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RESEARCH ARTICLE

SWEET SYNDROME : A RARE CASE.

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Abstract

11 yrs female, 2nd issue of non-consanguineous marriage with H/O Joint Pain & swelling since 8 months, fever since 4 days was admitted with us.
Patient was investigated and treated symptomatically. Peripheral smear showed dense neutrophilia. Skin biopsy confirmed the final diagnosis of 'Sweet Syndrome'. Symptoms relieved dramatically after MethylPrednisolone Pulse Therapy

Introduction:-

Sweet syndrome or **acute febrile neutrophilic dermatosis** is a skin disease characterized by

- sudden onset of fever
- an elevated white blood cell count and
- tender, red, well-demarcated papules and plaques that show
- dense infiltrates by neutrophil granulocytes on histologic examination

Case Presentation:-

- 11 yrs female , 2nd issue of non-consangnious marriage, admitted with chief complaints of
- Joint Pain & swelling since 8 months, fever since 2 days

Examination:-

- Patient was febrile, well nourished

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- Pulse- 110/ min , RR- 30/min
- BP – 100/60 mm Hg (50th Percentile)
- Pallor

Maculopapular rash on dorsum of joints:-
- ***GOTTON'S Papule*** **over knuckles**
- Swelling at elbow joint
- Restricted joint movement
- No mucous membrane involvement
- S/E : Normal

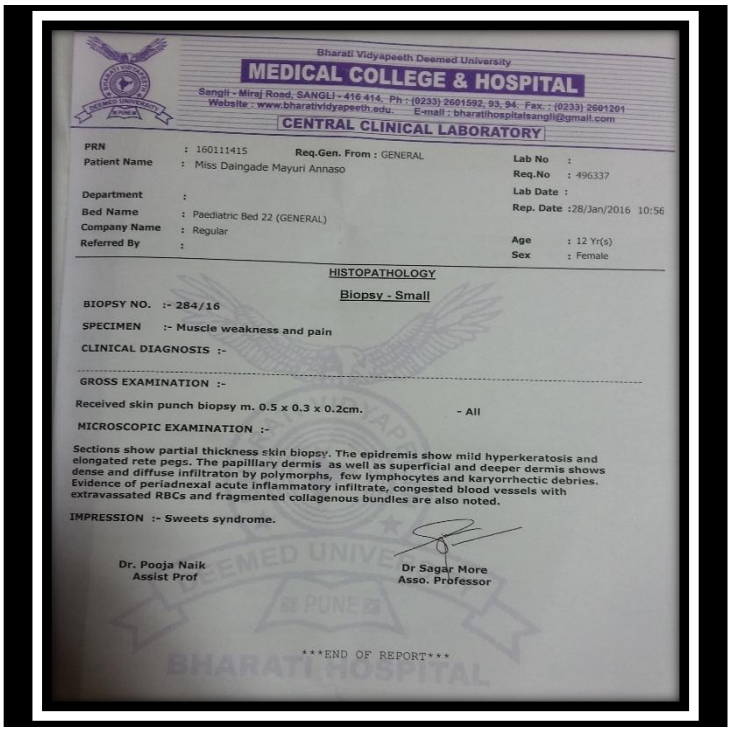
Provisional Diagnosis:-
Fever with Polyarticular Arthritis with Dermatomyositis
to r/o Mixed Connective Tissue Disorder

Differential Diagnosis:-
- Rheumatoid Arthritis
- Pyoderma Gangrenosum
- Erythema Multiforme
- Adverse drug reactions
- Urticaria
- Urticaria Vasculitis

Investigations:-
- **CBC:**
- Hb -9.2 gm %
- WBCs-13200 / mm^3
- P-84 %
- L-12 %
- Platelets-494000
- **ESR : 20 mm at the end of 1st hour**
- **Urine Routine** : Pus cells- 8-10 / mm^3
- CRP – Negative
- ANA- Negative
- ASO – Negative

- RA factor – Negative
- CPK – 86 IU/L
- LFTs – WNL

Skin Biopsy:-



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PRN : 160111415 Req.Gen. From : GENERAL
Patient Name : Miss Daingade Mayuri Annaso
Department :
Bed Name : Paediatric Bed 22 (GENERAL)
Company Name : Regular
Referred By :

Lab No :
Req.No : 496337
Lab Date :
Rep. Date :28/Jan/2016 10:56
Age : 12 Yr(s)
Sex : Female

HISTOPATHOLOGY

Biopsy - Small

BIOPSY NO. :- 284/16

SPECIMEN :- Muscle weakness and pain

CLINICAL DIAGNOSIS :-

GROSS EXAMINATION :-

Received skin punch biopsy m. 0.5 x 0.3 x 0.2cm. - All

MICROSCOPIC EXAMINATION :-

Sections show partial thickness skin biopsy. The epidermis show mild hyperkeratosis and elongated rete pegs. The papilllary dermis as well as superficial and deeper dermis shows dense and diffuse infiltraton by polymorphs, few lymphocytes and karyorrhectic debries. Evidence of periadnexal acute inflammatory infiltrate, congested blood vessels with extravassated RBCs and fragmented collagenous bundles are also noted.

