

Secretan Syndrome: A Fluctuating Case of Factitious Lymphedema

Deniz Demircioğlu, MD; Emel Öztürk Durmaz, MD; Engin Sezer, MD; Sedef Şahin, MD

PRACTICE POINTS

- Secretan syndrome is a recurrent or chronic form of factitious lymphedema that usually affects the dorsal aspect of the hand; it is accepted as a subtype of Munchausen syndrome.
- Secretan syndrome usually is induced by compression of the extremity by tourniquets, ligatures, cords, or similar equipment.
- This unconsciously motivated and consciously produced lymphedema is an expression of underlying psychiatric disease.

Secretan syndrome (SS) is a recurrent or chronic form of factitious lymphedema that usually affects the dorsal aspect of the hand and is accepted as a subtype of Munchausen syndrome. Secretan syndrome usually is induced by compression of the extremity by tourniquets, ligatures, cords, or similar equipment. This unconsciously motivated and consciously produced lymphedema is an expression of underlying psychiatric disease. Herein, we present a fluctuating case of SS involving the hand, with periods of severe lymphedema and those of complete amelioration, parallel to stressful and peaceful periods in the patient's personal life. We briefly review the current literature on SS to increase awareness among dermatologists, and we present information on the clinical portrait, diagnosis, and appropriate management of this peculiar and underreported disorder.

Cutis. 2021;108:E23-E25.

Secretan syndrome (SS) represents a recurrent or chronic form of factitious lymphedema, usually affecting the dorsal aspect of the hand.¹⁻³ It is accepted as a subtype of Munchausen syndrome whereby the patient self-inflicts and simulates lymphedema.^{1,2} Historically, many of the cases reported with the term

Charcot's oedème bleu are now believed to represent clinical variants of SS.⁴⁻⁶

Case Report

A 38-year-old Turkish woman presented with progressive swelling of the right hand of 2 years' duration that had caused difficulty in manual work and reduction in manual dexterity. She previously had sought medical treatment for this condition by visiting several hospitals. According to her medical record, the following laboratory or radiologic tests had revealed negative or normal findings, except for obvious soft-tissue edema: bacterial and fungal cultures, plain radiography, Doppler ultrasonography, lymphoscintigraphy, magnetic resonance imaging, fine needle aspiration, and punch biopsy. Reflex sympathetic dystrophy, compartment syndrome, filariasis, tuberculosis, and lymphatic and venous obstruction were all excluded by appropriate testing. Our patient was in good health prior to onset of this disorder, and her medical history was unremarkable. There was no family history of a similar condition.

Dermatologic examination revealed brawny, soft, pitting edema; erythema; and crusts affecting the dorsal aspect of the right hand and proximal parts of the fingers (Figure 1). The yellow discoloration of the skin and nails was attributed to potassium permanganate wet dressings. Under an elastic bandage at the wrist, which the patient unrolled herself, a sharp line of demarcation was evident, separating the lymphedematous and normal parts of the arm. There was no axillary lymphadenopathy.

The patient's affect was discordant to the manifestation of the cutaneous findings. She wanted to show every physician in the department how swollen her hand was and seemed to be happy with this condition.

From the Department of Dermatology, Acibadem Mehmet Ali Aydınlar University School of Medicine, İstanbul, Turkey.

The authors report no conflict of interest.

Correspondence: Emel Öztürk Durmaz, MD, Acibadem Maslak Hospital, Büyükdere Caddesi 40, Maslak 34457, İstanbul, Turkey

(emelerkek@yahoo.com).

doi:10.12788/cutis.0425

Although she displayed no signs of disturbance when the affected extremity was touched or handled, she reported severe pain and tenderness as well as difficulty in housework. She noted that she normally resided in a city and that the swelling had started at the time she had relocated to a rural village to take care of her bedridden mother-in-law. She was under an intensive workload in the village, and the condition of the hand was impeding manual work.



FIGURE 1. A, Brawny, soft, pitting edema; erythema; and crusting on the dorsal aspect of the right hand and proximal parts of the fingers. B, An evident line of demarcation was noted at the wrist.

Factitious lymphedema was considered, and hospitalization was recommended. The patient was then lost to follow-up; however, one of her relatives noted that the patient had returned to the city. When she presented again 1 year later, almost all physical signs had disappeared (Figure 2), and a psychiatric referral was recommended. A Minnesota Multiphasic Personality Inventory test yielded an invalid result due to the patient's exaggeration of her preexisting physical symptoms. Further psychiatric workup was rejected by the patient.

Almost a year after the psychiatric referral, the patient's follow-up photographs revealed that the lymphedema recurred when she went to visit her mother-in-law in the rural village and that it was completely ameliorated when she returned to the city. Thus, a positive "mother-in-law provocation test" was accepted as final proof of the self-inflicted nature of the condition.

