PRACTICAL ADVICE FOR PHYSICIANS

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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 as a cause of the clinical simptoms and signs, although up till then it was known it happens only as a consequence of orbital trauma. Soparkar et al. [5], in their paper published in 1994, introduced the term silent sinus syndrome. They described a large group of 14 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) [5, 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 [9, 13, 14].

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

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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 reccurent 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, 8, 15–17].

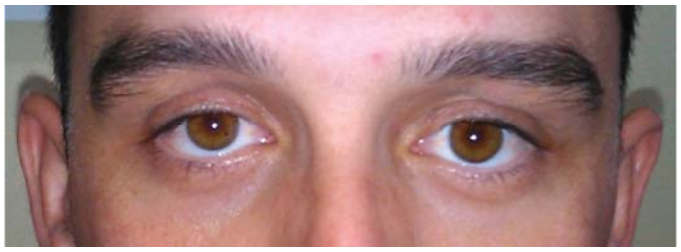

Fig. 1 – Typical ocular (facial) asymmetry in silent sinus syndrome (SSS) – hypoglobus and enophthalmos (3 mm) on the left side; upper-lid retraction, deepened upper-lid sulcus.

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].

The first theory is associated with prolonged negative pressure (continued negative pressure within the sinus) [3, 5, 15, 17–19]. 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 that creates vacuum, thus creating a suction effect of negative pressure within the maxillary sinus, vacuum may induce osteopenia, bone remodeling and sinus walls retraction (like eustachian tube dysfunction causing retraction in the middle ear) [15]. 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 enophtalmos 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 lateralized 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 ostiumobstruction, 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 supposingly 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, 9].

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 lateralized (middle meatus obscured due to lateral displacement of the middle turbinate 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-

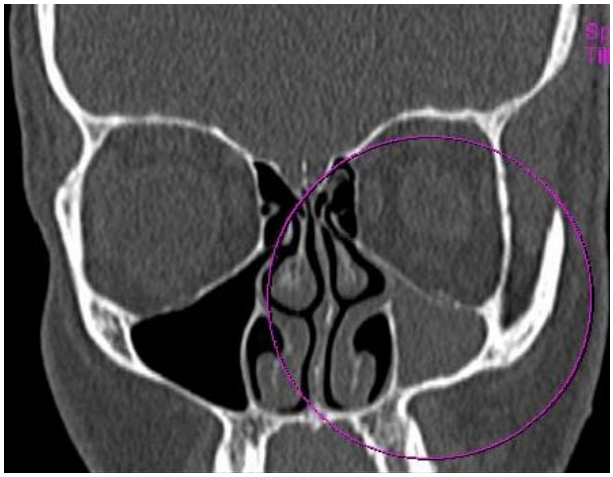

Fig. 2 – Coronal computed tomography (CT) scan: lateralized left uncinate process and medial maxillary sinus wall, enlarged left middle meatus, completely opacified left maxillary sinus, and its volume decreased.

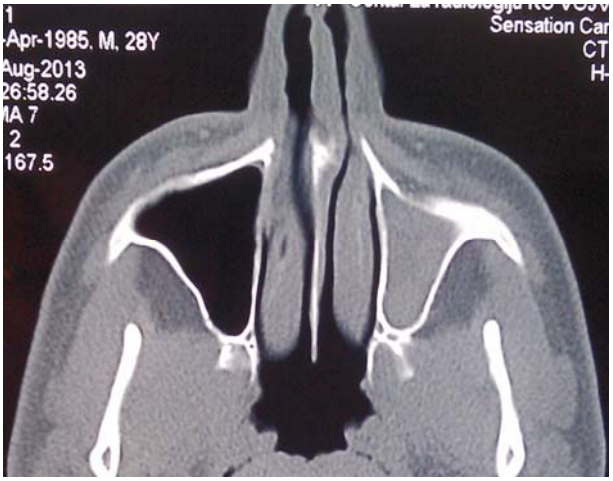

Fig. 3 – Axial computed tomography (CT) scan: inward retracted walls of the left maxillary sinus, left maxillary sinus completely opacified, and its volume decreased.

raction of anterior, posterior and medial wall into sinus lumen. There is also patchy loss of mineralisation. Typically, all 4 walls of the sinus are retracted, though one of the medial, anterior, or posterolateral walls may be spared [20].

