Silent sinus syndrome – one more reason for an ophthalmologist to have a rhinologist as a good friend

Sindrom tihog sinusa – još jedan razlog za oftalmologa da ima rinologa kao dobrog prijatelja

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Introduction

Silent sinus syndrome (SSS) is a rare condition involving the maxillary sinus, characterized by unilateral collapse of the maxillary sinus and orbital floor, associated with negative antral pressure in the absence of sinus symptoms 1,2. It is also known as imploding antrum syndrome and typical radiological findings are ipsilateral depression of the orbital floor and opacification of a collapsed maxillary sinus 1, 2. There has been some more than 105 cases of SSS published in English literature so far (1). The largest case series with 22 patients was published by Kass et al. 3 in 1997.

SSS is characterized by spontaneous and progressive enophthalmos (“sunken” eye-eye recession into globe) and hypoglobus (globe displaced downward; a drop in the pupillary level), so it is common that these patients first present to ophthalmologist 1, 2, 4. Its development is gradual and progressive, so after a few months up to a few years may become symptomatic 1. Since patients present with ophthalmological complains, without any nasal or sinus symptoms, with painless course and slow development, the term “silent sinus” was introduced 5.

The first report of this entity was in 1964 in a paper written by Montgomery 6. His report was about patients who had diplopia and enophthalmos associated with collapse of the maxillary sinus. Wilkins and Kulwin 7, in their paper published in 1981, emphasized that there was no orbital trauma in patients with spontaneous, unilateral enophthalmos and hypoglobus associated with “asymptomatic, bone thinning, maxillary sinus disease” [seen on computed tomography (CT) scans]. Imploding antrum syndrome can be primary or secondary. Primary or spontaneous (SSS) is idiopathic, whereas secondary may arise from mid-face trauma (including surgery), rhinosurgery, chronic rhinosinusitis and has also been reported in less than 1% of patients after orbital decompression in Graves ophthalmopathy (thyroid eye disease) 1, 5, 8, 9.

SSS most commonly presents unilaterally, although there are reports on it being bilateral 4, 10. It occurs exclusively in maxillary sinus (there is one report about it in the frontal sinus) 1, 11. SSS presents in the third to forth decades of life and seems to affect both genders equally 9. The incidence of SSS is similar in the left and right maxillary sinuses (there might be a slight predominance for presenting on the right side – (57%) 4, 9. The average duration of the progressive, characteristic orbitopathies until presentation is 3 months (range 10 days – 2 years) 1, 9. Average enophthalmos at presentation is 2,96 mm (± 0.16 mm), average hypoglobus at presentation is 2.78 (± 0.25 mm) 5, 9, 12. Although mostly observed in adults, there have been reports on SSS in children 5, 13.

The syndrome’s typical constellation of symptoms and signs are: spontaneous, gradual and progressive enophthalmos and hypoglobus (ocular asymmetry – a drop in the pupillary level, deep upper lid sulcus), so patients have a various
degree of facial asymmetry (esthetic problem) 1, 2, 8, 15–17. The eye is retracted into the orbit (one more prominent) and downward placed (Figure 1). There is normal ocular motility and vision, with no recurrent infections, pain or pressure. But, diplopia (usually in up gaze), cheek pressure, (intraorbital) facial pressure or mild pain, mild dental pain can also be the symptoms. Some other ophthalmological signs that can occur are abnormal eyelid signs (retraction, ptosis, absent crease), lid lag, and dry eyes from lagophthalmos 1, 2, 15–17.

Pathophysiology

SSS has two main theorized mechanisms: maxillary sinus atelectasis (MSA) which could be idiopathic, posttraumatic, or post-surgery, and maxillary sinus hypoplasia (MSH) 4.

The exact pathophysiology of SSS is unknown, and so far, there are three main theories 1, 3, 5, 9, 15, 17. A complete obstruction of the MS ostium results in hypoventilation and accumulation of secretion. In this enclosed cavity subsequent gas resorption leads to subatmospheric pressure (continued negative pressure within the sinus) 3, 5, 15, 17–19. The chronic negative pressure in the sinus slowly retracts the orbital floor, altering orbital anatomy and affecting the function of orbital contents. Continued negative pressure within the sinus activates the osteoclasts; in turn, these make the sinus walls thinner. The enophthalmos and hypoglobus are induced because the thinned orbital floor cannot support the pressure from the overlying orbital content, which gradually expands into the sinus 3, 5, 15, 17–19.

