Subcutaneous Sweet's Syndrome Associated with Progressive Bilateral Sensory Neural Hearing Loss: A Case Report and Review of Literature

Hiathem Abu Al-haija [1], Nidal A. Obaidat, [1] Mohammed Tawara, [1] Mamoun Athamneh, [1] Omar AshUKaibi, [2] Bayan Abu Al-haija [1]*

Abstract

Sweet's syndrome is an autoimmune inflammatory condition, characterized by erythematous plaques infiltrated by neutrophils limited to the dermis. If the infiltrate just present in or extend to the subcutaneous tissue, then it will be labeled as subcutaneous Sweet's syndrome. Due to the rarity of this condition, its association with other medical conditions is inconclusive.

Herein, we present a male patient who underwent cochlear implant after having progressive bilateral sensory neural hearing loss. Two months after the implant, he developed cutaneous lesions consistent with subcutaneous Sweet's syndrome, which were well controlled with systemic steroids and cyclosporine.

Four previous cases of Sweet's syndrome followed by progressive bilateral sensory neural hearing loss were reported in the literature. Twenty-one cases of subcutaneous Sweet's syndrome associated with different medical conditions were reported so far. The present case is the first, up to our knowledge, of progressive bilateral sensory neural hearing loss that was followed by subcutaneous Sweet's syndrome.

Keywords: Neutrophilic dermatosis, progressive bilateral sensory neural hearing loss, Sweet's syndrome, subcutaneous Sweet's syndrome.

(J Med J 2014; Vol. 48 (4):292- 304)

Received	***Accepted***
Jan. 5, 2014	*June 23, 2014*

Introduction

Sweet's syndrome (SS), or acute febrile neutrophilic dermatosis, was first described in 1964 by Robert Sweet in a group of 8 female patients[1]. It typically presents with the following clinical triad: erythematous cutaneous plaques infiltrated by mature neutrophils, in association with fever and leukocytosis[1, 2]. Criteria for diagnosing SS are outlined in table 1. Histologically, SS is characterized by diffuse neutrophilic infiltrate

1. Department of Dermatology, King Hussein Medical Center, Amman, Jordan.
2. Department of Pathology, King Hussein Medical Center, Amman, Jordan.

**** Correspondence should be addressed to:***
Dr. Hiathem Abu al-haija.
Section of Dermatology, Department of Internal Medicine.
King Hussein Medical Center.
Royal Medical Services.
E-Mail: hayathema@yahoo.com
P. O. Box: 1991, Amman 11821, Jordan.

© 2014 DAR Publishers / The University of Jordan. All Rights Reserved.

in the upper dermis with leukocytoclasia but no true vasculitis[2].

Table 1. Criteria for diagnosing Sweet's syndrome [4,8,10]

Major criteria	*1.* Abrupt onset of painful erythematous plaques or nodules. *2.* Histopathologic evidence of a dense neutrophilic infiltrate without evidence of leukocytoclastic vasculitis.
Minor criteria	*1.* Pyrexia >38°C *2.* Association with an underlying hematologic or visceral malignancy, inflammatory disease, or pregnancy, OR preceded by an upper respiratory or gastrointestinal infection or vaccination *3.* Excellent response to treatment with systemic corticosteroids or potassium iodide *4.* Abnormal laboratory values at presentation (three of four): erythrocyte sedimentation rate >20 mm/hr; positive C-reactive protein; >8,000 leukocytes; >70% neutrophils

** Two major and two minor criteria are needed for the diagnosis.*

The pathogenesis of this condition is still not well known, but an immune etiology has been implicated[3].SS presents in three clinical settings: classical (or idiopathic), malignancy-associated, and drug-induced[4].Classical SS is often preceded by an upper respiratory tract infection and may be associated with inflammatory bowel disease and pregnancy[4].Variants of SS include: plaque, bullous and vesicular, subcutaneous SS, neutrophilic dermatosis of dorsal hands, pustular variant with ulcerative colitis and erythema nodosum-like picture[4].

Although SS is primarily a dermal process, the neutrophilic infiltrate may spread to the underlying subcutaneous tissue[1,4,6,7].The term subcutaneous SS is recommended only for cases in which the neutrophilic infiltrate is exclusively or predominantly located in the subcutaneous tissue, regardless of whether the infiltrate predominates in the fat lobules or septae, although lobular predominance is more frequent[1,7-10]. The clinical manifestations in most cases of subcutaneous SS are nodules, whereas it is mostly plaques in classic SS[9,11].

Till now, there have been four reports of progressive bilateral sensory neural hearing loss (PBSNHL) in association with SS[12-15]. An autoimmune process targeting the inner ear may be responsible about PBSNHL[16, 17].We present a 43 year old male patient with PBSNHL who underwent cochlear implantation. Two months later he developed skin lesions consistent with subcutaneous SS, which were well controlled with systemic steroids and cyclosporine.