The orbital floor is always retracted, commonly thinned, while the other walls may be thinned, normal, or slightly thickened [1, 4, 20, 25]. Orbital floor thickening is also a possibility. Considering the meaning of the thinned or thickened sinus walls in SSS, Hourany et al. [20] discused the issue in their paper, illustrating it with the case that supports the notion that SSS is an aquired condition, since thickening of the sinus walls is probably related to chronic inflammation and not to underlying developmental hypoplasia [20].The opacification of the MS can be complete or near complete (total or near total) [16, 18]. Sanchez et al. [25] described an image of a "pseudo-pneumo-orbit" that can also be seen due to air trapped under the upper eyelid.

The cases of lateralized uncinate process and increased orbital volumes observed on CT scans, but lacking clinical enophthalmos and hypoglobus exist, and are a matter of discussion if it is, or it is not the SSS. Wise et al. [26] in their paper published in 2007 conclude that it potentially represents early SSS, before the development of clinical orbital findings, so such cases should be considered and treated as an early stage of SSS.

Differential diagnosis

The changed architecture of maxillary sinus seen in SSS should be differentiated from the MSH and chronic maxillary atelectasis (CMA) [27].

Maxillary sinus hypoplasia or failure of development (arrested pneumatisation), is an infrequent congenital anomaly, and can occur in the absence of disease or surgery [27]. This is often accompanied by hypoplasia of the uncinate process. The prevalence of 10.4% has been described [4], with a proposed classification of the degree of hypoplasia based on CT appearances in three types [4, 27].

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 [20].

Chronic maxillary atelectasis is the term that describes a persistent decrease in the sinus volume from inwardly bowing antral walls [4, 12]. 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 [12] 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 [12].

A clinician treating a patient presenting with enophthalmos and hypoglobus, must also consider a wide range of diseases [9]. The differential diagnosis for SSS includes trauma to the orbit (especially blow out fracture), prior orbital decompression for Graves orbithopathy, chronic rhinosinusitis, osteomyelitis, Wegener granulomatosis, orbital metastasis, human immunodeficiency virus (HIV) lipodystrophy, and prior orbital radiation therapy [9, 17, 18, 28]. 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 [9, 17, 18, 20, 28].

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

avoid infection [1, 14, 17]. These objectives are achieved in single- or two- stage surgery.

The treatment of SSS is surgical, performed by a rhinologist [14]. The general agreement is that sinus pathology should be treated as the first stage of the treatment. It is the functional endoscopic sinus surgery (FESS) that should be performed in patients with SSS, because it restores sinus drainage with no or minimal collateral damage [1, 9, 12, 29, 30]. 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 [1, 9, 30]. Then, a wide meatalantrostomy 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 enopthalmos. A wide antrostomy prevents future reobstructions, and good reaeration of the sinus helps to avoid recurrent enophthalmos [4, 16, 29]. 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 [1, 16, 29, 30]. In some cases an inferior meatal antrostomy with even endoscopic medial maxillectomy are done [9]. Inside the maxillary sinus with SSS, the mucus secretion (thick glue like) is often found, and removed [15, 30].

Besides the described traditional sinus treatment with FESS in SSS, there is a report on succesfull ball treatment with balloon sinuplasty [31].

The second stage in the treatment of SSS is a surgical procedure done to restore orbital volume and symmetry [1]. 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 [4, 32]. 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 [4, 32]. Most other authors think that drainage of the sinus as a single step should be enough [1, 4, 16, 29, 30, 32]. The maxillary sinus usually remodels after FESS, so it is rarely necessary to do the secondary repair of the orbital floor for aesthetic reasons. Orbitopathies (enophthalmos and hypoglobus – cosmetic or symptomatic) improve over 6 months after surgery [1, 9], so a natural resolution of orbital findings and subjective complains happen. The recommendation today is to do the two-stage approach to orbital repair, as described, and do the second operation, if necessary, after at least 6 months [1, 9]. If a clinically significant and symptomatic enophthalmos or hypoglobus persist at 6 months after sinus surgery, the orbital floor repair is absolutely indicated [1, 9].

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 ophtalmologists should be familiar with it.

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