Restless Genital Syndrome Before and After Clitoridectomy for Spontaneous Orgasms: A Case Report

Marcel D. Waldinger, MD, PhD,*† Pieter L. Venema, MD, PhD,‡ Ad P.G. van Gils, MD, PhD,§ Eltjo M.J. Schutter, MD, PhD,¶ and Dave H. Schweitzer, MD, PhD**

*Department of Psychiatry and Neurosexology, HagaHospital Leyenburg, The Hague, The Netherlands; †Department of Psychopharmacology, Utrecht Institute for Pharmaceutical Sciences and Rudolf Magnus Institute for Neurosciences, Utrecht University, Utrecht, The Netherlands; ‡Outpatient Department of Continence, HagaHospital Leyenburg, The Hague, The Netherlands; §Department of Radiology and Nuclear Medicine, HagaHospital Leyenburg, The Hague, The Netherlands; ¶Department of Obstetrics and Gynaecology, Medisch Spectrum Twente, Enschede, The Netherlands; **Department of Internal Medicine and Endocrinology, Reinier de Graaf Groep of Hospitals, Delft-Voorburg, The Netherlands

DOI: 10.1111/j.1743-6109.2009.01571.x

ABSTRACT

Introduction. Females despairing of restless genital syndrome (ReGS) may request clitoridectomy for treatment of unwanted genital sensations.
Aim. The aim of this study was to report persistence of ReGS despite clitoridectomy.
Methods. Following a clitoridectomy for spontaneous orgasms, a 77-year-old woman was referred to our clinic for persistent unwanted genital sensations and feelings of imminent orgasm. An in-depth interview, routine and hormonal investigations, electroencephalography (EEG) and magnetic resonance imaging (MRI) of the brain and pelvis were performed. The localizations of genital sensations were investigated by manual examination of the ramus inferior of the pubic bone (RIPB) and by sensory testing of the skin of the genital area with a cotton swab.
Main Outcome Measures. The main outcome measures included sensitivity of dorsal nerve of the clitoris in RIPB and MRI-pelvis.
Results. Genital dysesthesias, paresthesias, intolerance (allodynia) for tight clothes, aggravation of symptoms during sitting, restless legs, and overactive bladder were diagnosed. Laboratory assessments, and EEG and MRI of the brain were in agreement with aging, but all results were within the normal range. MRI of the pelvis disclosed varices of the uterus and of the left ovarian vein, and a visible scar in the region of the clitoris. Sensory testing of the genital area showed various points of static mechanical hyperesthesia at the left dermatome of the pudendal nerve. Manual examination of the RIPB also elicited the genital sensations at the left side of the vagina at about the 3 o'clock position.
Conclusions. This patient fulfilled all clinical criteria of ReGS that is believed to be caused by neuropathy of the left pudendal nerve. Clitoridectomy abolished spontaneous orgasms for a great part but not completely, and it did not diminish the typical dysesthesias, paresthesias, and feelings of imminent orgasms that typically belong to ReGS. Clitoridectomy is no optional treatment of ReGS. There is a need for publications of ReGS in general medical journals. **Waldinger MD, Venema PL, van Gils APG, Schutter EMJ, and Schweitzer DH. Restless genital syndrome before and after clitoridectomy for spontaneous orgasms: A case report. J Sex Med **;**:**–**.**

Key Words. ReGS; PGAD; Persistent Sexual Arousal Syndrome; RLS; OAB; Pudendal Nerve; Dorsal Nerve of the Clitoris; Clitoridectomy

Introduction

In 2001, persistent sexual arousal syndrome (PSAS) was first reported in the medical literature by Leiblum and Nathan [1]. In 2006, it has been renamed persistent genital arousal disorder (PGAD) by Goldmeier and Leiblum [2]. In 2009, the combination of PGAD with restless legs syndrome (RLS) and/or overactive bladder syndrome (OAB) and/or urethral hypersensitivity has been called restless genital syndrome (ReGS) by Waldinger et al. [3,4]. The unwanted genital