There is a hypothesis suggesting that lateraled middle turbinate may act as one-way pneumatic valve, leading to progressive reduction of air in the antrum and subsequent collapse of the maxillary sinus 1, 16.

However, given the rarity of SSS and the very high prevalence of maxillary sinus ostiomeatal obstruction, Hourany et al. 20 has placed this first theory explanation under question. The prolonged negative pressure theory also fails to offer an explanation for the exclusive involvement of the maxillary sinus, so he supposed that some other compounding factors such as trauma or anatomic predisposition play a role 20.

The second theory of SSS pathogenesis is inflammatory erosion 15. Chronic inflammation could induce the erosion of the orbital floor. Inflammatory cells can produce cytokines that inhibit the replication of osteoblasts and collagen synthesis, thus favoring the osteopenic process. Subclinical inflammatory process can induce osteopenia, resulting in maxillary bone loss, but the negative pressure in the maxillary sinus must also take part in the mechanisms.

The third theory involves hypoplasia 15. SSS is supposedly the result of an infection in a congenitally hypoplastic maxillary sinus, but one third of the patients have no history of sinus disease in childhood and SSS occurs in normal and well-developed maxillary sinus.

Diagnosis

The diagnosis of SSS is made by typical clinical features – gradual onset of enophthalmos and/or hypoglobus, in the absence of orbital trauma (including surgery) or sinus disease, nasal endoscopy and CT scans of the nose and paranasal sinuses 1, 8.

Nasal endoscopy will show either a completely normal finding, or one of the two typical pictures: firstly, widened middle meatus on the affected side with inward retraction of the uncinate process 15 and secondly, middle turbinate lateralyzed (middle meatus obscured due to lateral displacement of the middle turbinates toward the uncinate process) 9, 15, 21.

Imaging of the choice for SSS is CT 16, 22–24. CT finding is typical and definitely confirms the diagnosis 1, 4, 16, 20, 22–24. At coronal CT scans, we shall see the uncinate process and medial sinus wall retracted laterally; medial meatus enlarged, orbital floor retracted into sinus lumen, increased orbital volume, inward retraction of medial and superior walls of the MS, decrease in the MS volume and its total opacification (Figure 2). The maxillary sinus can be normally developed or hypoplastic, but is opacified, and the infundibulum is always occluded 1, 20. Occlusion is usually caused by lateral retraction of the uncinate process with the uncinate process being retracted against the inferomedial aspect of the orbital wall. The middle meatus is correspondingly enlarged with varying degrees of lateral retraction of the middle turbinate. The most characteristic imaging feature of the silent sinus syndrome is the inward retraction of the sinus walls into the sinus lumen, with associated decrease in sinus volume, which can be nicely seen on axial CT scans. The orbital floor (maxillary roof) is always retracted and commonly thinned, demineralized 1, 4, 20. Axial CT scans will show maxillary sinus opacity with inward bowing (“implosion”) of the maxillary sinus walls and associated lateral displacement of the middle turbinate (Figure 3). Maxillary sinus “implosion” means ret-

Fig. 1 – Typical ocular (facial) asymmetry in silent sinus syndrome (SSS) – hypoglobus and enophthalmos (3 mm) on the left side; upper-lid retraction, deepened upper-lid sulcus.

Coronal computed tomography (CT) scan: lateralized left uncinate process and medial maxillary sinus wall, enlarged left middle meatus, completely opacified left maxillary sinus, and its volume decreased.

Axial computed tomography (CT) scan: inward retracted walls of the left maxillary sinus, left maxillary sinus completely opacified, and its volume decreased.

The distinction between the imaging appearance of hypoplastic maxillary sinus and silent sinus syndrome is not well understood. Some authors believe that a congenital underdevelopment of the maxillary sinus is responsible for the development of SSS, but the acquired nature of this condition is now more readily apparent.

Chronic maxillary atelectasis is the term that describes a persistent decrease in the sinus volume from inwardly bowing antral walls. CMA was also differentiated in 3 stages based on the observed anatomical changes on CT scans. Stage 3 CMA (clinical deformity) is diagnosed when enophthalmos, hypoglobus, and/or midfacial deformity is noted. The presence of sinus-related symptoms distinguishes CMA stage III from SSS. Brandt and Wright are some of the authors that support the concept that SSS and CMA are the same clinical entity, because SSS fits within the staging classification of CMA. They suggested abandoning the term SSS and recommend universal adoption of the CMA staging system, which uses nomenclature that more accurately portrays the pathophysiology and natural history of this condition.

