SAR Journal of Medical Case Reports

Abbreviated Key Title: *SAR J Med Case Rep*

Home page: http://sarmedjournals.com/sarjmcr/home

ISSN 2707-7780 (P)
ISSN 2709-6947 (O)

Case Report

Silent Sinus Syndrome

Dr. Dewan Chengappa B D[1], Dr. Elsa Mary Gipson[1], Dr. Preethi Poonja[2], Dr. Gowri P Bhandarkar[3], Dr. Prasanna Kumar Rao[4], Dr. Raghavendra Kini[5]*

[1]Intern, Department of Oral Medicine and Radiology, A.J Institute of Dental Sciences, Mangalore, Karnataka, India.
[2]Assistant Professor, Department of Oral Medicine and Radiology, A.J Institute of Dental Sciences, Mangalore, Karnataka, India.
[3]Reader, Department of Oral Medicine and Radiology, A.J Institute of Dental Sciences, Mangalore, Karnataka, India.
[4]Professor, Department of Oral Medicine and Radiology, A.J Institute of Dental Sciences, Mangalore, Karnataka, India.
[5]Professor and Head, Department of Oral Medicine and Radiology, A.J Institute of Dental Sciences, Mangalore, Karnataka, India

***Corresponding Author**
Dr. Raghavendra Kini

Article History: | Received: 25.02.2020 | Accepted: 14.03.2020 | Published: 28.03.2020 |

Abstract: Silent sinus syndrome (SSS) is a rare entity that is associated with spontaneous, painless, unilateral enophthalmos and hypoglobus resulting from downward bowing of the orbital floor in the absence of any symptomatic sinonasal disease. The diagnosis is suspected clinically, but it is confirmed radiologically by its characteristic imaging features that include maxillary sinus outlet obstruction, sinus opacification and sinus volume loss caused by inward retraction of the sinus walls. Ostium obstruction appears to play a critical role in the development of SSS. Treatment involves reconstructive procedure of orbital floor and functional endoscopic sinus surgery. The purpose of this article is to acquaint the reader with the clinical and imaging features of the SSS, along with a brief discussion about its pathogenesis and treatment, which is relatively unknown among oral and maxillofacial radiologists.

Keywords: Chronic maxillary atelectasia, displacement, enophthalmos, hypoglobus, endoscope.

INTRODUCTION

Silent Sinus Syndrome (SSS) is a rare condition that causes facial symmetry, unilateral painless enophthalmos and hypoglobus resulting from downward blowing of the orbital floor in the absence of any symptomatic sinonasal disease and diplopia with chronic negative pressure within the sinus (Choudhary, S. H. *et al*.,2016) (Sridhar, T. *et al*., 2018).

This diseases spectrum is also known by alternate names: “chronic maxillary atelectasis” and “imploding antrum syndrome (Burkat, C.N. *et al*.,). Some believe that congenital development of maxillary sinus may also be responsible for the development of SSS.

The diagnosis is suspected clinically, but it is confirmed radiologically by its characteristic imaging features that include maxillary sinus outlet obstruction, sinus opacification and sinus volume loss caused by inward retraction of the sinus walls (Choudhary, S. H. *et al*., 2016). Ostium obstruction appears to play a critical role in the development of SSS (Sridhar, T. *et al*., 208). Ocular manifestations are asymmetry due to exophthalmos in uninvolved contralateral eye or enophthalmos or ptosis of the affected eye. No histories of rhinosinusitis or significant sinus symptoms are evident. An orbital/paranasal sinus view in computerized tomography (CT) and magnetic resonance imaging (MRI) demonstrates opacification and collapse of orbit floor (Monos, T. *et al*., 2005).