Case Report:

A 43 year old male patient was followed at the Ear Nose Throat(ENT) clinic as a case of idiopathic progressive bilateral sensory neural hearing loss (PBSNHL) since 3 years, who

underwent cochlear implantation. Two years after the onset of PBSNHL, he developed scleritis and followed at the Ophthalmology clinic. He was given intravenous methylprednisolone 1000mg for 3 days then he was maintained on oral prednisolone 60 mg per day for 1 week which was tapered slowly over 4 months.

He was referred to our clinic, two months after cochlear implant, due to linear (sporotrichoid) tender erythematous elevated painful nodules over both forearms. One week later he presented with painful erythematous plaques over left leg, which ulcerated over the next couple of weeks (figure 1). Skin biopsy was done from the leg nodule; showing spongiosis and mild neutrophilic infiltration of the epidermis with perivascular mixed infiltrate in the dermis (figure 2A). The main pathology was located in the subcutaneous tissue; showing dense neutrophilic infiltrate within the fat lobules, (figure 2B, 2C). Ziehl-Neelsen (ZN) and Periodic-Acid Schiff (PAS) stains were negative.

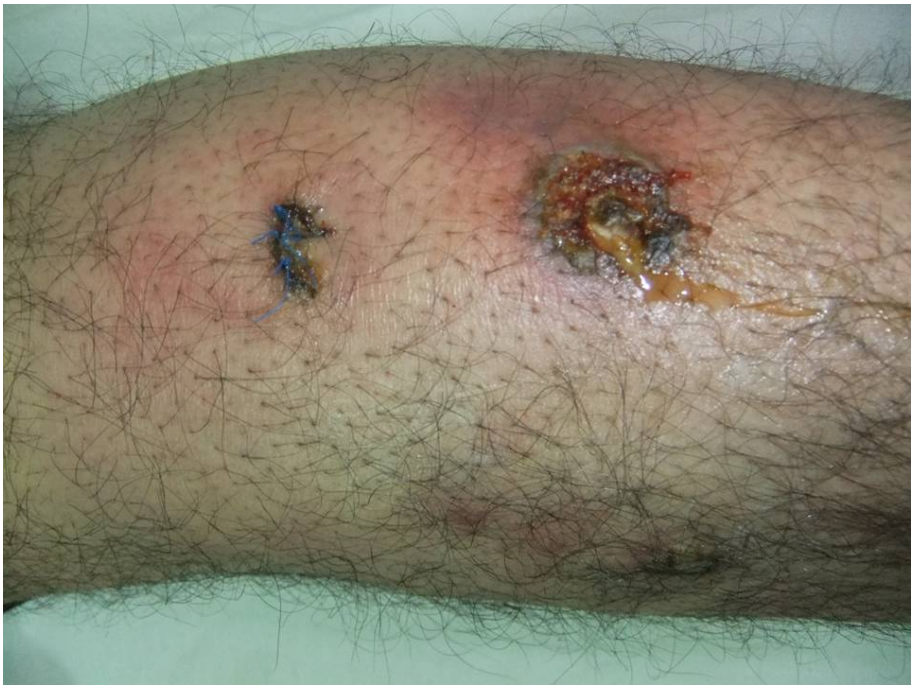

Figure 1: Ulcerated subcutaneous nodule over leg

A provisional diagnosis of neutrophilic lobular panniculitis was made.The underlying causes for the neutrophilic panniculitis were sought. Laboratory investigations showed elevated absolute leukocyte count: $12.5\times10^3/\mu l$, C-reactive protein (CRP): 24mg/l and erythrocyte sedimentation rate (ESR): 60mm/hr. While liver function, kidney function and amylase were within normal level. Anti-streptolysin O (ASO) titer, antinuclear antibodies (ANA), Carcinoembryonic antigen (CEA) and Ca 19-9 were negative. Antitrypsin: 213mg/dl(90-200),prostate specific antigen (PSA): 2.14(within normal limit), purified protein derivative (PPD)skin test: negative, chest X-ray (CXR) and leg X-ray were normal. Skin swab and tissue cultures for bacteria and mycobacteria were negative.

After exclusion of other causes of neutrophilic panniculitis, the patient was labeled to have subcutaneous SS, as he fulfilled the criteria for the diagnosis of SS, and the neutrophilic infiltrate was predominantly limited to the subcutaneous tissue.

