotics and topical nasal steroidal sprays at our centre for a period of 2 months with poor response. Following which we performed revision FESS for this patient. Polypoid tissue was biopsied and sent for the histopathological examination. Intra-operatively bilateral hypo plastic frontal sinusand dehiscent lamina papyracea were noted.Post-operative histopathologicalreport was suggestive of inflammatory benign nasal polyp with cellular infiltrates predominantly composed of lymphocytes, plasma cells and neutrophils with few eosinophils. Patient was followed up in outpatient clinic at 1 month and 3 months post-operatively. On follow up visits, there were no further complaints of nasal blockage noted with good bilateral air blast.

Case 2- 15 year old female, sister of case -1, came with similar complaints for 1year (Figure 4). Similar to her sibling, nasal polyps were refractory to multiple courses of antibiotic and corticosteroid therapy. She also underwent FESS surgery 2 years back with recurrence of pre-operative symptoms within few months. The clinical presentation, endoscopic and radiological findings were comparable to elder sibling. This patient also showed similar picture on HRCT thorax. At our centre she was also treated with a course of antibiotics and steroid nasal spray for 2 months; but no significant benefits were noted. Revision FESS was performed and polypoid tissues were sent for the histopathological examination. Intra-operatively, similar to case 1bilateralhypo plastic frontal sinus was noted. As nasal deformity was more pronounced in this case, septorhinoplasty by endonasal approach was performed in the same sitting(Figure 5).Histopathological report obtained in this case was also comparable to case-1.This patient also received same post-operative care .During 3rd month follow up, there was no recurrence of nasal polyp noted with symptomatic relief. Nasal contour cosmetically improved with better definition of nasolabial fold. (Figure 6).

Case 3-13 year old female sibling came with similar complaints for 6 months.Compared to her siblings she only had minimal nasal deformity (Figure 7)... Clinical examination, anterior rhinoscopy, diagnostic nasal endoscopy were comparable to previous cases. CT PNS showed nasal polyposis occupying bilateral nasal cavity with pan-sinusitis. HRCT Thorax showed bronchiolitis with few fibro nodular areas with sub segmental consolidation .Multifocal bronchial stenosis with bronchiectasis in both lungs with no regional predominance.

	CASE 1	CASE 2	CASE 3
AGE	17	15	13
GENDER	MALE	FEMALE	FEMALE
H/O NASAL OBSTRUCTION	5 YEARS	3 YEARS	6 MONTHS
H/O PREVIOUS SURGERY	+	+	-
OTHER COMPLAINTS	Progressive deformity of nose, bilateral mild decreased hearing, chronic cough	Progressive deformity of nose, bilateral mild decreased hearing, chronic cough	Chronic cough Left ear discharge
ANTERIOR RHINOSCOPY	Bilateral nasal polyposis	Bilateral nasal polyposis	Bilateral nasal polyposis
DIAGNOSTIC NASAL ENDOSCOPY	Bilateral nasal polyposis with thick viscid secretion	Bilateral nasal polyposis with thick viscid secretion	Bilateral nasal polyposis with thick viscid secretion
SERUM IgE	177 IU/mL	238 IU/mL	303IU/Ml
SWEAT CHLORIDE TEST	NEGATIVE	NEGATIVE	NEGATIVE
CT PNS	Nasal polyposis occupying bilateral nasal cavity with pansinusitis.	Nasal polyposis occupying bilateral nasal cavity with pansinusitis.	Nasal polyposis occupying bilateral nasal cavity with pansinusitis.
HRCT THORAX	Bilateral symmetrical central bronchiectasis with upper lobe predominance. No evidence of situs inversus.	Bilateral symmetrical central bronchiectasis with upper lobe predominance. No evidence of situs inversus.	Bronchiolitis with few fibro nodular areas with sub segmental consolidation with Multifocal bronchial stenosis with bronchiectasis in both lungs with no regional predominance. No evidence of situs inversus.
MEDICAL MANAGEMENT	Antibiotic and steroid nasal spray for 2 months	Antibiotic and steroid nasal spray for 2 months	Antibiotic and steroid nasal spray for 2 months
SURGICAL MANAGEMENT	Revision bilateral full house FESS	Revision bilateral full house FESS with septorhinoplasty.	Bilateral full house FESS
HISTOPATHOLOGY REPORT OF POLYPOID TISSUE	Inflammatory benign nasal polyp with cellular infiltrates predominantly composed of lymphocytes, plasma cells, neutrophils with few eosinophils.	Inflammatory benign nasal polyp with cellular infiltrate predominantly of lymphocytes, plasma cells with few eosinophils	Inflammatory benign nasal polyp with lymphoplasmacytic infiltrate, with scant neutrophils and eosinophils

In contrast to the other two cases, this was a primary case with no history of previous surgeries. After a failed medical management, bilateral FESS was performed. Post-operative histopathological report and post-operative follow up were in line with the previous 2cases.