HISTORY

In 1964, the first case of maxillary sinus opacification and collapse was reported by Montgomery,(Montgomery, W.W. 1964) but the name “Silent Sinus Syndrome” was coined 30 years later by Soparkar *et al. in 1994.* In 1997 Kass *et al.* suggested the name “chronic maxillary atelectasis” (CMA), as a term describing 22 patients with similar findings. CMA, however, may be diagnosed in patients with chronic rhinosinusitis symptoms while the criteria of SSS stipulate that affected patients must have the absence of

these symptoms for diagnosis (Soparkar, C.N. et a., 1994; Kass, E.S. *et al*., 1997).

Additionally, CMA is divided into three stages, with stage I and II describing maxillary deformity in the absence of hypoglobus or enophthalmos (Kass, E.S. *et al*., 1997). While some strictly distinguish CMA and SSS as separate clinical entities distinguished by presence or absence of chronic sinusitis symptoms, others suggest that SSS is actually part of the spectrum of CMA (Brandt, M.G., & Wright, E.D. 2008). The tertiary stage of CMA, manifest clinical deformity including enophthalmos and hypoglobus, closely overlays the definition of SSS (Kass, E.S. *et al*., 1997).

More recently, Rose *et al*. have suggested the term "imploding antrum syndrome" to clarify and describe the more acute and symptomatic event of sinus implosion that happens rapidly after long periods of chronic underlying atelectasis (Rose, G.E. *et al*., 2003). Recently, in 2008, Brandt and Wright required SSS to be included under the common term chronic maxillary atelectasis (Cobb, A.R. *et al*., 2012).

EPIDEMIOLOGY

Silent Sinus Syndrome is usually diagnosed in patients in their 30s and 40s with a mean age of 39 and a range of 19 to 82 (Brandt, M.G., & Wright, E.D. 2008) (Rose, G.E. *et al*., 2003). There is no bias in gender. There is an equal reported incidence of right and left-sided disease with a slight predominance of right-sided disease. Cases of SSS reported in the literature are limited mostly to single case reports, with only a small number of larger case series that have been described till date. Of these reported cases, only a few describe true SSS (Choudhary, S. H. *et al*., 2016).

RISK FACTORS

Underlying aberrant nasal anatomy is thought to be the largest risk factor for silent sinus syndrome. Specifically, studies describe characteristic anatomical variations including ipsilateral nasal septum deviation, laterally deviated middle turbinate, and otherwise narrowed infundibular passage (Kass, E.S. *et al*., 1997; Rose, G.E. *et al*., 2003; Babar-Craig, H. *et al*., 2011; Rose, G.E. 2004). These anatomical variations are hypothesized to predispose to a "flap-valve" occlusion of the natural maxillary sinus ostium wherein the sinus becomes completely occluded allowing gas resorption and development of negative pressure in the sinus antrum (Kass, E. S. *et al*., 1996).

ETIOLOGY AND PATHOGENESIS

The etiology of SSS is not clearly understood. Several theories have been proposed to explain the sequelae of events. Soparkar *et al*. suggested maxillary hypoplasia to be the predisposing factor in some individuals that results in development of SSS. They assumed that SSS is the result of infection in the congenitally hypoplastic maxillary sinus (Choudhury, N., & Marais, J. 2010).

In their early reports, Montgomery named a mucocele as the cause of enophthalmos while Wilkins and Kulwin noted a strong association with ipsilateral chronic maxillary sinus disease (Montgomery, W.W. 1964) (Wilkins, R. B., & Kulwin, D. R. 1981). When silent sinus syndrome was described in 1994 by Soparkar *et al*., the disease process was initially thought to be related to pre-existing sinus hypoplasia (Soparkar, C.N. *et al*., 1994) This association, however, was disproven after a 1999 case report by Davidson *et al.* documented pre-disease imaging demonstrating normal maxillary sinus anatomy several years prior to the onset of symptoms and manifestations of SSS (Davidson, J. K. *et al*., 1999). This report thus strongly suggested that SSS occurs in patients with normal sinus size prior to disease development (Burkat, C.N. *et al*.,).