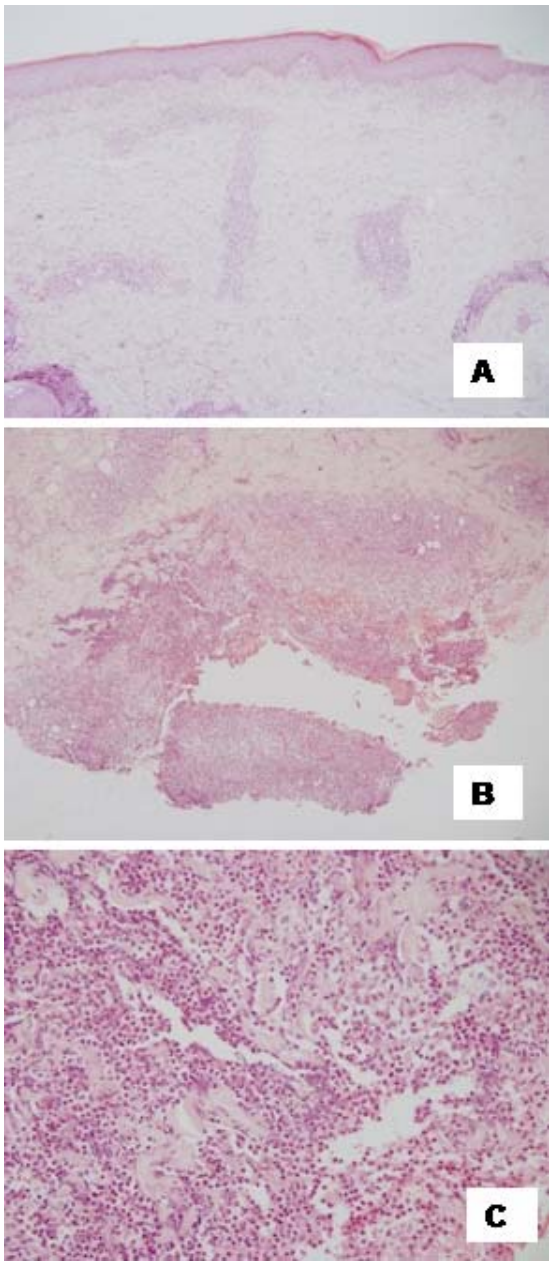

Figure 2: Hematoxylin and eosin stainof lesional skin. .

A. Spongiosis of the epidermis with perivascular mixed infiltrate in the dermis and subcutaneous tissue.

B. The main pathology located in the subcutaneous tissue

C. Dense neutrophils and nuclear dust in the subcutaneous infiltrate.

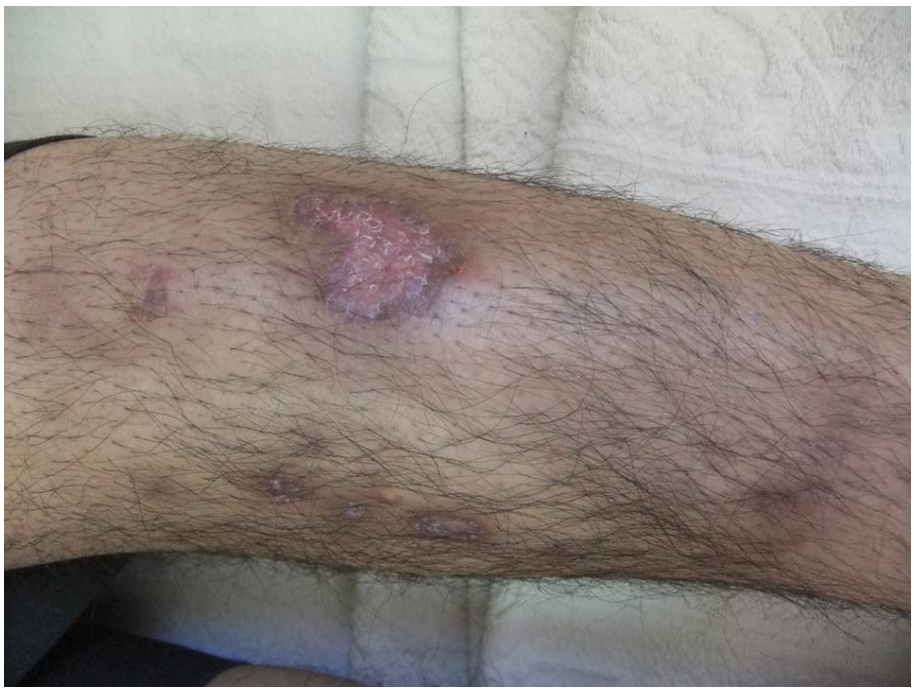

Figure 3: Atrophic scar and post inflammatory hyperpigmentation at site of previous lesions

The patient was given oral clarithromycin 500 mg twice daily for two weeks and prednisolone 40 mg per day. The skin ulcer and the indurated erythematous plaques improved after 5 weeks of treatment. The lesions recurred after six months and did not respond to prednisolone (40 mg/day) alone; so cyclosporine 300mg per day was added after 8 weeks of initiating prednisolone. The patient was maintained on this regimen for three months, before tapering was initiated over the following 7 months. The patient is currently off treatment and the lesions completely resolved (figure 3).

Discussion:

Sweet's syndromeis considered a multisystemic inflammatory condition. The diagnostic criteria for classical SS were originally proposed by Su and Liu in 1986[8], and modified by von den Driesch in 1994[10], as shown in table 1.Our patient fulfilled these criteria; as he presented with painful erythematous nodules that showed dense mature neutrophilic infiltrate in the subcutaneous tissue; increased ESR, CRP and leukocytosis and he responded to treatment with systemic steroids.