Comment

In 1901, Henri Francois Secretan, a Swiss physician, reported workmen who had persistent hard swellings on the dorsal aspect of the hands after minor work-related trauma for which they had compensation claims.⁷ In his original report, Secretan did not suggest self-inflicted trauma in the etiology of this disorder.^{5,8,9} In 1890, Jean Martin Charcot, a French neurologist, described *oedème bleu*, a term that is now believed to denote a condition similar to SS.⁴⁻⁶ Currently, SS is attributed to self-inflicted injury and is considered a form of factitious lymphedema.⁹ As in dermatitis artefacta, most patients with SS



FIGURE 2. Complete regression of lymphedema was noted 1 year after the initial presentation.

are young women, and male patients with the condition tend to be older.^{3,8}

The mechanism used to provoke this factitious lymphedema might be of traumatic or obstructive nature. Secretan syndrome either is induced by intermittent or constant application of a tourniquet, ligature, cord, elastic bandage, scarf, kerchief, rubber band, or compress around the affected extremity, or by repetitive blunt trauma, force, or skin irritation.^{1,4,5,8-10} There was an underlying psychopathology in all reported cases.^{1,8,11} Factitious lymphedema is unconsciously motivated and consciously produced.^{4,12} The affected patient often is experiencing a serious emotional conflict and is unlikely to be a malingerer, although exaggeration of symptoms may occur, as in our patient.¹² Psychiatric evaluation in SS may uncover neurosis, hysteria, frank psychosis, schizophrenia, masochism, depression, or an abnormal personality disorder.^{1,12}

Patients with SS present with recurrent or chronic lymphedema, usually affecting the dominant hand.¹ Involvement usually is unilateral; bilateral cases are rare.^{3,6} Secretan syndrome is not solely limited to the hands; it also may involve the upper and lower extremities, including the feet.^{3,11} There may be a clear line of demarcation, a ring, sulcus, distinct circumferential linear bands of erythema, discoloration, or ecchymoses, separating the normal and lymphedematous parts of the extremity.^{1,4,6,8-10,12} Patients usually attempt to hide the constricted areas from sight.¹ Over time, flexion contractures may develop due to peritendinous fibrosis.⁶ Histopathology displays a hematoma with adhesions to the extensor tendons; a hematoma surrounded by a thickened scar; or changes similar to ganglion tissue with cystic areas of mucin, fibrosis, and myxoid degeneration.^{4,6}

Factitious lymphedema can only be definitively diagnosed when the patient confesses or is caught self-inflicting the injury. Nevertheless, a diagnosis by exclusion is possible.⁴ Lymphangiography, lymphoscintigraphy, vascular Doppler ultrasonography, and magnetic resonance imaging may be helpful in excluding congenital and acquired causes of lymphedema and venous obstruction.^{1,3,9,11} Magnetic resonance imaging may show soft tissue and tendon edema as well as diffuse peritendinous fibrosis extending to the fascia of the dorsal interosseous muscles.^{3,4}

Factitious lymphedema should be suspected in all patients with recurrent or chronic unilateral lymphedema without an explicable or apparent predisposing factor.^{4,11,12} Patients with SS typically visit several hospitals or institutions; see many physicians; and willingly accept, request, and undergo unnecessary extensive, invasive, and costly diagnostic and therapeutic procedures and prolonged hospitalizations.^{1,2,5,12} The disorder promptly responds to immobilization and elevation of the limb.^{2,4} Plaster casts may prove useful in prevention of compression and thus amelioration of the lymphedema.^{1,4,6} Once the diagnosis is confirmed, direct confrontation should be avoided and ideally the patient should be referred for psychiatric evaluation.^{1,2,4,5,8,12} If the patient admits self-inflicting behavior, psychotherapy and/or behavior modification therapy along with psychotropic medications may be helpful to relieve emotional and behavioral symptoms.¹² Unfortunately, if the patient denies a self-inflicting role in the occurrence of lymphedema and persists in self-injurious behavior, psychotherapy or psychotropic medications will be futile.⁹

REFERENCES

1. Miyamoto Y, Hamanaka T, Yokoyama S, et al. Factitious lymphedema of the upper limb. *Kawasaki Med J*. 1979;5:39-45.
2. de Oliveira RK, Bayer LR, Lauxen D, et al. Factitious lesions of the hand. *Rev Bras Ortop*. 2013;48:381-386.
3. Hahm MH, Yi JH. A case report of Secretan's disease in both hands. *J Korean Soc Radiol*. 2013;68:511-514.
4. Eldridge MP, Grunert BK, Matloub HS. Streamlined classification of psychopathological hand disorders: a literature review. *Hand (NY)*. 2008;3:118-128.
5. Ostlere LS, Harris D, Denton C, et al. Boxing-glove hand: an unusual presentation of dermatitis artefacta. *J Am Acad Dermatol*. 1993;28:120-122.
6. Winkelmann RK, Barker SM. Factitial traumatic panniculitis. *J Am Acad Dermatol*. 1985;13:988-994.
7. Secretan H. Oederne dur et hyperplasie traumatique du metacarpe dorsal. *RevMed Suisse Romande*. 1901;21:409-416.
8. Barth JH, Pegum JS. The case of the speckled band: acquired lymphedema due to constriction bands. *J Am Acad Dermatol*. 1986;15:296-297.
9. Birman MV, Lee DH. Factitious disorders of the upper extremity. *J Am Acad Orthop Surg*. 2012;20:78-85.
10. Nwaejike N, Archbold H, Wilson DS. Factitious lymphoedema as a psychiatric condition mimicking reflex sympathetic dystrophy: a case report. *J Med Case Rep*. 2008;2:216.
11. De Fátima Guerreiro Godoy M, Pereira De Godoy JM. Factitious lymphedema of the arm: case report and review of publications. *Eur J Phys Rehabil Med*. 2015;51:337-339.
12. Abhari SAA, Alimalayeri N, Abhari SSA, et al. Factitious lymphedema of the hand. *Iran J Psychiatry*. 2006;1:166-168.