The most popular theory suggested by several authors is that hypoventilation of the maxillary sinus secondary to obstruction of ostium leads to the formation of chronic negative pressure. Several mechanisms of ostial occlusion propose that occlusion by persistent mucoid impaction, hypermobile infundibular wall or lateralized middle turbinate, mucocele or nasal polyp results in occluding of ostium or antrum (Sridhar, T. *et al*., 2018). This is similar to middle ear atelectasis that occurs secondary to Eustachian tube dysfunction.

In the presence or absence of sinusitis, the inflamed mucosa of hypoplastic maxillary sinus and antrum causes decreased ostial width, resulting in more frequent occurrence of occlusion. (Bolger, W. E. *et al*., 1990) The hypoventilation and occlusion of the sinus may lead to the development of negative pressure. This negative pressure may cause thinning and serious structural damage to sinus walls, including the orbital floor, leading to hypoglobus (Sridhar, T. *et al*., 2018).

In case of opening of maxillary ostium, the sinus fluid, which initially provided support for the thin orbital floor, may drain, resulting in further depression of the globe and orbital floor. (Sridhar, T. *et al*., 2018) Once the sinus becomes occluded and negative pressure develops, the sinus cavity is filled with a combination of mucus and acellular transudate (Kass, E. S. *et al*., 1996) due to reabsorption of the aqueous component of the natural sinus mucosal secretions (Soparkar, C.N. *et al*.,2004). This process of aeration loss is hypothesized to create chronic, low grade mucosal inflammation.

Secondary enophthalmos after bone removing orbital decompression, the severe nasal inflammation with marked expansion, and pressure of altered turbinate may cause prolonged impairment of antral drainage. Subatmospheric pressure associated with

retained secretions causes the implosion (Sridhar, T. *et al*., 2018).

Cigarette smoking and incidence of SSS has been reported as a negative association. Whereas the observers of this phenomenon have hypothesized that chronic inflammation and sinonasal disease caused by smoking may lead smokers to present with symptoms much earlier (Rose, G.E. *et al*., 2003; Soparkar, C.N. *et al*., 2004).

There has been an observed association of the late development of iatrogenic SSS after some bone-removing orbital decompressions for thyroid eye disease. Secondarily prolapsed fat into the ethmoidal infundibulum during the surgery occludes the maxillary sinus ostium. Development of SSS after orbital decompression may also be related to the removal of inferomedial bone strut important to maintaining the normal maxilla ethmoidal interface and sinus aeration (Rose, G.E. *et al*., 2003; Soparkar, C.N. *et al*., 2004).

CLINICAL PRESENTATION

Time course:

Silent sinus syndrome is most commonly a gradual and progressive disorder with a time course of weeks to months. The average duration of symptoms before presentation range from 4 to 8 months (Rose, G.E. *et al*., 2003; Soparkar, C.N. *et al*., 2004; Numa, W.A. *et al*., 2005).

Signs & symptoms:

Patients with silent sinus syndrome presents to an ophthalmologist for evaluation of a change in facial appearance, either with the “sunken eye” appearance or deepening of sulcus of the upper lid or with discomfort of an intermittent vague around the affected orbits. Enophthalmos is present in 98% to 100% present of cases with a mean measurement of 3.4 mm. Hypoglobus may be slightly less common than enophthalmos with a mean of 3.2mm (Rose, G.E. *et al*., 2003; Soparkar, C.N. *et al*., 2004; Sridhar, T. *et al*., 2018).

Some other signs that may be associated including upper eyelid retraction, superior orbital sulcus deepening, fat loss in lower eyelid, malar depression, widening of middle meatus and ipsilateral retraction of the middle turbinate toward the affected side (Figure 1). Exophthalmos of the uninvolved contralateral eye may be reported occasionally. Usually, the visual function is unaffected, but a few patients have been reported with alterations in ocular motility or muscle imbalance resulting in diplopia (Choudhary, S. H. *et al*., 2016).

The proposed mechanism of diplopia is secondary to limitation of the superior and inferior oblique muscles, as their attachments to the bony orbit make them vulnerable to dysfunction when the globe becomes displaced in relationship to bony orbit (Soparkar, C.N. *et al*., 2004; Numa, W.A. *et al*., 2005).