sensations of ReGS are typically dysesthesias and/or paresthesias, and are often felt as an imminent orgasm in the absence of sexual desire or fantasies. In two systematic studies [3,5] it was found that the onset of ReGS usually occurs in perimenopausal and postmenopausal women with clinical characteristics of small fiber sensory neuropathy (SFSN) of the pudendal nerve, particularly the dorsal nerve of the clitoris (DNC). Moreover, the majority of women affected by ReGS has mild-to-moderate varices in the vagina, labia minora and/or majora, and uterus. However, whether pelvic varices are causally related to ReGS is still questioned, as pelvic varices are rather common in elderly women. Although ReGS is not associated with premorbid psychiatric disturbances, nearly all of these women report varying degrees of social withdrawal, desperate feelings, dysthymia, agitation, and depressed mood as a result of the persistent presence of the unwanted genital sensations. Some females become so desperate that they insist on undergoing clitoridectomy despite a lack of testimonials regarding the results of the surgery [3,4]. In this case report, we show that clitoridectomy in a female with ReGS may largely, but not completely, abolish spontaneous orgasms. It does not diminish unwanted restless genital sensations and feelings of imminent orgasm.

Case Report

Mrs. A. is a 77-year-old single woman who became widow at the age of 54 years. She is the mother of two children. At the age of 34 years she underwent reconstructive surgery for a prolapsed uterus, and at the age of 49 years she underwent an appendectomy. Her medical history also revealed a history of bronchitis. Nevertheless, the patient reported being a regular tobacco user. At 72 years the patient experienced spontaneous orgasms, originating in the clitoris, and a simultaneous absence of sexual desire, thoughts, or fantasies. These spontaneous orgasms, which were accompanied by movements of the extremities, gradually increased to two to three episodes each day. In addition, she experienced unusual throbbing sensations in the genitals as if on the verge of an orgasm. These genital sensations were aggravated during sitting, but diminished with walking. The sensations were associated with a stiff feeling in the abdominal muscles. She also experienced intolerance for wearing her clothes. The genital sensations and spontaneous orgasms upset her, and she felt desperate and finally became suicidal. Before her genital sensations started, she reported always having satisfying orgasms. There had never been complaints of depression, anxiety disorders, or obsessive–compulsive disorder. The genital sensations coincided with restless legs that usually occurred while she was asleep, and caused sleep disturbance almost every night. Additionally, soon after the onset of the genital sensations, she also experienced frequent urges to void (10–12 times a day), but with only small micturition. No incontinence of urine or bowel habits was recorded. Since she had lost her husband at the age of 54 years, she had not engaged in intercourse. At the age of 57 years she went into menopause, but prior to menopause, at the age of 51 years, she experienced the first genital sensation episode along with spontaneously occurring orgasms. The orgasms during this episode were less intense and less frequent, while the total period lasted for 1–1.5 years. Her complaints in this instance were part of a second episode that started at the age of 72 years. It took her 3 years before she dared to consult her general practitioner for her complaints. Initially, she was referred to a sexologist, but there was no therapy available to alleviate her symptoms. Later on, her general practitioner referred her to a gynecologist. Physical examination at that time disclosed a somewhat edematous clitoral region with a glans clitoridis that was covered by a fused labia minora as can be seen with lichen sclerosis et atroficans. The vaginal introitus was small, and the vaginal epithelium was atrophic. Routine laboratory determinations including urinary workup revealed no abnormalities. Her complaints were diagnosed at the time as clitoral dysesthesia presumably because of a chronic inflammatory reaction of the clitoris. As her symptoms persisted, she became desperate, and in a suicidal mood, she insisted in undergoing clitoridectomy. Initially, her gynecologist turned down this urgent request on the grounds that clitoridectomy would be disadvantageous or disappointing for her. Moreover, she was informed that after clitoridectomy, she would no longer have the ability to experience an orgasm. However, despite such medical objections to perform surgery, she continued to insist on an operation.

The clitoris and crura were completely resected, and on histology, all clitoral stromal tissues appeared normal but included an epidermal cyst. At that time it was concluded that this cyst must have been the cause of her complaints. Indeed, spontaneous orgasms did not recur post-surgery, for which she expressed her gratitude.