Skin lesions of SS are typically tender, purple-red, papules, plaques or nodules[6]. The eruption is often distributed asymmetrically and presents as either a single lesion or multiple lesions. The most common sites are the upper extremities, face, and neck[4]. The lesions may also be distributed in a sporotrichoid pattern[18], which was seen in our patient.

Since its initial description, SS has been reported to be associated with numerous conditions[4,9,10,12], as outlined in table 2.

Table 2. Sweet's syndrome and probably associated conditions[4-6]

1. Classical SS:	
A. Infections	Most commonly of the upper respiratory tract (streptococcosis) and the gastrointestinal tract (salmonellosis and yersiniosis), cholecystitis, pyelonephritis, otitis media
B. Inflammatory bowel disease	Crohn's disease and ulcerative colitis
C. Pregnancy	
2. Cancer – associated	Hematologic malignancies (most commonly acute myelogenousleukemia) and solid tumors (most commonly carcinomas of thegenitourinary organs, prostate, breast, and gastrointestinal tract)
3. Drug -induced	Granulocyte colony-stimulating factor (most common), apomoorphine, carbamazepine, diazepam, diclofenac, minocycline, hydralazine, nitrofurantoin, estradiol

Table 3. Classification of Neutrophilic dermatoses according to the location of Neutrophilic infiltrate

Location of Neutrophilic infiltrate	**The diagnosis**
Epidermis	Subcorneal pustulosis(Sneddon-Wilkinson disease)
Superficial dermis	Sweet's syndrome
Dermal vessels	Erythema elevatum diutinum
Eccrine glands	Neutrophilic eccrine hidradenitis
Dermis &subcutaneous tissue	Pyoderma gangrenosum
Subcutaneous tissue	Subcutaneous Sweet's syndrome
Subcutaneous tissue & Internal organs	Aseptic abscess

Females are affected 4 times more than males with an average age of 52.6 years[4, 10]. More than 75% of patients have other systemic symptoms apart from fever as arthritis, arthralgia or myalgia in two third of cases and conjunctivitis or episcleritis in one third of cases. Ocular manifestations may be the presenting feature of SS, as was seen in our patient, since scleritis was his first manifestation of SS. However, ocular lesions of SS are uncommon in the malignancy-associated and druginduced forms of the dermatosis[4].Oral lesions resembling aphthae occur in 2-3% of patient, but in 10% of patient with hematological malignancies[18,19]. Cough, dyspnea and pleuritis may represent pulmonary involvement. Rarely there may be cardiac, renal, hepatic, intestinal and neurological manifestations. Multifocal sterile osteomyelitis may also occur.[2]

The pathogenesis of SS is mostly related to abnormal regulation of the immune system, involving various inflammatory cells[4, 10, 20]. The rapid response of SS to systemic corticosteroids is in support of its immunological etiology[4,12,21]. A classification of neutrophilic dermatoses according to the main location of the neutrophilic infiltrate has been proposed[9], as summarized in table 3.

Systemic corticosteroids are the first line and best choice treatment for SS. Other first-line oral systemic medications are potassium iodide and colchicine. Second-line oral systemic treatments include cyclosporine, dapsone, indomethacin and clofazimine[4, 10, 12].

Cyclosporine was used effectively as a steroid-sparing agent in our patient.

Although SS is primarily a dermal process, the neutrophilic infiltrate may spread into the underlying subcutaneous tissue with an associated neutrophilic panniculitis[1, 4, 6, 7].

The term subcutaneous SS is recommended only for cases in which the neutrophilic infiltrate is exclusively or predominantly located in the subcutaneous tissue[1, 7-10]. In our patient, there was a neutrophilic lobular panniculitis. We performed many tests and imaging studies to look for a possible cause of his neutrophilic lobular panniculitis; these causes are shown in table 4. After exclusion of these causes, we applied the diagnostic criteria for the diagnosis of SS, and he fulfilled them. Therefore, our case fits well the diagnosis of subcutaneous SS.

Up to date only 21well documented cases of subcutaneous SS have been reported in the literature (table 5) [5,22-38]. Most of these cases are associated with myeloid disorders. In general, the associated diseases in patients who have subcutaneous SS are the same as those of classic SS[9], including both hematologic dyscrasias[39,40] and solid tumors[42].

In subcutaneous SS, lesions consist of multipleerythematous tender nodules or plaques [9, 11, 23,28,31]. Lower extremities are most commonly involved and rarely the lesions may become ulcerated[23,28,31]. Lesions usually appear and resolve in a short period of time[9]. When lesions resolve, they can leave hyper pigmentation and/or atrophicscars[25]. Skin lesions are usually accompanied by systemic symptoms like fever and malaise[24,26,31]. Subcutaneous SS in our patient was unusual in having ulceration and absence of constitutional symptoms, but was similar to other reported cases in term of rapid resolution, recurrence and healing with mild atrophic scar and post inflammatory hyper pigmentation.