Figure 1. Pseudoretraction of right upper eyelid due to silent sinus syndrome

DIAGNOSTIC CRITERIA

Criteria include: Z(Soparkar, C.N. *et al*., 1994; randt, M.G., & Wright, E.D. 2008; Burkat, C.N. *et al*.,)

1. Spontaneous Enophthalmos and/or hypoglobus
2. Absence of symptoms of sinonasal inflammatory disease
3. CT imaging showing contraction of maxillary sinus
4. Absence of history of orbital trauma, absence of enophthalmos due to another etiology
5. Absence of documented congenital deformity

RADIOGRAPHIC FEATURES

The diagnosis is usually done by otolaryngologists and ophthalmologists and the classical radiographic features are unique to SSS which can be clearly appreciated on computed tomographic (CT) (Figure 2) scan and MRI (Figure 3) (Sridhar, T. *et al*., 2018; Choudhary, S. H. *et al*., 2016). Focal absence of the sinus wall is most commonly seen in the medial wall and less frequently in the posterior, anterior, and lateral walls. The most characteristic feature of SSS is inward retraction of sinus wall into the lumen causes diminishing of sinus volume, and complete opacification of the affected maxillary sinus (Soparkar, C.N. *et al*., 1994).

Other radiographic findings include nasal septum deviation to the affected side, (Rose, G.E. *et al*., 2003; Soparkar, C.N. *et al*., 2004) enlarged middle meatus and anomalous direction of middle turbinate (Rose, G.E. *et al*., 2003). Rarely, concurrent collapse of a neighboring, ipsilateral ethmoid sinus has been documented (Rose, G.E. *et al*., 2003).

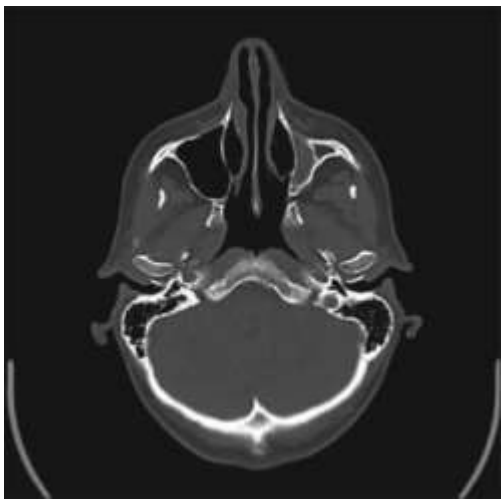

Figure 2: Computed tomography section

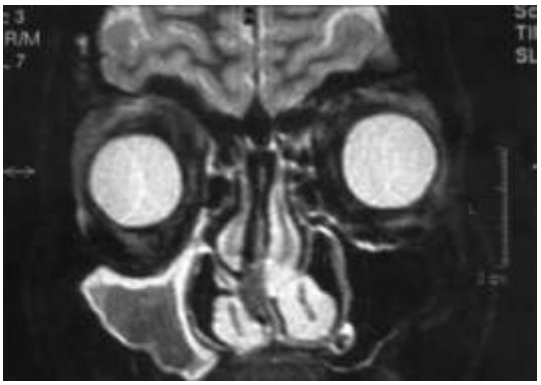

Figure 3: Magnetic resonance imaging

DIFFERENTIAL DIAGNOSIS

The differential diagnosis of silent sinus syndrome includes chronic sinusitis, osteomyelitis, malignant sinus infiltration, orbital trauma, linear scleroderma, pseudo-enophthalmos (contralateral exophthalmos, Horner'syndrome), atrophy of orbital contents and Parry- Romberg syndrome. Orbital content atrophy or contraction may be secondary granulomatous polyangitis, irradiation, or sclerosing pseudotumor (Sridhar, T. *et al*., 2018; Choudhary, S. H. *et al*., 2016; Burroughs, J.R. H. *et al*., 2003). In case of Parry–Romberg syndrome and scleroderma, there is no marked hypoglobus, and periorbital soft-tissue or contour changes are more frequent than in SSS (Burroughs, J.R. H. *et al*., 2003).