A clinician treating a patient presenting with enophthalmos and hypoglobus, must also consider a wide range of diseases. The differential diagnosis for SSS includes trauma to the orbit (especially blow out fracture), prior orbital decompression for Graves orbitopathy, chronic rhinosinusitis, osteomyelitis, Wegener granulomatosis, orbital metastasis, human immunodeficiency virus (HIV) lipodystrophy, and prior orbital radiation therapy. There are also some really rare conditions to consider like orbital fat atrophy, Recklinghausen disease (the absence of the sphenoid wing), linear scleroderma, Parry-Romberg syndrome (progressive hemifacial atrophy) and pseudoenophthalmos.

Treatment

The treatment of SSS has objectives to restore the eye position and orbital floor height, prevent progression of enophthalmos, restore ventilation and drainage of sinus and

Differential diagnosis

The changed architecture of maxillary sinus seen in SSS should be differentiated from the MSH and chronic maxillary atelectasis (CMA).

Maxillary sinus hypoplasia or failure of development (arrested pneumatisation), is an infrequent congenital anomaly, and can occur in the absence of disease or surgery. This is often accompanied by hypoplasia of the uncinate process. The prevalence of 10.4% has been described, with a proposed classification of the degree of hypoplasia based on CT appearances in three types.
Eight. Pic medial maxillectomy are done. Inside the maxillary sinus, in some cases an inferior meatal antrostomy with even endoscopic sinus surgery (FESS) that should be performed in patients with SSS, because it restores sinus drainage with no or minimal collateral damage. The procedure must be done with extreme caution, since unintentional entry into the orbit happens much more often. Surgery starts as usually with uncinectomy which should be performed with extra care, as the inward implosion of the antral walls, in combination with a depressed orbital floor, places the orbital contents at particular risk of injury. Then, a wide meatallantrostomy must be made, which provides aeration to the maxillary sinus. Antrostomy typically results in the release of negative sinus pressure and re-expansion of the collapsed cavity leading to the reduction of maxillary ostium and wide middle meatal antrostomy. So, a rhinosurgeon should perform a complete uncinectomy, anterior ethmoidectomy (adds exposure of the hiatus semilunaris and medial orbital wall), trimming of the inferior third of the middle nasal turbinate with gentle medial displacement (if it is lateralized) to prevent reocclusion of the natural maxillary ostium and wide middle meatal antrostomy.

In some cases an inferior meatal antrostomy with even endoscopic maxilllectomy are done. Inside the maxillary sinus, the mucus secretion (thick glue like) is often found, which provides aeration to the maxillary sinus. Antrostomy typically results in the release of negative sinus pressure and re-expansion of the collapsed cavity leading to the reduction of maxillary ostium and wide middle meatal antrostomy. So, a rhinosurgeon should perform a complete uncinectomy, anterior ethmoidectomy (adds exposure of the hiatus semilunaris and medial orbital wall), trimming of the inferior third of the middle nasal turbinate with gentle medial displacement (if it is lateralized) to prevent reocclusion of the natural maxillary ostium and wide middle meatal antrostomy. In some cases an inferior meatal antrostomy with even endoscopic maxilllectomy are done. Inside the maxillary sinus, the mucus secretion (thick glue like) is often found, and removed.

Besides the described traditional sinus treatment with FESS in SSS, there is a report on successful ball treatment with balloon sinuplasty.

The second stage in the treatment of SSS is a surgical procedure done to restore orbital volume and symmetry. Should an orbital wall repair be performed at the same time with sinus surgery (FESS), depends on the severity of diplopia, the degree of cosmetic alterations, and the postsurgical evaluation of the sinus. Actually, the timing for management of the orbital floor is still under debate. As suggested by some authors, orbital floor reconstruction should be performed simultaneously with sinus treatment. Most other authors think that drainage of the sinus as a single step should be enough. Orbital floor repair is absolutely indicated.

**Conclusion**

Silent sinus syndrome is a rare entity of spontaneous progressive asymptomatic collapse of the maxillary sinus. The diagnosis is based on the gradual onset of enophthalmos and/or hypoglobus, in the absence of orbital trauma (including surgery) or prior symptoms of sinus disease. Treatment is surgical, meaning functional endoscopic sinus surgery as the first and necessary step, and orbital floor repair performed in some cases, as the second step.

Silent sinus syndrome describes a constellation of ocular and sinonasal findings, so both otorhinolaryngologists and ophthalmologists should be familiar with it.

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