DISCUSSION

Woakes syndrome, as described by Woakes in 1885 hadfour characteristics: childhood onset bilateral nasal polyposis in the middle meatus, ethmoiditis, nasal pyramid deformity and therapeutic failure, with constant and rapid recurrences (5).In the current case series, the clinical presentation of all three siblings met the diagnostic criteria of Woakes syndrome. In 1979, Kellerhals and De Uthemann reported4 cases ofrecurrent infantile nasal polyposis with broadening of the nose, frontal sinus aplasia, bronchiectasis and nasal dyscrinia, describing it as Woakes' syndrome (4). According to this definition, our patients met all the diagnostic criteria except frontal sinus aplasia similar to Abbud-Neme et al (6).As frontal sinus development usually occurs by 7 years of age and the cases described by Kellerhals and De Uthemann belonged to early childhood, these patients presented with frontal sinus aplasia. In contrary, our patients were teenagers and hence presented with bilateral hypoplastic frontal sinus. Most cases of Woakes' syndrome occur in children and young adults due to the plasticity of the developing facial bony structures (7, 8, and 9). Similar cases of childhood nasal polyposis are also seen in allergy, cystic fibrosis and primary ciliary dyskinesia. The diagnosis of Woakes syndrome was confirmed only after excluding above mentioned conditions. Serum IgE levels within normal range in all three siblings excluded allergic etiology for nasal polyposis. A diagnosis of cystic fibrosis was ruled out in view of negative sweat chloride test and absence of abdominal symptoms (10). As the radiological imaging lacked any evidence of situs inversus, primary ciliary dyskinesia was unlikely (5).

In literature, few studies described synchronous occurrence of Woakes syndrome in two siblings (5, 11), making heredity a potential contributing factor (4, 5). But precise etiopathogenesis for the same still remains unknown.Similar to other studies in the literature, there was no evidence of these syndromic presentations in any previous ancestors of these siblings. Isolated involvement of three siblings of the same family makes non-inherited unknown genetic factor a possible etiology for this syndrome. Functional Endoscopic Sinus Surgery (FESS) remains gold standard in managing nasal polyp. It also restores normal sinus ventilation and drainage, relieving obstruction of osteomeatal complex. Though there is a general consensus over a functional endoscopic sinus surgery for Woakes syndrome, addressing the nasal deformity is still disputable. Laff performed nasal plastic repair with osteotomy (12).Foze etal. described septorhinoplasty to restore nasal function and to improve cosmetic appearance (13). Schoenenberger U et al. described simple facial digital compression without osteotomies toimprove cosmetic appearance in a case of adult onset Woakes syndrome in view of atrophy of nasal bone (14).Ueda et al did Rhinoplasty via midface degloving approach in a severe case of Woakes syndrome with severe deformity including excessive prominence of the malar region (15).In this case series, only one out of three patients had marked nasal deformity warranting FESS with Septorhinoplasty in same sitting. After taking into consideration the recurrent nature of nasal polyposis in this syndrome, the authors came to a consensus that a regular and thorough post-operative follow-up for a prolonged period should be given an equal thrust as surgery alone in the management of Woakes syndrome.

Conclusion

To conclude, Woakes syndrome is a clinical condition characterised by a tetrad of recurrent nasal polyposis, nasal pyramid deformity, frontal aplasia or hypoplasia and

bronchiectasis. Etiopathogenesis of this syndrome still remains unclear due to its rarity and sparse documentation. Non-inherited unknown genetic factor may be a possible etiology, and needs further detailed genetic studies. As these cases are usually refractory to medical management, meticulous surgery in the form of Functional endoscopic sinus surgery forms the mainstay of the treatment with regular post-operative care for prolonged duration carrying equal importance. Rhinoplasty surgery helps the patient to give cosmetic results and better face. Furthermore, detailed study over a prolonged period is required in such patients to comment upon specific duration of follow-up required.

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