SSS can be differentiated from all the above-mentioned entities by its unique clinical and radiological features which include facial asymmetry due to asymptomatic unilateral enophthalmos and hypoglobus that usually progresses over a period of months with the absence of any history of ocular disease, orbital trauma or sinonasal disease. Radiographic features include inward retraction of the sinus walls resulting in volume loss, downward bowing of the orbital wall causing increased orbital volume, without any signs of fracture and enlarged middle meatus in some cases (Choudhary, S. H. *et al*., 2016).

MANAGEMENT

Surgical Management:

Management of SSS is mainly by two steps: **(a)** Clearing the sinus and recontouring a functional drainage passage and **(b)** reconstructing the normal orbital architecture. Earlier, surgical treatment was done by performing a Caldwell–Luc procedure, transconjunctival repair of the orbital floor (Choudhary, S. H. *et al*., 2016; Choudhury, N., & Marais, J. 2010;) or functional endoscopic sinus surgery (FESS) as an initial treatment followed by the reconstructive procedure with the placement of titanium mesh, autogenous nasal septal cartilage, spilt-thickness bone, or auricular concha cartilage onto the floor of the orbit. In some patients, the FESS alone can minimize the enophthalmos (Choudhary, S. H. *et al*., 2016; Soparkar, C.N. *et al*., 1994; Cobb, A.R. *et al*., 2012; Choudhury, N., & Marais, J. 2010).

Due to advances in endoscopic surgery, currently, endoscopic sinus surgery along with orbital floor repair – a single-stage procedure for creating a wide maxillary antrostomy is considered the surgical gold standard treatment for SSS. Common material of

choice for floor reconstruction of SSS are Medpor implant, silicone blocks, and Teflon sheets (Choudhury, N., & Marais, J. 2010; Numa, W.A. *et al.*).

Non-Surgical Management:

In 2011, nonsurgical approach using hyaluronic acid gel to correct enophthalmos after middle–meatal antrostomy was reported (Mavrikakis, I. *et al*., 2012). The gel was placed as a single bolus in the intra- and extraconal posterior orbit with a 21-gauge needle. Immediate improvement of the patient's unresolved enophthalmos and superior sulcus deformity was described.

CONCLUSION

Silent sinus syndrome is a rare acquired diseased condition with striking clinical and radiological features which is often misdiagnosed with controversial pathology. Significant feature is the absence of sinusitis symptoms and the history is usually not contributory. There is the need for all clinicians exposed to head and neck pathology to be familiar with SSS to ensure the correct diagnosis is made and optimal treatment is provided.