Regrettably, all typical dysesthesias, paresthesias, and feelings of imminent orgasms remained persistent. These sensations were present throughout the whole day and night with a pounding intensity that made her desperate. The sensations were located at the left side of the clitoris, around the vagina, and at the pubic bone. The unusual genital sensations persisted in the absence of sexual arousal or desire. Clonazepam was initially prescribed by her general physician, but with transient and limited effect. Finally, her general physician referred the patient to our Outpatient Department of Neurosexology in February 2009.

Here, the first author, following an evaluation procedure according to standard protocols, investigated the patient. After a neuropsychiatric and medical sexological interview that took 1 hour, the patient was clinically diagnosed as having ReGS. The woman had difficulties in describing the nature of her genital feelings, as it appeared to her that there were no adequate words. She described them as restless, irritating, and "weird" feelings; electric currents; and little spasms. The sensations were not described in terms of (sexual) arousal but as feelings that were similar as if she was about to get an orgasm. These pre-orgasmic feelings were continuously present. Routine laboratory assessments and endocrine laboratory screening were performed as well as electroencephalography (EEG) and magnetic resonance imaging (MRI) of the brain and pelvis. Finally, the urological examination included sensory testing with a cotton swab at the genital and pubic bone area, and included manual examination of the ramus inferior of the pubic bone (RIPB). The last examination was performed by the first and second authors in the presence of a trained nurse. According to the regulations of the medical ethical committee, official permission for study participation into our research of ReGS was not required, as the study is not placebo controlled and study drugs are not taken. The patient provided written informed consent for publication of details of her medical record.

After her referral to our clinic, the patient reported to have experienced two incidental full orgasms. The interview with this patient did not reveal prior child abuse, mood or anxiety disorder, compulsive disorder, traumatic sexual experiences, and alcohol or substance abuse. Routine laboratory assessments, including glucose and iron status, as well as hormonal assessments with thyroid screen were normal. The EEG was also normal. MRI of the brain was in agreement with aging. MRI of the pelvis, conducted without a Valsalva maneuver of the patient, disclosed varices in the anterior and posterior wall of the uterus and a dilated left-sided ovarian vein, and the signal intensity of scar tissue at the region of the clitoris (Figure 1). Sensory testing of the genital area showed various points that provoked the undesired sensations concentrated at the left side of the vagina. Furthermore, manual examination of the RIPB also elicited the genital sensations at the left side of the vagina at about 3 PM. Based on clinical symptoms, the diagnosis "small fiber sensory neuropathy of the left pudendal nerve, particularly the DNC" was established. In order to investigate the effect of local anesthetizing infiltration, the patient agreed to local injections with 1 cc bupivacaine hydrochloride monohydrate 0.5%. This anesthetic was injected under finger control in the vicinity of two trigger points. The infiltration anesthesia of the left pudendal nerve including the dorsal terminal branch to the clitoris led to a *transient*, but complete, disappearance of complaints, including RLS and the symptoms of OAB. This local infiltration therapy was effective for 72 hours, but by Day 4, all ReGS symptoms, including RLS and OAB, had recurred.

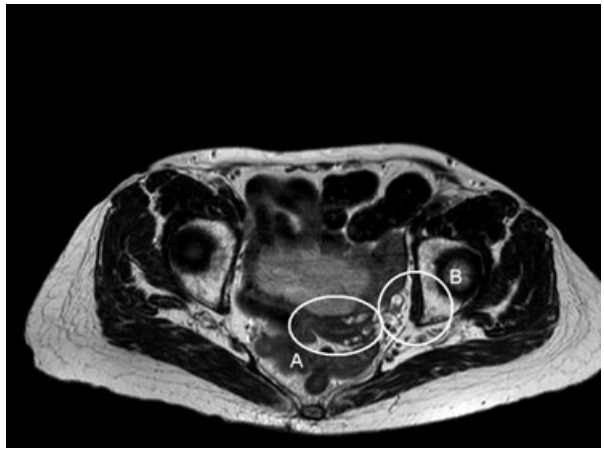

Figure 1 Transversal section of the pelvis. A = the small postmenopausal uterus contains varices in the anterior and posterior wall; B = the left ovarian vein is dilated.