Table 4. Differential diagnosis of the lobular subset of neutrophilic panniculitis: [7]

Differential diagnosis of lobular neutrophilic panniculitis
1. Infections
2. Pancreatitis
3. Alpha 1-antitrypsin deficiency syndrome
4. Rheumatoid arthritis
5. Subcutaneous Sweet's syndrome
6. Factitial

Table 5. Reported Cases of Subcutaneous Sweet's syndrome

Author/Yr	Age/ Sex	Associated medical illnesses	Clinical picture	Site of lesions	Constitutional symptoms	Course	Septal vs lobular infiltrate	WBC Count
Cooper[22] 1983	52/F	AML	Erythematous nodules	Arms+ Thighs	Fever	Resolution	lobular	16.900
Morioka[28] 1990	48/F	MDS	Nodules	Legs	Not available (N.A.)	Recurrent nodules over 4 yr controlled by prednisolone	Not available	(N.A.)
Cullity[29] 1991	65/F	MDS to AML	Plaques	Cheecks+ Supraclavicular fossa	Fever	Resolution	Not available	(N.A.)
Vignon-pennamen[30]/1991	60/F	Breast carcinoma	Nodules+ Plaques	Nodules on legs+plaques on arms +hands	Fever	Resolution in 4 wk	Septal	10.900
Matsumura[24] 1997	74/F	MDS	Nodules+ Plaques	Legs	Fever	Resolution	Both	13.800
Florez[23] 1999	60/M	Salmonella enteritidis infection	Nodules+ Plaques	Extensor sites of upper +lower legs	Subfebrile tempreture	Resolution	lobular	12.700
Carvalho[31] 2001	28/M	Crohn's disease	Nodules with spontaneous discharge of purulent material	Legs	Fever	Resolution	Both	17.500
Sutra-loubet 2004[25]	57/M	MDS	Nodules	Legs, arms + Trunk	None	Resolution (prior to prednisolne he was treated with colchicines and dapsone without good improvement)	Lobular	(N.A.)
Chen[26] 2004	59/M	MDS	Papules+pustuleses	Whole body	Fever+malaise	Resolution	Not available	10.700
Jagdeo [32] (2007)	(N.A.)	AML-M3 (APML) on ATRA	Single 1-cm red nodule	Shin		Spontaneous resolution	lobular	(N.A.)
Teng/2007 [33]	56/F	Metastatic Breast Cancer	Erythematous nodules	Neck, arms, lower limbs	malaise	Resolution with steroid in 1 wk	lobules	13.200
Uhara [34] (2008)	(N.A.)	AML-M6	Erythematous Indurations and plaques	Buttock,eye lid,ear,chest wall	fever	Resolution with steroids in 2 wk	lobular	(N.A.)
Becherer [35] (2009)	(N.A.)	MDS on G-CSF	Tender erythematous nodules	Whole body	none	Not specified	Septal	(N.A.)
Hood [36] (2010)	(N.A.)	MDS	erythematous, and indurated nodules	Submental area	fever	Unknown	Both	(N.A.)

Kim/2010 [37]	71/M	Spinal Metastasis from prostate cancer	Erythematous plaques And nodules	Trunk, arms	fever	Resolved with steroid s and colchicine	lobular	(N.A.)
Kim [38] (2012)	(N.A.)	MDS 2 cases;	Erythematous nodules	1.lower limbs 2.abdomen	1.fever 2.fever	Prompt resolution with steroid.	lobular	(N.A.)
Llamas/2013[5]	77/F	AML on pegylated G-CSF	Plaques,nodules	Neck,legs,upper limbs	fever	Resolution with steroid	lobular	(N.A.)
Chan[27]/2013 1.	53/M	AML	Tender nodules	Buttocks, thigh	Fever	Resolved spontaneously	lobular	8.500
2.	53/F	AML	Plaques and nodules	Upper and lower limbs	Fever	Resolved spontaneously	lobular	97.700
3.	41/M	AML/M2	plaques	Arms, thighs	Fever	Resolved spontaneously	Septal	1.100
4.	62/F	MDS	nodules	Lower limbs	unknown	Resolved with prednisone	Septal	2.300
Current case/2013	43/M	PBSNHL	Plaques and nodules	Upper and lower limbs	absent	Resolved over 7 months with steroids and cyclosporine	Lobular	12.500

Histopathologic description of subcutaneous SS is not mentioned in most reported cases[9]. Usually biopsies show a dense infiltrate of mature neutrophils in either the adipose tissue alone or it may be seen mainly in the subcutaneous fat with a less impressive infiltrate in the overlying dermis[7,27]. Within the subcutaneous fat, the neutrophilic infiltrate is most frequently present in the lobules[20,22,23,25,33,34,37,38], less commonly in the septae[20,27,30,35], or both[24,31,39,36,40,41]. Vasculitis was absent in all reported cases[9]. Occasionally, some mononuclear epithelioid cells may be found in the subcutaneous tissue[24], without necrosis of the adipocytes.

