Laporan Kasus

Woakes syndrome

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ABSTRACT

Background: Extensive nasal polyp growth in the paranasal sinuses can lead to bone erosion of the sinus walls and cause facial disfigurement due to continuous pressure or chronic inflammation. This extremely rare phenomenon is called Woakes syndrome. This syndrome consist of several symptoms include the destruction of ethmoid sinus that cause broadening of the bridge of the nose, frontal sinus aplasia and bronchiectasis. **Purpose**: To give complete information about the diagnosis and management of Woakes Syndrome. **Case**: A 16-year-old boy with deformity of the left nose, nasal obstruction and frequent episodes of rhinorrhea since 4 months before admission. Nasoendoscopic evaluation showed huge nasal polyps filling the left nasal cavity, pushing the septum and narrowing the right nasal cavity. Histopathology result was edematous polyp with necrosis and massive bleeding without signs of malignancy. **Management**: Patient was managed in two stages operations. First, nasal polyp removal by FESS technique in general anesthesia, and the second stage four months later, was septorhinoplasty for aesthetic bridge reconstruction. **Conclusion**: Nasal polyps could be related to Woakes syndrome, characterized by broadening of nasal bridge which needs functional and aesthetic surgery.

Keyword: Woakes Syndrome, nasal polyps, Functional Endoscopic Sinus Surgery,Septorhinoplasty

ABSTRAK

Latar Belakang: *Polip hidung besar yang meluas dalam sinus paranasal dapat menyebabkan erosi dinding sinus dan menyebabkan cacat wajah akibat tekanan terus-menerus atau peradangan kronis. Fenomena ini sangat langka dan disebut sebagai sindrom Woakes. Sindrom ini terdiri dari beberapa gejala termasuk kerusakan dinding sinus etmoid yang menyebabkan hidung melebar, aplasia sinus frontal dan bronkiektasis.* ***Tujuan****: Untuk memberikan informasi yang lengkap tentang diagnosis dan penatalaksanaan Woakes Syndrome.* ***Kasus****: Seorang anak laki-laki 16 tahun dengan deformitas hidung kiri, hidung tersumbat dan pilek berulang sejak 4 bulan. Evaluasi nasoendokopi menunjukkan polip hidung masif mengisi rongga hidung kiri, mendorong septum dan menyempitkan rongga hidung kanan. Pemeriksan histo-patologi memperlihatkan polip edematosa dengan nekrosis dan perdarahan masif tanpa tanda-tanda keganasan.* ***Penatalaksanaan****: Pada pasien dilakukan dua tahap tindakan. Pertama, dilakukan Bedah Sinus Endoskopik Fungsional (BSEF) dan polipektomi dalam anestesi umum, dan empat bulan kemudian pasien menjalani septorinoplasti untuk rekonstruksi wajah.* ***Kesimpulan****: Polip hidung pada kasus ini kemungkinan terkait dengan sindrom Woakes, ditandai dengan pelebaran pyramid hidung yang membutuhkan tindakan operasi fungsional dan estetika.*

Kata kunci*: sindroma Woakes, polip hidung, Bedah Sinus Endoskopik Fungsional, Septorinoplasti.*

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INTRODUCTION

Woakes syndrome or ethmoiditis was first described by Woakes in 1885. The clinical hallmark of this entity with severe recurrent growth of nasal polyposis is broadening of the nose and hypertelorism. Polyps in Woakes' syndrome do not differ histologically from nasal polyps but are more often caused by a non-eosinophilic dominated inflammation.[1]

Woakes syndrome is a very rare case. No more than 10 cases had been reported in the published journals. Abud-Neme et al in 1986 reported one case of 17-year-old girl with Woakes syndrome who underwent resection of polyps and fibrous tissue and reconstruction of the nasal pyramid. Kellerhals et al in 1977 reported a 5-year-old-girl with Woakes syndrome managed with polyp resection and planned for reconstruction after puberty.[2, 3]

The occurrence of nasal polyps is a multifactorial disease resulting from chronic immune inflammatory processes within the paranasal sinuses. The pathophysiological processes of the underlying inflammation causing chronic hyperplastic sinusitis and nasal polyposis are still unclear, and the patho-etiology of this syndrome is also still obscure. Some authors have suggested genetic factors, as the syndrome has occurred more frequently among sisters, as mentioned by Groman et al,[1,4] and others implied infectious origin, primarily syphilitic. External noxious agents and allergies, could accelerates the growth of polyps. However, in many cases, no agents or allergy could be found. This indicates that the syndrome with its deforming and recurrent polyps is only a clinical entity. The extreme broadening of the nose is explained as the result of the chronic pressure of the polyps. The chronic pressure also leads to a bolding of the lamina papyracea with progressive protrusiobulbi to the lateral side and to a risk of slowly progressing blindness due to compression of the optic nerve.[1]

