Case Report

Adult-onset woakes’ syndrome: Report of two cases

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\textbf{A R T I C L E   I N F O}

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\textbf{A B S T R A C T}

Introduction: Woakes’ syndrome is a rare condition commonly defined as recurrent sinonasal polypsis with consecutive destruction of the nasal pyramid. Till now, only a few cases have been reported in the literature. The purpose of this paper is to present the features of woakes’ syndrome through two new clinical cases, adding some valuable insight to the recently reported cases.

Case report: We report a series of two consecutive adults male and female patients, aged 55 and 58 years, with Samter’s triad, who presented recurrent nasal polypsis and progressive widening of the nasal dorsum. Facial CT showed in both patients the same radiologic pattern of nasal and paranasal cavities obliteration with nasal bone deformation. Both patients underwent functional endoscopic sinus surgery and correction of the bony nasal vault deformity without osteotomies. At 3 months follow-up, the nasal air passage remained free and aesthetic outcomes were observed.

Discussion: having been described over 130 years ago, the etiology of woakes’ syndrome remains unclear. Treatment includes topical treatment and sinonasal surgery. Surgical treatment of the nasal dorsum deformity is rarely addressed.

Conclusion: These observations suggest that the external nose deformity may be successfully corrected by digital compression, in combination with endoscopic sinus surgery.

\textbf{1. Introduction}

Woakes’ syndrome is a very rare entity described for the first time in 1885 by Woakes [1]. It was defined as severe recurrent nasal polypsis with consecutive deformation of the nasal pyramid [1]. Till now, only a few cases were reported in the literature, most of them occur in children and young adults with the precise etiology being poorly understood [2]. Surgical treatment of the external nose deformity in these cases has rarely been addressed. The purpose of this paper is to present the features of woakes’ syndrome through two new clinical cases, adding some valuable insight to the recently reported cases. This case report is in line with the SCARE criteria [3].

\textbf{2. Patient and observation}

\textbf{Case 1.} A 55-year-old male patient consulted our department, in 2012, with severe and progressive nasal obstruction. Symptoms had begun 4 years earlier but had been left untreated. He was still known to have an aspirin intolerance. additionally, he had suffered from asthma since his youth and does not mention any positive family history for nasal polypsis. Physical findings documented at the time an obstruction of the nasal cavities by polyps, anosmia and a normal appearing external nose.

After 06 years of medical treatment with topical steroids failed, an endoscopic surgery was proposed to the patient, but he refused. Afterward, he was lost to follow-up and started self-medication with topical and oral corticosteroids. In 2020, the patient returned to our department for an aggravation and persistence of symptoms, including complete bilateral nasal obstruction by polyps visible in both nares and worsened anosmia. Additionally, there was a progressive broadening of the nasal pyramid. Physical examination showed a distinct widening and bilateral enlargement of the nasal pyramid (Fig. 1). Anterior rhinoscopy revealed nasal polyps obstructing the vestibule of the nose on both sides. On a CT scan, a total obliteration of the nasal cavities and paranasal sinuses as well as deformation and massive expansion of the nasal bones (Fig. 2).

The patient’s test results with Sniffin’ sticks showed an anosmia (identification of 2 out of 12 possible odors). Endoscopic endonasal
polypectomy, bilateral maxillary sinus fenestration and functional ethmoidectomy were performed. Simultaneously, the bony nasal vault was narrowed by forced external digital compression without osteotomies. No nasal packing was inserted. A nasal splint was applied for 2 weeks. Histology of polyps showed a squamous epithelium and respiratory mucosa with polymorphic inflammatory infiltrate without signs of malignancy. Nasal saline irrigation and topical corticosteroids were prescribed postoperatively. At the 3 months' follow-up, the nasal air passage remained free, the sense of smell improved and the form of the exterior nose remained stable.