Progressive Bilateral Sensory Neural Hearing Loss was first described by McCabe in 1979[16]. The clinical picture is variable with rapidly fluctuating progressive loss of hearing in both ears over a period of weeks to months[16]. Nearly 15% of patients have other systemic "autoimmune" diseases such as multiple sclerosis, inflammatory bowel disease, systemic lupus erythematosus, rheumatoid arthritis, and ankylosing spondylitis[12, 43]. The hearing was restored in most patients after the administration of high doses of oral steroids[16]. The observed clinical response to corticosteroid and immunosuppressant therapy supports an immunological etiology targeting the inner ear[43].Some patients with PBSNHL fail to respond to any medical intervention and the gradual hearing loss progresses to complete deafness, which necessitates cochlear implantation[12,17]. Ourpatient also failed to respond to systemic treatment, so he underwent cochlear implantation as well.

Till now, there have been four case reports of classical SS following PBSNHL (table 6)[12-15]. Our case is the first, up to our knowledge, of PBSNHL that is followed by

subcutaneous SS. The PBSNHL did not improve with conservative measures and the patient underwent cochlear implant as seen in two other patients who had PBSNHL associated with SS[13,14].

Table 6. Reported cases of Sweet's syndrome associated with PBSNHL

Author/Yr	Sex/Age	Duration of sweet's before PBSNHL	Treatment of PBSNHL	Other associated medical illnesses
Pharis[12]/2000	F/48	2 Yr	Improved transiently with steroids, steroids sparing agents and plasmapheresis	Hysterectomy and bilateral oophrectomy, silicone filled breast implants
Wada[15]/2006	M/33	Not available	Not available	Not available
Cheng[13]/2010	F/63	10 Yr	Improved transiently with steroids, steroids sparing agents then after 12 months underwent cochlear implant	MDS
Aynehchi[14]/2012	F/49	2 Yr	Not improved with steroids then after 1 wk underwent cochlear implant	Not mentioned
Current case/2013	M/43	PBSNHL developed 3 Yrs before subcutaneous Sweet's syndrome	After 2.8 Yr, underwent cochlear implant	Medically free, apart from PBSNHL, SS.

Definitive classification of either SS or PBSNHL as an autoimmune disorder cannot be made with any degree of certainty at this time[43]. Whether SS and PBSNHL are related to a single common immunologic defect in our patient is something that we cannot assume. However, the triggering factor for PBSNHL may have resulted in an immunologic cascade that led to the development of subcutaneous SS.

The association of both SS and PBSNHL with more common systemic autoimmune diseases, such as inflammatory bowel disease and rheumatoid arthritis, suggests that the affected patients must be investigated and studied to elucidate a common immunologic role and to rule out other associated autoimmune diseases[12].

Conclusion:

The precise association between subcutaneous SS and PBSNHL has not been clearly elucidated. However, the association of both with each other supports the immunological etiology of both. The affected patients must be thoroughly investigated and studied to elucidate a common immunologic role and to rule out other associated immune mediated diseases.