Discussion

ReGS is a genital syndrome in women that usually begins during peri- and menopause. In a clinical series of 23 women who were diagnosed with ReGS and underwent in-depth interviewing, routine and hormonal investigations, EEG and

MRI of the brain and pelvis, physical examination including sensory testing of the dermatomes of the genital area, and manual examination of the RIPB, it was found that the majority is deeply suffering. This study, being the largest clinical series published so far, also revealed that women were ashamed for having inappropriate genital feelings but had decided to keep it for themselves. The study showed that 65% of women were in menopause with a mean age of 60.0 ± 6.7 years. They sought medical treatment 4.2 ± 3.9 years after the onset of the genital symptoms. The premenopausal women (35%) in these series were 43.3 ± 6.7 years, and decided to seek medical treatment at 0.9 ± 1.0 years after the initial onset of symptoms. A patient delay of 3 years, as what happened in the current case, is therefore in line with previously reported data of menopausal women.

Two clinical studies have shown that ReGS forms a cluster of well-established clinical symptoms including RLS and OAB [3,5]. Finally, ReGS includes persistent feelings of imminent orgasm but has not been reported with persistent spontaneous orgasms. The presence of spontaneous orgasms, as in the current patient, does not argue against the diagnosis of "ReGS." In this respect, it appears that this clinical presentation adds new important symptoms to previous descriptions of ReGS. The current lady is the first subject ever published who underwent clitoridectomy in an attempt to get rid of her complaints. Her family physician who had initially referred her to a sexologist and later to a gynecologist was at the time unaware of the existing, but scarce, literature on PSAS, PGAD, or ReGS. Similarly, her gynecologist later reported to us that he did not know the literature on this disorder. The patient explained to us that she herself insisted on, and deliberately convinced, her gynecologist to perform clitoridectomy. She was so desperate that she even wanted to commit suicide if clitoridectomy would have been rejected. Suicidal thoughts or intentions are certainly not exceptional for women affected by ReGS. In our first series of 18 women with ReGS, it appeared that 4 (22%) women temporarily insisted to undergo clitoridectomy, of whom some individuals could hardly accept that such a radical approach would not do any good to their situation [3,4]. A few other female patients reported considering clitoridectomy but realized that this would probably not be an adequate solution for the torments they were suffering from. Thoughts or even persistent desires for clitoridectomy in mentally, completely normal women with no history of psychiatric disorders show how immensely tormenting ReGS can be for women affected by it.

The consulted gynecologist described that prior to clitoridectomy, the labia were fused over the intact clitoris and the presence of a small atrophic vagina, however incompatible with lichen sclerosis et atrophicans. There were no biopsies taken. We confirmed this professional judgment after referral to our outpatient department. However, we disagreed with the initial working hypothesis of the gynecologist of clinical significance and consequence of a small clitoral epidermal cyst that was found on histology of the removed clitoris. Epidermal cysts of the clitoris are slowly growing intradermal or subcutaneous lesions with a wall composed of true epidermis [6]. The prevalence of clitoral cysts is very low [6,7]. Its occurrence has been associated with one of the possible complications of genital mutilation (female circumcision) [8,9]. However, as earlier mentioned, there were no prior traumatic sexual experiences or female circumcision in this patient.

The current case report provides first evidence that clitoridectomy is an inappropriate therapeutic measure for ReGS. The clitoridectomy resulted in a near disappearance of her spontaneous orgasms, but her persistent restless genital feelings remained. Besides, clitoridectomy to alleviate ReGS and spontaneous orgasms can even have undesired effects, as no data are available on tissue scarring, local pain, or paradoxal aggravation of ReGS post-surgery.

In line with our previous research findings [5], the patient had difficulties in describing the nature of her genital sensations. Like other women, she did not want to describe the sensations in terms of (sexual) arousal but preferred terms such as restlessness, electric currents, and little spasms, thereby noting that it felt as if she was about to get an orgasm. Also in line with our previous research findings [5] was the sensory testing of the genital area. Physical examination of the RIPB revealed signs and symptoms of ReGS, and included static mechanical hyperesthesia of the left pudendal dermatome. The presence of SFSN of the left pudendal nerve including its dorsal branch to the clitoris was clinically apparent by the findings of dysesthesias and allodynia, as we have also previously described [5]. However, SFSN can only be confirmed by specific electrophysiological tests. However, as these tests may be extremely painful in the genital region, it was not acceptable in this case.