The etiology of nasal polyps is unknown. Some theories consider polyps as consequence of conditions which cause chronic inflammation in the nose and nasal sinuses characterized by stromal edema and variable cellular infiltrate. While many aspects have been documented to support this theory, the initiating cause remains unknown and might be different in many cases.[1-4] Medical conditions commonly associated with polyps include asthma, bronchiectasis, and cystic fibrosis There is a well recognized subgroup of patients with Samnter's Triad comprising polyposis, asthma, and aspirin hypersensitivity which makes up almost 10% of cases of nasal polyps.[5-8]

Extensive polyp growth in the paranasal sinuses and facial disfigurement due to continuous pressure or chronic inflammation and ethmoiditis.The clinical hallmark of this entity with severe recurrent growth of nasal polyposis is broadening of the nose and hypertelorism. The syndrome has also been described with bronchiectasis in children.[1] Caversaccio et al,[1] reported following four characteristics (1) bilateral nasal polyps in the middle meatus beginning during childhood, (2) ethmoiditis, (3) hypertrophic process with nasal pyramid deformation and (4) therapeutic failure with constant and rapid recurrences.

Patient with Woakes syndrome can complaint about abundant nasal discharge and the secretions being of a rubber-like viscosity. Further examination revealed frontal sinus aplasia, bronchiectasis with clubbed fingers, deformation of the nasal bridge and no sign of allergies.[3,4]

Laboratory findings showed no signs of allergic disease. AlI blood examination seems to be normal. CT scan showed a complete obliteration of the paranasal sinuses on both sides with a bulging of the ethmoid walls into the orbits.[1]

Management of Woakes syndrome consists of two stages. The first is by complete

resection of the nasal polyps, best executed by Functional Endoscopic Sinus Surgery (FESS) followed by the second stage of reconstruction. Septorhinoplasty on Woakes syndrome has a goal in restoring nasal function by maximizing nasal airflow and improving cosmetic appearance.[9]

The aim of publishing this case is to remind us about the nature of chronic polypoid inflammatory disease that may cause facial disfigurement as distinctive as broadening of nasal pyramid and its management.

CASE REPORT

A 16 year-old-boy was referred from a district hospital with a diagnosis of sinonasal tumor. Patient complained of recurrent common colds with nasal blockage since 3 years before admission. Six months earlier, patient could not breathe through the nose. The last four months patient felt impaired sense of smell and found that his left nose became enlarged. He had moderate headache especially when the nasal blockage become severe. There was no history of facial pain, allergy or trauma. Patient did not complain of nosebleeds, hearing loss, double vision, or neck lymph node enlargement.

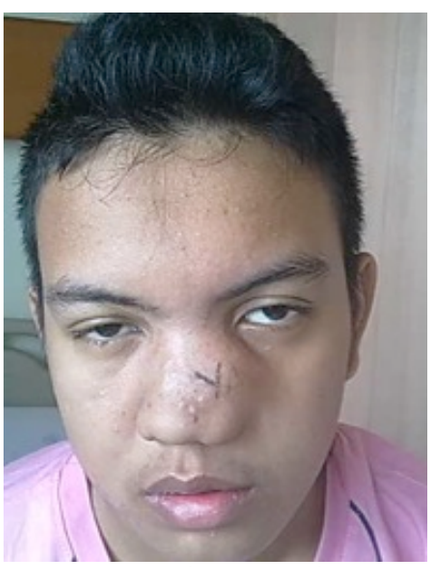

Fig. 1 External nose enlargement

Physical examination revealed polypoid mass filling the entire nasal cavity with enlargement of the nose. Frontal view appearance showed nose enlargement with no distinctive border of the left nose. From the basal view appearance, the tip of the nose is deviated to the right side. Sinus paranasal CT Scan taken on December 19th, 2012 showed opacities on the right and left nasal cavities, both choana and nasopharyngeal space, bilateral maxillary sinuses, bilateral ethmoid sinuses, left sphenoid sinus and left frontal sinus. There were signs of erosion of the nasal septum, bilateral medial maxillary sinus walls, left posterior maxillary sinus wall, left orbital inferomedial wall, and left inferior sphenoid sinus wall. There was thickening of right sphenoid sinus mucosa.