References

1. Sweet RD. An acute febrile neutrophilic dermatosis. Br J Dermatol 1964; 74: 349-56 .
2. Barnhill R., Busam K. Vascular Diseases. In: Elder D, Elenitsas R, Jaworsky C, Johnson Jr. B, editors. Lever's histopathology of the skin. 8th ed. Lippincott-Raven: Philadelphia; 1997. p. 204-6.
3. Barham K., Jorizzo J., Cox NH. Rook's textbook of dermatology. 7th ed. Oxford: Wiley-Blackwell; 2004.Vasculitis and Neutrophilic Vascular Reactions; p. 49.33-49.36.
4. Cohen PR. Sweet's syndrome-a comprehensive review of an acute febrile neutrophilic dermatosis. Orphanet J Rare Dis.2007, 2: 34 doi: 10. 1186/1750-1172-2-34. This article is available from: http://www.OJRD.com/ content/2/1/34
5. Llamas-Velasco M, Garcia-Martin P, Sanchez-Perez J, Garcia-Diez A. Sweet's syndrome with subcutaneous involvement associated with pegfilgrastim treatment: first reported case. Journal of Cutaneous Pathology, 40: 46-49, January 2013.
6. Cohen PR, Kurzrock R: Sweet's syndrome revisited: a review of disease concepts. Int J Dermatol 2003, 42: 761-778.[CR]
7. Cohen PR. Subcutaneous Sweet's syndrome. A variant ofacute febrile neutrophilic dermatosis that is included in the his topathologic differential diagnosis of neutrophilic panniculitis. J Am Acad Dermatol 2005; 52: 927-8.
8. Su WPD, Liu HNH. Diagnostic criteria for Sweet's syndrome.Cutis. 1986; 37: 167-74.
9. Guillermo guhl. Subcutaneous sweet syndrome. Dermatol Clin 26 (2008) 541-551
10. Von den Driesch P. sweet's syndrome (acute febrile neutrophilic dermatosis). J Am Acad Dermatol 1994; 31:535-56.
11. Moschella S., Davis M. Neutrophilic Dermatoses. In: Bolognia J., Jorizzo J., Rapini R., editors. Dermatology. 2nd ed. MOSBY ELSEVIER; 2008. 379-383.
12. Pharis DB, Cerenko D, Caughman SW. Sweet's syndrome in a patient with idiopathic progressive bilateral sensorineural hearing loss. J Am Acad Dermatol. 2000 May; 42 (5 Pt 2): 932-5.
13. Cheng S, da Cruz M.Sweet's disease and profound, bilateral, sensorineural hearing loss. J Laryngol Otol. 2010 Jan; 124 (1): 105-7. Epub 2009 Oct 20.
14. Aynehchi BB & Hanson MB .Challenges in Cochlear Implantation for Sudden Idiopathic Sensorineural Hearing Loss Associated with Neutrophilic Dermatosis (Sweet's Syndrome). 2012 Meeting of The American Laryngological, Rhinological and Otological Society (The Triological society), January 26-28, 2012 Florida, USA.
15. Wada T, Sato H, Izumi S, Kubota Y, Ueki Y, Fujisaki T et al. Neuro-Sweet's disease in a patient with bilateral recurrent acute sensorineural hearing loss. Audiol Jpn 2006; 49: 254-9.
16. McCabe BF. Autoimmune sensorineural hearing loss. Ann Otol Rhinol Laryngol 1979; 88:585-9.
17. Hsieh D., Young Y. Bilateral Rapidly Progressive Hearing Loss Followed by Loss of Vestibular Function — Case Report Tzu Chi Med J 2006; 18: 141-144.
18. Cohen PR, Kurzrock R: Sweet's syndrome: a neutrophilic dermatosis classically associated with acute onset and fever. Clin Dermatol 2000, 18: 265-282.[CR]
19. Cohen PR, Talpaz M, Kurzrock R: Malignancy-associated Sweet's syndrome: Review of the world literature. J Clin Oncol 1988, 6:1887-1897.[CR]
20. Jordan HF: Acute febrile neutrophilic dermatosis. A histopathological study of 37 patients and a review of the literature. Am J Dermatopathol 1989, 11:99-111.[CR]
21. Going JJ, Going SM, Myskow MW, Beveridge GW: Sweet's syndrome: histological and immunohistochemical study of 15 cases. J Clin Pathol 1987, 40:175-179.[CR]
22. Cooper PH, Frierson HF, Greer KE. Subcutaneous neutrophilic infiltrate in acute