Notably, the disappearance of symptoms after a locally injected bupivacaine in a non-edematous or inflamed residual scar tissue near the vicinity of the resected clitoris argues against the hypothesis of clitoral tissue abnormalities as a cause of ReGS. Lastly, the coincidental finding of a small cyst imbedded in stromal tissue does not explain the clinical symptoms.

The strong association with pelvic varices, as reported previously [3–5], suggests that high-pressure moments (labor, chronic constipation, or a prolapsed womb as in the current case) could have played a role in the pathogenesis. Even in the absence of Valsalva maneuvers during MRI, MRI scans showed clear pelvic varices in the uterus walls and in the left ovarian vein.

A remarkable expression of ReGS in the current patient was the occurrence of spontaneous orgasms and the incidental occurrence of ReGS at the onset of menopause followed by a long-lasting paucity of symptoms. Recurrence of symptoms occurred many years later without any obvious warning symptom such as trauma, or strenuous or stressful periods. A biphasic expression of ReGS during life has been described previously. In our study of 18 women with ReGS, 3 women reported that ReGS symptoms had also been present during pregnancy, and 4 women experienced ReGS symptoms in the second half of the menstrual cycle, long before the outbreak of ReGS later in life.

A new finding revealed local injection with 1 cc bupivacaine hydrochloride monohydrate 0.5% in the dermatome of the left pudendal nerve led to a *transient*, but complete, disappearance of complaints, including RLS and the symptoms of OAB, for the duration of 72 hours, after which, all ReGS symptoms, including RLS and OAB, had recurred. The continuation of ReGS symptoms after clitoridectomy, the incidental occurrence of full orgasms despite clitoridectomy, and the complete disappearance of symptoms after local infiltration indicate sparing of the left distal branch of the pudendal nerve despite the surgery. However, the complete disappearance of ReGS, RLS, and OAB after local anesthetic infiltration supports our view that ReGS is clinically and causally related to disturbances of the peripheral nervous system originating in the sacral spinal cord (S1-S5). The clinical symptoms of ReGS, such as dysesthesias, paresthesias, burning sensations, hyperesthesia, and allodynia, fit into small fiber sensory neuropathy (SFSN) [5]. As static mechanical hyperesthesia was elicited within the margins of the left dermatome of the pudendal nerve, it is most likely that ReGS of the current patient was associated with SFSN of the left pudendal nerve and its distal branches. This is in line with our previous study in which we found clinical evidence that ReGS is associated with SFSN of the pudendal nerve [5].

It has been shown that SFSN is associated with RLS [10]. In other words, peripheral neuropathy because of SFSN may give rise to RLS. The relationship of peripheral neuropathy-induced RLS and RLS that is caused by central dopaminergic dysfunction is unclear [11–13]. We presume that, in ReGS, certain spinal factors, other than dopaminergic neurotransmission, are associated with pudendal neuropathy-induced RLS. Although a central dopaminergic cause for PGAD has recently been postulated [14], there currently are no data that show that RLS in ReGS is directly associated with central dopaminergic dysfunction. For a better insight and understanding, electrophysiological and clinical studies are needed. Such studies are being undertaken currently by our group.

Conclusion

In conclusion, this case report confirms that ReGS is most likely the manifestation of SFSN of the pudendal nerve, and particularly of the DNC. ReGS was originally described by our group without spontaneous orgasms, but in this case, spontaneous orgasms were part of the genital syndrome. Clitoridectomy did not completely suppress spontaneous orgasms, nor was it effective to suppress dysesthesias, paresthesias, allodynia, feelings of imminent orgasm, static mechanical hyperesthesia, RLS, and OAB. Consequently, clitoridectomy is considered inappropriate to treat ReGS. If an awareness of PSAS, PGAD, or ReGS had been greater, then clitoridectomy in the current patient would most likely have not been performed. This genital syndrome deserves more attention from clinicians as ignorance may lead to spurious treatments. The initial treatment with clonazepam was partially effective, as has also been reported in our previous study [3,4]. The message of this case report is clear: clitoridectomy is never an option to treat women with ReGS even if they try to persuade physicians. Suicidal thoughts of women with ReGS may also never form the argument for performing surgical genital mutilation. In addition, this case report signals the urgent need to provide more information on the existence and characteristics of PSAS, PGAD, and ReGS in general medical journals read by generalists who

practice outside the field of sexual medicine so that physicians become aware of the syndrome.