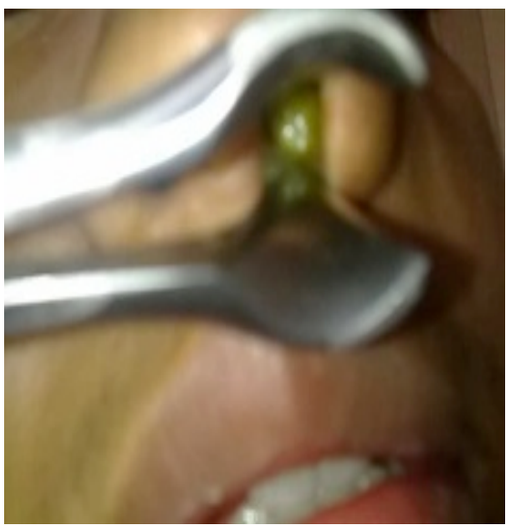

Fig. 2 Anterior rhinoscopy

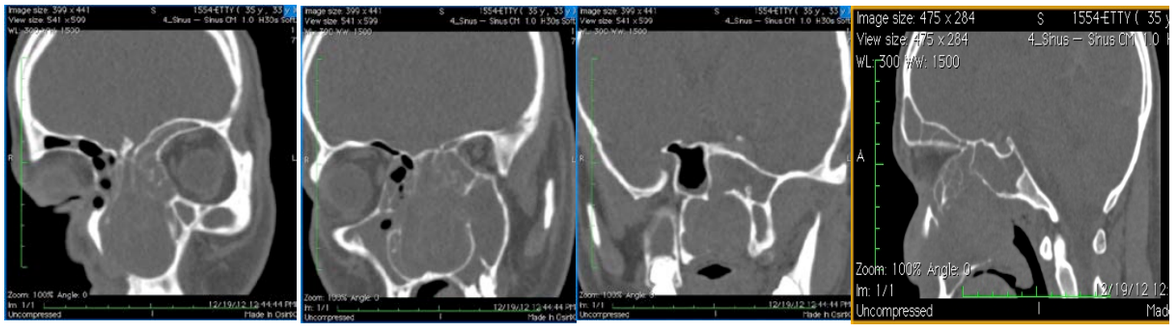

Fig. 3 Paranasal CT Scan of the patient with WoakesSyndrome

Patient underwent biopsy of the nasal mass in January 15th, 2013 with the result of chronic rhinitis and nasopharyngitis. FESS was performed in March 8th, 2013. Polypoid mass was extracted with forceps. Inferior and medial turbinate was difficult to recognize due to the expansion of the polypoid mass. The bleeding from the sphenopalatine artery was cauterized. Uncinectomy was performed with backbiting forceps, followed by middle meatal antrostomy finding no polyp inside the maxillary antrum.

Histo-pathology examination showed edematous polyps with necrosis and massive bleeding and no signs of malignancy. Patient was discharged on the fifth day after hospitalization and was prescribed coamoxiclav 3x625 mg, mefenamic acid 3x500 mg and normosaline solution for nasal rinsing. First post operative follow up was done on March 14th 2013 and nasoendoscopy examination revealed wide left nasal cavity, there was clotting and no mass. Right nasal cavity still narrow due to septal deviation. Patient did not complain of nasal blockage anymore and could breathe through the nose. Four months after surgery, patient prepared for second stage surgery by Plastic Reconstruction ENT Division for rhinoplasty correction.

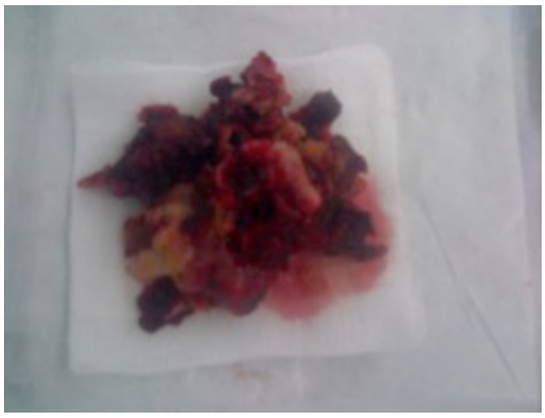

Fig.4 Polypoid tissue of surgical specimen.

Septorhinoplasty was performed on July 8th 2013. Graft augmentation was taken from left auricle cartilage. Transcollumelar incision was carried out, followed by infracartilage incision. Septal cartilage was incised sparing superior and posterior part. Septal extension graft was done with interdomal suture. Osteotomy was performed at right and left side. Nasal splint was maintained for 2 weeks and removed on July 25th, 2013. Patient was satisfied with the result.

