Woakes’ Syndrome; Rare Manifestation of Nasal Polyposis: Experience of Seven Patients

ABSTRACT

Background: Woakes’ syndrome, first described by Woakes in 1885 which is also better known as ethmoiditis, is a very uncommon entity of severe nasal polyposis causing disfigured facial appearance. In this study, we aimed to present the diagnosis and management of seven patients’ series with Woakes’ syndrome in the light of current literature.

Methods: The medical records of seven patients who had functional endoscopic sinus surgery because of nasal polyposis that caused nasal roof deformity between January 2013 and June 2020 in a tertiary hospital otorhinolaryngology department were evaluated retrospectively. Demographic aspects, examination data, rhinological surgery history, Lund-Mackay scores at admission, surgical procedures performed, and follow-up periods were analyzed.

Results: All patients had bilateral severe nasal polyposis and significant nasal bone expansion. Bilateral functional endoscopic sinus surgery was applied to all patients and additionally septoplasty was applied to one patient and septorhinoplasty to two patients in the same session. Five patients stated they did not want septorhinoplasty in the same session. We tried reduction of nasal bones with a finger pressure in these patients. While we achieved a satisfactory reduction in three of these patients, it was unsuccessful in two cases.

Conclusion: Treatment of nasal polyps should be achieved by functional endoscopic sinus surgery for Woakes’ syndrome. Topical and systemic treatment of the nose should slow down or evade the recurrence of the nasal polyps. Approach to nasal deformity should be addressed during the initial surgery if feasible.

Keywords: Woakes’ syndrome, nasal polyposis, disfiguration, functional endoscopic sinus surgery

INTRODUCTION

Nasal polyposis (NP) is a recurrent inflammatory condition that develops when the mucosa lining of the nasal cavity hangs toward the middle meatus. It arises principally from the ethmoid sinus mucosa and appears as smooth surface, pale, edematous masses. Over the last years, Woakes’ syndrome has mostly been specified as severe nasal polyps, with the consecutive destruction of the nasal pyramid leading to the disfiguration of the nose due to constant pressure of the polyroid masses.3 The majority of Woakes’ syndrome cases appear in childhood and youth due to the plasticity of the osseous structures of the developing face. The etiology is still unknown. There are just a few cases presented in the literature.4,5 As far as we have reviewed the literature, this study has the largest number of Woakes’ syndrome patients. The purpose of this study was to present the diagnosis and management of seven patients with Woakes’ syndrome in the light of current literature.

METHODS

The study was approved by the Institutional Review Board and the Ethics Board of the Afyonkarahisar Health Sciences University—number of approval IRB 204/27–4–20. Signed consent of patients was obtained for permission to publish photographs, radiologic images, and medical data.

The medical records of seven patients who had functional endoscopic sinus surgery (FESS) because of NP that caused nasal roof deformity between January
2013 and June 2020 at Afyonkarahisar Health Sciences University Otorhinolaryngology department were evaluated retrospectively. Patients with nasal deformation because of trauma were not included in the study. The files of the cases were assessed retrospectively. Demographic aspects, examination data, rhinological surgery history, Lund-Mackay scores (Table 1) at admission, surgical procedures performed, and follow-up periods were noted. All patients were given 1 mg/kg methylprednisolone per oral before the surgery, with a dose reduction of 15 mg every 3 days. Preoperative paranasal sinus tomography was requested from all patients. All patients underwent bilateral FESS under general anesthesia for NP, and some patients underwent septorhinoplasty/septoplasty procedures in the same session, depending on the patient’s request. We attempted nasal bone reduction with a finger pressure in all patients during the surgery. Messerklinger and Wigand technique was used as the surgical technique for FESS. Preoperatively, topical anesthesia and vasoconstriction were provided by using cotton pads impregnated with 1/1000 adrenaline and 2% panto
caine. After topical anesthesia, infiltration anesthesia with lidocaine and 1/1000 epinephrine were performed for sphenopalatine ganglion blockade. We did not apply nasal packing to the patients who did not have active bleeding at the end of the surgery. In the surgeries, 4 and 2.7 mm 0\°, 30\°, and 45\° nasal telescope, xenon light source, and monitor were used. We removed the nasal packings on the third postoperative day. We removed the packings and external nasal splints of the patients who underwent septorhinoplasty on the seventh day. Nasal washing with isotonic saline was recommended for nasal secretion and clearing of crusts. For endonasal examination, the patients were called for control on the tenth postoperative day, and nasal topical steroid was started.