Corresponding Author: Marcel D. Waldinger, MD, PhD, Department of Psychiatry and Neurosexology, HagaHospital Leyenburg, Leyweg 275, 2545 CH, The Hague, The Netherlands. Tel: +31-70-210-2086; Fax: +31-70-210-4902; E-mail: md@waldinger.demon.nl

Conflict of Interest: None.

Statement of Authorship

Category 1

(a) Conception and Design
Marcel D. Waldinger

(b) Acquisition of Data
Marcel D. Waldinger; Pieter L. Venema; Ad P.G. van Gils

(c) Analysis and Interpretation of Data
Marcel D. Waldinger; Pieter L. Venema; Ad P.G. van Gils

Category 2

(a) Drafting the Article
Marcel D. Waldinger

(b) Revising It for Intellectual Content
Marcel D. Waldinger; Eltjo M.J. Schutter; Dave H. Schweitzer

Category 3

(a) Final Approval of the Completed Article
Marcel D. Waldinger; Pieter L. Venema; Ad P.G. van Gils; Eltjo M.J. Schutter; Dave H. Schweitzer

References

1 Leiblum SR, Nathan SG. Persistent sexual arousal syndrome: A newly discovered pattern of female sexuality. J Sex Marital Ther 2001;27:365–80.
2 Goldmeier D, Leiblum SR. Persistent genital arousal in women—A new syndrome entity. Int J STD AIDS 2006;17:215–6.
3 Waldinger MD, van Gils APG, Ottervanger HP, Vandenbroucke WVA, Tavy DLJ. Persistent genital arousal disorder in 18 Dutch women: Part 1. MRI, EEG and transvaginal ultrasonography investigations. J Sex Med 2009;6:474–81.
4 Waldinger MD, Schweitzer DH. Persistent genital arousal disorder in 18 Dutch women: Part II. A syndrome clustered with restless legs and overactive bladder. J Sex Med 2009;6:482–97.
5 Waldinger MD, Venema PL, van Gils APG, Schweitzer DH. New insights into restless genital syndrome: Static mechanical hyperesthesia and neuropathy of the nervus dorsalis clitoridis. J Sex Med 2009;6:2778–87.
6 Schmidt A, Lang U, Kiess W. Epidermal cyst of the clitoris: A rare clitorimegaly. Eur J Obstet Gynecol Reprod Biol 1999;87:163–5.
7 Teague JL, Anglo L. Clitoral cyst: An unusual cause of clitorimegaly. J Urol 1996;156:2057.
8 Rouzi AA, Sindi O, Radhan B, Ba'ageel H. Epidermal clitoral inclusion cyst after type I female genital mutilation. Am J Obstet Gynecol 2001;185:569–71.
9 Yoong WC, Shakya R, Sanders BT, Lind J. Clitoral inclusion cyst: A complication of type I female genital mutilation. J Obstet Gynaecol 2004;24: 98–9.
10 Polydefkis M, Allen RP, Hauer P, Earley CJ, Griffin JW, McArthur JC. Subclinical sensory neuropathy in late-onset restless legs syndrome. Neurology 2000;55:1115–21.
11 Gemignani F, Brindani F, Vitetta F, Marbini A, Calzetti S. Restless legs syndrome in diabetic neuropathy: A frequent manifestation of small fiber neuropathy. J Peripher Nerv Syst 2007;12: 50–3.
12 Trenkwalder C, Paulus W. Why do restless legs occur at rest? Pathophysiology of neuronal structures in RLS. Neurophysiology of RLS (part 2). Clin Neurophysiol 2004;115:1975–88.
13 Clemens S, Rye D, Hochman S. Restless legs syndrome. Revisiting the dopamine hypothesis from the spinal cord perspective. Neurology 2006; 67:125–30.
14 Korda JB, Pfaus JG, Goldstein I. Persistent genital arousal disorder: A case report in a woman with lifelong PGAD where serendipitous administration of varenicline tartrate resulted in symptomatic improvement. J Sex Med 2009;6:1479–86.