# A Case of PUO

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# ABSTRACT

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This case report describes a 68 year old patient with fever since 2 months and generalised painless body eruptions since 1 month. Complete evaluation was done including skin biopsy of the nodular eruptions.

Sweet's syndrome or acute febrile Neutrophilic Dermatosis is a condition characterised by sudden onset of fever, leucocytosis and tender erythematous, well demarcated papules and plaques which show dense neutrophilic infiltrates on histologic examination.

Keywords: Fever, Generalized eruption

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A 68 year old Madhavan, a manual labourer presented with fever since 2 months and generalised body eruption since 1 month. Fever was high grade, intermittent, with occasional chills and rigor and more towards evening. No history of diarrhea, vomiting. No history of travel outside Kerala, No history of vomiting, diarrhea, blood in stools, icterus, dysuria, altered consciousness etc Had body pain and fatigue. Generalised painless, body eruptions were seen since 1 month, more over face, arms and legs. No itching, no loss of sensations, no itching, ulceration, discharge etc.

Past history of recurrent episodes of breathlessness was observed for the past one year. No history of similar rashes before. Not on regular treatment, No history of DM, Hypertension, TB, IHD, Epilepsy, Malignancies. Takes mixed diet, Chronic smoker for



Figure 1. Papules, nodules and plaques more common in the face, neck and arms.

past 40 years (2 packs a day). Stopped since 1 year, not alcoholic, appetite decreased, sleep decreased, bowel and bladder habits were normal. No history of similar complaints, DM, Hypertension, malignancies, asthma or TB in the family.

On examination he was moderately built and nourished, Pallor +, Clubbing +; Grade II

No icterus, cyanosis, lymphadenopathy, BP : 110/70 mm Hg, Pulse : 78/min

Peripheral pulses feeble in both legs, Oral hygiene poor, Tobacco stains present in mouth

A single Lipoma present over the back.

Skin lesions- Extensive nodular erythematous eruptions present all over the body, more over the face, upper arms and legs, less over the trunk, normal vesicular breath sounds heard, air entry equal on both sides and bilateral rhonchi and crepitations were present in all areas, Liver and spleen not palpable, No lymphadenopathy. GIT, CVS, CNS were normal.

#### Lab investigations

On 24/9/2009 - Hb: **9.6** g%, ESR: **80** mm/hr, TC: **31800**, N 90 L8 E2, Platelet count: 3.32 lakhs, RBS: 105 mg%, B.Urea: 20 mg%, S. Creatinine: 0.6, Na+: 122 meq/L and 139 (29/9/09), K+: 4.1,3.9meq/L, CPK: 17U, Serum bilirubin, Total: 1.9mg%, Direct: 0.6, Protein Total: 7.4 gm%, Albumin: 3.3, AST: 56 U/LALT: 35U/L, Alkaline phosphatase: 480U/L, PSA (6/10/09): 0.5 ng/ml, TFT- Normal, RA factor Negative, Stool Occult Blood – Negative.

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Figure 2. Site of lesion in the arm taken for biopsy

**Peripheral Smear** - RBC: red cells are mainly normocytic normochromic, No normoblasts or parasites seen, WBC : count is increased with neutrophilia, N85 L6 M8 E1, Polymorphs and lymphocytes show normal morphology, No immature cells or blasts seen, Platelet seen scattered and count is adequate, Impression: Moderate polymorphonuclear leucocytosis, Normocytic normochromic Anaemia.

**Bone Marrow Smear -** Dated 6/10/2009, Cellularity increased. Myleoid : Erythroid ratio increased. Erythroid series show normal maturation. Myleoid series are increased in number, with normal maturation. Ratio of plasma cells and lymphocytes normal

No abnormal cells seen. Megakaryocytes series show normal maturation

Diagnosis: BM smear show decrease in erythroid series and increase in myeloid series

**Skin Biopsy-** Site: Right upper arm, Skin shows normal epidermis and dermis showing a dense perivascular neutrophilic infiltration, also with some histiocytes and lymphocytes

Small blood vessels show swelling ofendothelium and oedema of upper dermis, Occasional vessels show vasculitis with necrosis of vessel wall, PAS and Gram stain : No specific pathogen identified. Diagnosis: consistent with Acute Febrile Neutrophilic Dermatosis (SWEET SYNDROME).

Dated 11/10/2009 - Hb: 9.6 g%, ESR : 80 mm/hr, TC : 18200, N82 L13 E5

Diagnosis: Acute Febrile Neutrophilic Dermatosis (Sweet Syndrome)

Course in the Hospital - His symptoms improved with

a course of antibiotics and antipyretic agents. Fever subsided. He was put on a tapering dose of steroids. Dermatologist and ophthalmologist were consulted.

# DISCUSSION

Sweet's syndrome or acute febrile Neutrophilic Dermatosis is a condition characterised by sudden onset of fever, leucocytosis and tender erythematous, well demarcated papules and plaques which show dense neutrophilic infiltrates on histologic examination. It is named after Robert Douglas Sweet. It was originally described in women with elevated WBC count. Ironically this disease now is usually seen in neutropenic patients with cancer more often with acute leukemia. It presents as red or bluish red papules or nodules that may coalesce and form sharply bordered plaques. The edema may suggest vesicles, but on palpation the lesions are solid, and the vesicles probably never arise in this disease. The lesions are most common in face, neck and arms. On the legs, they may be confused with erythema nodosum. Although it may occur alone, it is often associated with hematologic diseases like leukemia and immunologic diseases like RA or IBD

Genetic association has been suggested, but not yet been proved. Approximately 20 % of cases are associated with malignancy predominantly hematological especially acute myeloid leukemia. 50% of cases have an underlying condition like streptococcal infection, IBD, other hematologic malignancies, solid tumours, pregnancy. Attacks of SS may precede the hematologic diagnosis by 3 months to 6 years. So a close evaluation of patients in idiopathic group is required. Acute tender erythematous plaques, nodes, pseudovesicles and occasionally blisters with an annular or acriform pattern occur on head neck legs and arms. Trunk is rarely involved. It is associated with fever and elevated ESR. Both the lesions and temperature respond to glucocorticoid administration. Arthralgia, Eye involvement (conjuctivitis or irido-cyclitis), Oral aphthae may be presenting symptoms.

Laboratory Investigations - Moderate neutrophilia, Elevated ESR (>30mm/hr), Increase in alkaline phosphatase, Skin biopsy shows a papillary and mid dermal mixed infiltrate of polymorphonuclear leucocytes with nuclear fragmentation and histiocytic cells. The infiltrate is predominantly perivascular with endothelial cell swelling in some vessels, Vasculitic changes are absent in early lesions, Vasculitis occur secondary to noxious products released from neutrophils. Treatment- Systemic corticosteroids - Prednisolone 0.5 to 1.5 mg/kg body wt produce rapid improvement and are the gold standard of treatment, Frequent relapses can occur, Steroids are then tapered within 2 to 6 weeks to zero. Resolution is occasionally followed by milia and scarring. Other Treatment Options are Topical or intralesional corticosteroids, Oral potassium iodide, Colchicine, Indomethacin. Alternatives to corticosteroid treatment include dapsone, Doxycycline, clofazimine and cyclosporine.

# **END NOTE**

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#### Conflict of Interest: None declared

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