### RESULTS

Of the 7 patients included in the study, 5 were male and 2 were female. The ages of the cases were between 19 and 51 (mean 37.8). The preoperative Lund-Mackay scores ranged from 17 to 24 (mean 20.7). All patients had bilateral diffuse NP and significant nasal bone expansion (Figures 1 and 2). Bilateral FESS was applied to all patients and additionally septoplasty was applied to 1 patient and septorhinoplasty to 2 patients in the same session. Five patients stated they did not want septorhinoplasty in the same session. We tried reduction of nasal bones with a finger pressure in these patients. While we achieved a satisfactory reduction in 3 of these patients, it was unsuccessful in 2 cases. We observed no perioperative complications in any of the patients. We applied anterior packing in patients who developed postoperative mucosal bleeding. Small mucosal adhesions were opened with local anesthesia during patient follow-up. The mean follow-up period was 21.1 months (12–32 months) (Table 2). We observed no recurrence of NP in any of the patients during the follow-ups.

### MAIN POINTS

- Woakes syndrome is a rare disorder commonly defined as recurring nasal polyposis with consequent destruction of the nasal pyramid.
- This study has the largest number of Woakes’ syndrome patients, while there are few case reports presented in the literature.
- The removal of nasal polyps should be the first aim of treatment. However, external nose deformation correction by finger pressure during sinus surgery can be tried for all patients, if not possible, rhinoplasty should be suggested during or after surgery up to patient expectation.
DISCUSSION

The nasal polyp is derived from the phrase “polyposus,” poly foot (poly: poly, pous: foot) in ancient Greek. Hippocrates reported nasal polyps to result from an imbalance between fire, water, soil, and air. Nasal polyps, considered a neoplasm in the early 1800s, were reported by Paget in 1854 as mostly benign tumors. In 1885, Woakes has suggested nasal polyps are not tumors, but inflammatory lesions. In 1872, Voltani reported nasal polyps were seen with asthma. In 1922, Widal et al. reported that asthma, aspirin intolerance, and nasal polyps were common together.6

Nasal polyps are an inflammatory condition in the nasal region and are generally encountered in otorhinolaryngology clinics. Nasal polyps are usually defined as nasal mucosal membrane enlargement, occluding the bilateral nasal airways and usually accompanied by chronic sinusitis, nasal or postnasal discharge. They are the most frequent mass lesions of the nose, and they recur despite medical and surgical treatments. Although the physiopathology of nasal polyps is not fully understood, epithelium may play a critical role in inflammatory nasal diseases. Numerous pathogenic theories have been proposed for the etiology. These theories include adenoma and fibroma theories, glandular cyst theories, mucosal exudate theories, blockage theories, glandular hyperplasia theories, other gland formation theories, ion transport theories, pylephlebitis theory, secretory duct cystic dilatation, and vascular occlusion theory.5–7

Woakes’ syndrome is better known as ethmoiditis; it is an unusual condition causing disfigured nose by severe NP growth in the nasal cavity and paranasal sinuses.1 It was further detailed by Société Française de Laryngology with these features: deformed nasal pyramid because of hypertrophic process, ethmoiditis, a childhood disease of bilateral nasal polyps in the middle meatus, and failure in treatment with recurrences.8 In our study, all patients fulfilled the criteria of Société Française de Laryngology except being child and recurrence of polyps.

FESS is a gold standard for managing nasal polyps. The treatment of NP involves topical and oral corticosteroids. FESS is used to remove the polyps of the sinuses and nasal cavity and to re-establish normal nasal ventilation. Foze et al described rhinoplasty as part of the treatment in Woakes’ syndrome, but, apart from this description, the treatment appears to be mostly limited to FESS.6 However, a simple facial finger pressure without osteotomies can still be performed to correct cosmetic appearance if nasal bones allow.5–12 Schoenenberger and Tasman showed that external nose deformation can be well corrected by finger pressure with FESS.13

The surgery varies depending on polyposis and nasal deformity, the expertise of the surgeon, the material and technology accessible, the condition, and expectations of the patient.

Briefly, Woakes’ syndrome is rarely reported in the literature and its exact etiology remains unclear. As far as we have reviewed the literature, this study has the largest number of Woakes’ syndrome patients. There is a consensus regarding the treatment of nasal polyps by FESS. Topical and systemic treatment of the nose should slow down or evade the recurrence of the nasal polyps. Approach to nasal deformity should be addressed during the initial surgery, if feasible.

REFERENCES


Table 2. Dermographic and Clinical Findings of Seven Patients

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FESS, Functional Endoscopic Sinus Surgery; SRP, Septorhinoplasty.