febrile neutrophilic Dermatosis. Arch Dermatol 1983; 119:610-1.
23. Florez A, Sanchez-aguilar D, Roson E, et al. Sweet's syndrome associated with salmonella enteritidis infection. Clin Exp Dermatol 1999; 24: 239-40.
24. Matsumura Y, Tanabe H, Wada Y, at al. Neutrophilic panniculitis associated with myelodysplastic syndromes. Br J Dermatol 1997;136:142-4.
25. Soutra Loubet C, Carlotti A, Guillemette J, et al. Neutrophilic lobular Dermatosis. J Am Acad Dermatol 2004;50:280-5.
26. Chen HC, Kao WY, Chang DM, et al. Neutrophilic panniculitis with myelodysplastic syndromes presenting as pustulosis: case report and review of the literature. Am J Hematol 2004; 76: 61-5.
27. Chan M., Duncan L., Nazarian R. Subcutaneous Sweet syndrome in the setting of myeloid disorders: A case series and review of the literature. J Am Acad Dermatol 2013; 68:1006-15.http://dx.doi.org/10.1016/j.jaad. 2012.12.954.
28. Morioka N, Otsuka F, Nogita T, et al. Neutrophilic dermatosis with myelodysplastic syndrome: nuclear segmentation anomalies of neutrophils in the skin lesion and in peripheral blood. J Am Acad Dermatol 1990; 23:247-9.
29. Cullity J, Maguire B, Gebauer K. Sweet's panniculitis. Australas J Dermatol 1991; 32:61-4.
30. Vignon Pennamen MD, Wallach D. Cutaneous manifestations of neutrophilic disease. A study of seven cases. Dermatologica 1991; 183:255-64.
31. Carvalho P, Cordel N, Courville PH, et al. Cutaneous aseptic abscesses, manifestations of neutrophilic diseases. Ann Dermatol Venereol 2001; 128:641-3.
32. Jagdeo J, Campbell R, Long T, Muglia J, Telang G, Robinson-Bostom L. Sweet's syndrome-like neutrophilic lobular panniculitis associated with all-trans-retinoic acid chemotherapy in a patient with acute promyelocytic leukemia. J Am Acad Dermatol 2007; 56:690-3.[CR]
33. Teng J. Draper B, Boyd A, Sweet's panniculitis associated with metastatic breast cancer.Journal of the American Academy of Dermatology. 56 (2) Supplement: S61-S62, February 2007.
34. Uhara H, Saida T, Nakazawa H, Ito T. Neutrophilic dermatoses with acute myeloid leukemia associated with an increase of serum colony-stimulating factor. J Am Acad Dermatol 2008; 59 (Suppl): S10-2.[CR]
35. Becherer K, Golda N, Feldman M, Diaz-Arias A, Caldwell C. Neutrophilic panniculitis associated with myelodysplastic syndrome with abnormal nuclear forms. J Cutan Pathol 2009; 36: 1024-6. [CR]
36. Hood M, Yu K, Magro C, Reisacher W. Pathology quiz case 2: subcutaneous Sweet syndrome of the neck. Arch Otolaryngol Head Neck Surg 2010; 136:1038, 1040-1.[CR]
37. Kim J, Choi S, Lee K. A Case of Sweet's Panniculitis Associated with Spinal Metastasis from Prostate Cancer. Ann Dermatol. 2010 November; 22 (4): 478-481.
38. Kim IH, Youn JH, Shin SH, Yahng SA, Lee SE, Kwon JC, et al. Neutrophilic panniculitis following azacitidine treatment formyelodysplastic syndromes. Leuk Res 2012; 36: e146-8.[CR]
39. Matsumura Y, Tanabe H, Wada Y, Ohta K, Okamoto H, Imamura S: Neutrophilic panniculitis associated with myelodysplastic syndromes. Br J Dermatol 1997, 136: 142-144.[CR]
40. Sutra-Loubet C, Carlotti A, Guillemette J, Wallach D: Panniculite neutrophilique (P151 at the Journees Dermatologiques de Paris, December 6–9, 2000). Ann Dermatol Venereol (Paris) 2000, 127: 4S176-4S177.[CR]
41. Hasegawa M, Sato S, Nakada M, Nitta H, Shirasaki H, Kasahara K, Takehara K: Sweet's syndrome associated with granulocyte colony-stimulating factor. Eur J Dermatol 1998, 8: 503-505.
42. Shinojima Y, Toma Y, Terui T: Sweet syndrome associated with intrahepatic cholangiocarcinoma producing granulocyte colony-stimulating factor. Br J Dermatol 2006, 154: 1103-1104 [CR].
43. Cao MY, Thonnard J, Deggouj N, Gersdorff M, Philippe M, Osselaer JC, et al. HLA class II associated genetic susceptibility in idiopathic progressive sensorineural hearing loss. Ann Otol Rhino1 Laryngol 1996; 105: 628-33.

متلازمة سويت تحت الجلدي المرتبطة بفقدان السمع الحسي العصبي التدريجي

هيثم قاسم أبو الهيجاء،[1] نضال عبيدات،[1] محمد الطوره،[1] عمر الشخيبي،[2] مأمون حتاملة،[1] بيان أبو الهيجاء[1]

1- قسم الجلدية، مركز الملك حسين الطبي، عمان، الأردن.
2- قسم الباثولوجيا، مركز الملك حسين الطبي، عمان، الأردن.

الملخص

متلازمة سويت هي حالة إلتهاب جلدي ذاتي المناعة تتميز بلويحات حماميه مخترقه من قبل كريات الدم البيضاء (العدلات) مقتصرة على الأدمة، إذا كان هناك ارتشاح ممتد إلى أنسجة تحت الجلد أو مقتصر عليها، عندها تعد نادرة جلديه جديدة يتم وصفها وهي متلازمة سويت تحت الجلدي. نظراً لندرة هذه الحالة فإن ارتباطها مع حالات مرضيه أخرى غير محسوم.

هنا في هذه الدراسة سيتم عرض حالة مريض يعاني من فقدان السمع الحسي العصبي التدريجي، تم إجراء زراعة قوقعه له وبعد شهرين من هذه العملية، ظهر عنده لويحات حمامية جلدية تتفق مع الوصف المرضي لمتلازمة سويت تحت الجلدي. وقد تم علاجها والسيطرة عليها بعلاجي الكورتيكوستيرويد والسيكلوسبورين.

بعد مراجعة المقالات الطبية المنشورة سابقا تم ذكر أربع حالات مرضية من متلازمة سويت تلاها ظهور فقدان السمع الحسي العصبي التدريجي، وواحد وعشرين حالة من متلازمة سويت تحت الجلدي مرتبطة بحالات مرضية مختلفة. و لكن هذه الحالة هي الأولى التي يعاني فيها المريض من فقدان السمع الحسي العصبي التدريجي وبعد ذلك ظهر عنده متلازمة سويت تحت الجلدي. لذلك اقتضى التنويه إليها ونشرها.

الكلمات الدالة: متلازمة سويت تحت الجلدي، فقدان السمع الحسّي العصبي.