Case 2. A 58-year-old women presented to our ENT department, in 2019, with the same complaints as the previous patient (ASA triad with complete bilateral nasal obstruction and progressive broadening of the nasal pyramid). Symptoms had begun 8 years earlier but had been left untreated. She had suffered from asthma since her youth and reported a positive family history for nasal polyposis. Physical examination showed a distinct widening and bilateral enlargement of the nasal pyramid with nasal polyps obstructing the vestibule of the nose on both sides (Fig. 3). A CT scan showed the same pattern of opacification seen in the previous patient with deformation and massive expansion of the nasal bones (Fig. 4). Endoscopic polypectomy, bilateral maxillary sinus fenestration and functional ethmoidectomy were performed. After that, nasal deformity was corrected by an external digital compression without osteotomies. A nasal splint was applied for 3 weeks. Histology of polyps showed the same result without signs of malignancy. Nasal saline irrigation and topical corticosteroids were prescribed postoperatively. At the 6th months' follow-up, the nasal air passage remained free, the sense of smell improved and the form of the exterior nose remained stable.

3. Discussion

Woakes’ syndrome was described for the first time, in 1885, as a form of necrotising ethmoiditis and mucosal polyps of the nasal cavities, sometimes producing a broadening of the bridge of the nose, when Edward Woakes’ presented a paper entitled the relation of necrosing ethmoiditis to nasal polypus to the Medical Society of London [1]. In recent years, Woakes’ syndrome is described as severe recurrent sino-nasal polyps with consecutive destruction and widening of the nasal pyramid due to chronic pressure of the polyps [2]. Most cases of Woakes’ syndrome seem to occur in children and young adults because of the plasticity of the bony nasal vault [4]. The etiology of this syndrome remains unknown; however, its heredity appears to be a potential contributing factor [5].

Our patients presented several risk factors for nasal polyps with an aspirin intolerance, multiple allergies and a positive family history for the second patient. Even so, it remains unclear why a few patients with nasal polyps develop a deformation of the nasal pyramid while most show only a deformity limited to the paranasal sinuses [6]. Based on our observations, the development of the condition over a period of more than 08 years during adulthood without treatment for the first patient, and self-medication with oral corticoid for the second patient seemed to be the origin of that deformation.

Our patients fulfilled the criteria for Woakes’ syndrome as defined by Appaix and Robert [7], as well as the criteria for Fernand Widal’s syndrome [8].

The treatment of sinonasal polyposis is based on topical and oral corticosteroid. Endoscopic sinus surgery is used to evacuate polyps from the osteomeatal complex and to restore normal nasal ventilation [9]. For Foze et al. Woakes’ syndrome treatment appears to be limited to endoscopic sinus surgery, rhinoplasty may be associated with the treatment for aesthetic purposes [10]. Schoenenberger and Tasman had shown that the external nose deformity may be successfully corrected by digital compression during or after endoscopic sinus surgery [1]. Our patients presented several risk factors for nasal polyps with an aspirin intolerance, multiple allergies and a positive family history for the second patient. Even so, it remains unclear why a few patients with nasal polyps develop a deformation of the nasal pyramid while most show only a deformity limited to the paranasal sinuses [6]. Based on our observations, the development of the condition over a period of more than 08 years during adulthood without treatment for the first patient, and self-medication with oral corticoid for the second patient seemed to be the origin of that deformation.

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cases illustrate the feasibility of a forced external digital compression without osteotomies to correct the external nasal deformity.

4. Conclusion

Woakes’ syndrome is a rare clinical entity that still well described in the literature even if its exact etiology remains unclear. An adequate functional treatment of the nasal polyps by endoscopic sinus surgery, local and, if necessary, general corticosteroid therapy could be factors to slow down or avoid recurrence of the nasal polyps. The treatment of the external nose deformity should be addressed during the primary surgery if possible.

Declaration of competing interest

All authors disclose any conflicts of interest.

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Ethical approval

The study committee of the university hospital center approves the favorable opinion to publish this work.

Consent

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

Author’s contribution

Dr. IC, Dr. RB, Dr. AL, Dr. DB, Dr. AA, Dr. FE have analysed and performed the literature research, Pr. RG performed the examination and performed the scientific validation of the manuscript. Dr. Ilham CHENNOUFI was the major contributor to the writing of the manuscript. All authors read and approved the manuscript.

Registration of research studies

Not Applicable.

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Availability of data and material

The datasets in this article are available in the repository of the ENT database, Chu Mohamed VI Oujda, upon request, from the corresponding author.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.amsu.2021.102695.

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