REFERENCES

1. Choudhary, S. H., Kale, L., Mishra, S. S., & Choudhary, A. K. (2016). Silent sinus syndrome: An imploding antrum syndrome. *Journal of Indian Academy of Oral Medicine and Radiology*, *28*(1), 30.
2. Sridhar, T., Karthika, B., Pavani, M., Iswarya, A. S., Shoban, D., & Priya, A. (2018). A review on silent sinus syndrome. *J Dent Allied Sci 7:81-3.*
3. Burkat, C.N., Merritt, H., & Silkiss, R.Z. Silent Sinus Syndrome. American Academy of Ophthalmology (//aao.org)
4. Monos, T., Levy, J., Lifshitz, T., & Puterman, M. (2005). The silent sinus syndrome. *Isr Med Assoc J;7:333-5.*
5. Montgomery, W.W. (1964). Mucocele of the maxillary sinus causing enophthalmos. *Eye Ear Nose Throat Mon 43:41-4.*
6. Soparkar, C.N., Patrinely, J.R., Cuaycong, M.J., *et al.*, (1994). The silent sinus syndrome. A cause of spontaneous enophthalmos. *Ophthalmology. 101(4):772-778.*
7. Kass, E.S., Salman, S., Rubin, P.A., Weber, A.L., & Montgomery, W.W. (1997). Chronic maxillary atelectasis. *Ann Otol Rhinol Laryngol.* 106(2),109-116
8. Brandt, M.G., & Wright, E.D. (2008).The silent sinus syndrome is a form of chronic maxillary atelectasis: A systematic review of all reported cases. *Am J Rhinol.* 22(1),68-73.
9. Rose, G.E., Sandy, C., Hallberg, L., & Moseley, I. (2003). Clinical and radiologic characteristics of the imploding antrum, or "silent sinus,"syndrome. *Ophthalmology.* 110(4), 811-818.
10. Cobb, A.R., Murthy, R., Cousin, G.C., El-Rasheed, A., Toma, A., Uddin, J., *et al., (2012).* Silent sinus syndrome. *Br J OralMaxillofac Surg 50:e81-5.*
11. Babar-Craig, H., Kayhanian, H., De Silva, D.J., Rose, G.E., Lund, V.J. (2011). Spontaneous silent sinus syndrome (imploding antrum syndrome): Case series of 16 patients. Rhinology. 49(3), 315-317.
12. Rose, G.E. (2004). Silent sinus syndrome -- new perspectives? Author reply. Ophthalmology (Rochester, Minn). 111(2), 415-416.
13. Kass, E. S., Salman, S., & Montgomery, W. W. (1996). Manometric study of complete ostial occlusion in chronic maxillary atelectasis. *The Laryngoscope*, *106*(10), 1255-1258.
14. Choudhury, N., & Marais, J. (2010). The silent sinus syndrome: A clinical review. *An Int J Clin RhinolClinical Rhinology: An International Journal*, *3*, 69-72.
15. Wilkins, R. B., & Kulwin, D. R. (1981). Spontaneous enophthalmos associated with chronic maxillary sinusitis. *Ophthalmology*, *88*(9), 981-985.
16. Davidson, J. K., Soparkar, C. N., Williams, J. B., & Patrinely, J. R. (1999). Negative sinus pressure and normal predisease imaging in silent sinus syndrome. *Archives of ophthalmology*, *117*(12), 1653-1654.
17. Bolger, W. E., Woodruff Jr, W. W., Morehead, J., & Parsons, D. S. (1990). Maxillary sinus hypoplasia: classification and description of associated uncinate process hypoplasia. *Otolaryngology—Head and Neck Surgery*, *103*(5), 759-765.
18. Soparkar, C.N., Patrinely, J.R., & Davidson, J.K. (2004). Silent sinus syndrome-new perspectives? Ophthalmology. 111(2), 414-5; author reply 415-6.
19. Rose, G.E., & Lund, V.J. (2003). Clinical features and treatment of late enophthalmos after orbital decompression: A condition suggesting cause for idiopathic "imploding antrum" (silent sinus) syndrome. Ophthalmology. 110(4), 819-826.
20. Numa, W.A., Desai, U., Gold, D.R., Heher, K.L., & Annino, D.J. (2005). Silent sinus syndrome: A case presentation and comprehensive review of all 84 reported cases. Ann Otol Rhinol Laryngol. 114(9), 688-694.
21. Burroughs, J.R. H., Cospín, J.R., Soparkar, C.N., & Patrinely, J.R. (2003). Misdiagnosis of silent sinus syndrome. Ophthalmic Plast Reconstr Surg 19,449-54.
22. Numa, W.A., Desai, U., Gold, D.R., Heher, K.L., & Annino, D.J. Silent sinus syndrome: A case presentation and comprehensive review of all 84 reported cases. *Ann Otol Rhinol Laryngol 114:688-94.*
23. Mavrikakis, I., Detorakis, E.T., Yiotakis, I., & Kandiloros, D. (2012). Nonsurgical management of silent sinus syndrome with hyaluronic acid gel. Ophthal Plast Reconstr Surg. 28(1),e6-7.