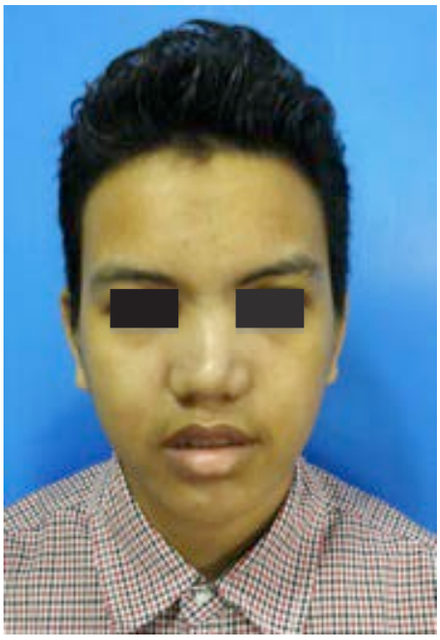

Fig.5 patient 16 years old boy with Woakes syndrome after septorinoplasty

DISCUSSION

According to Slavin et al as quoted by Casale,[6] nasal polyps are present in approximately 1% to 4% of the population, more often in males than females. This is similar with the case reported in this paper.

Case reports in published journals showed age incidence of Woakes syndrome under 10 years old.[3] Our case was a male teenager.

Patient in this case presented with nasal blockage and frequent episodes of rhinorrea since 3 years previously. He also complained loss of smell and moderate headache. According to Tritt et al,[10] patients with nasal polyposis may present clinically with complaints of nasal obstruction, congestion, hyposmia, rhinorrhea, epistaxis, postnasal drip, headaches, and snoring. Patient also complained of enlargement of his left nose. Broadening of the nose is a characteristic of Woakes syndrome as described by Caversaccio et al.[1]

Physical examination revealed clear polypoid mass filling the left nasal cavity. Nasal polyps typically present bilaterally but can present unilaterally. Unilateral nasal masses may be benign unilateral nasal polyps in relation with chronic rhinosinusitis. Biopsy from nasal and nasopharyngeal region showed chronic rhinitis and nasopharyngitis, which conclude that the patient has unilateral nasal polyp.[7]

The extreme broadening of the nose in our patient is explained by the ongoing pressure of the polyps. This is in accordance with hypertrophic process and nasal pyramid deformation, one of the four characteristics of Woakes syndrome.[1]

Paranasal sinus CT scan is needed to prove the frontal sinus aplasia. These findings added to the biopsy result suggest a Woakes Syndrome. Woakes' syndrome was defined as necrotizing ethmoiditis and nasal polyps with broadening of the nose. The syndrome was defined by the following four characteristics: (1) bilateral nasal polyps in the middle meatus beginning during childhood, (2) ethmoiditis, (3) hypertrophic process with nasal pyramid deformation and (4) therapeutic failure with constant and rapid recurrences.[1] In our case, frontal sinus aplasia was not found, but there was full opacification of left frontal sinus without any polps inside it. The left frontal sinusitis is caused by obstruction of the frontal recess.

We extirpate the bulk of nasal mass by nasoendoscopic procedure and sent it for histo-pathological examination. Functional Endoscopic Sinus Surgery (FESS) is considered the gold standard for the surgical treatment of chronic rhinosinusitis with or without nasal polyps. It is performed with a rigid endoscope which allowed a complete visualization of the nasal and paranasal cavities. The aims of this surgical technique is to restore normal sinus ventilation and

drainage removing polyps or other tissue obstructing the osteomeatal complex. FESS could minimized the recurrence rate of the disease. The extent of surgery varies with the extent of disease, the surgeon's expertise, the available equipments and technology, and patient's condition and expectation.[11-13]

Four months afterwards septorhinoplasty was performed. The goal of septorhinoplasty and rhinoplasty are to restore nasal function by maximizing nasal airflow and to improve cosmetic appearance. There are two completely different methods of rhinoplasty for correcting nasal pyramid deformations; the first is in which the osteo-cartilaginous resection of the nasal dorsum is carried out and the second is based on preservation of the dorsum.[9,14] The two techniques were never combined because osteo-cartilaginous resection of the nasal hump prevents complete disarticulation of the septal cartilage from its attachment on the perpendicular plate of the ethmoid bone and the vomer, an essential step in Cottle septoplasty. Indeed, once the septal cartilage is completely detached from its posterior and inferior edges, as performed in Cottle septoplasty, it can no longer be separated from the upper lateral cartilages as performed in nasal hump resection. This would involve total detachment with major risk of post-operative saddling and, moreover, stenosis of the internal nasal valve due to the collapse of the upper lateral cartilages.[2,14]

In the literatures reviewed, there was only one case of Woakes syndrome which underwent septorhinoplasty.[2]

In this paper, a case of Woakes syndrome was reported. The patient underwent two stages surgery, the first was FESS to clear out the polyp mass and the second surgery was septorhinoplasty to restore physiologic function and aesthetic appearance of the nose.

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