IMPRESSION :- Sweets syndrome.

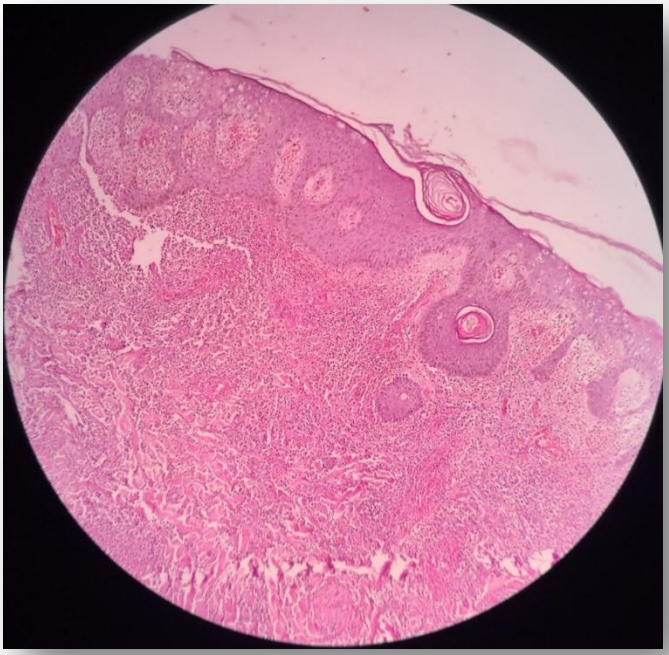
Dr. Pooja Naik
Assist Prof

Dr Sagar More
Asso. Professor

END OF REPORT

Histopathology:-

Histopathology showed a diffuse infiltrate consisting predominantly of mature neutrophils located in the upper dermis



Final Diagnosis
Sweet syndrome or
Acute Febrile Neutrophilic Dermatosis

Treatment:-
- Treatment was commenced with Methylprednisolone Pulse Therapy
- Ibuprofen and Paracetamol was given initially for pain relief
- Patient was discharged on oral steroids which were given over a period of 6 weeks and then tapered off.

Outcome:-
- Treated as per management protocol of Sweet Syndrome
- Patient responded to therapy
- Patient is on regular follow up for last 10 months and is symptom free

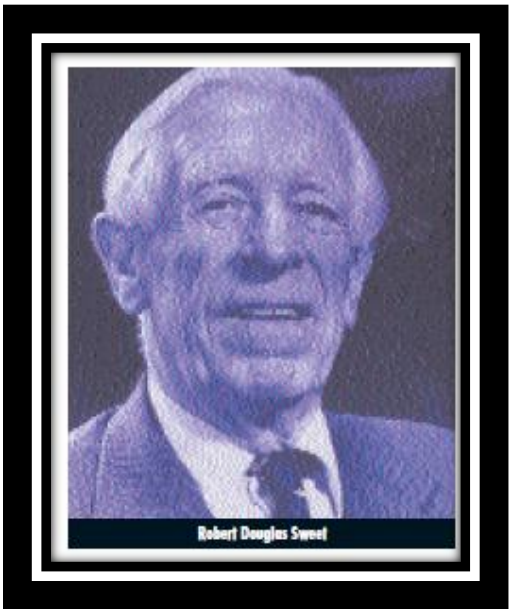
Conclusion:-
- Sweet Syndrome is an uncommon skin disorder in children characterized by fever and appearance of painful violaceous erythematous skin lesions.
- The clinical diagnosis done by diagnostic criteria (2 major , 2 minor) and confirmation on skin biopsy.
- The gold standard treatment is Corticosteroids.

What is Sweet's Syndrome?
- Sweet's syndrome (also known as acute febrile neutrophilic dermatosis) is a rare skin disorder characterized by a fever and the appearance of tender solid red lumps on the skin.
- It is a reactive condition with a number of potential triggers & is not a hereditary condition.
- It is not contagious and is not skin cancer.
- In approximately half of those affected it is not possible to identify a cause
- It is much more common in females between 20-40 years old a result of reaction to infection such as upper respiratory tract infections or to different medicinal drugs.
- In some patients it can be a manifestation of systemic diseases such lupus erythematosus or rheumatoid arthritis.
- In a few cases, it is a result of hormonal changes
- In some cases it may be a sign of an underlying blood disorder or an internal cancer.

Diagnostic criteria (Both major criteria and 2 of the 4 minor criteria are required to establish the diagnosis)
Major criteria:-
- Abrupt onset of painful erythematous plaques or nodules
- Histological evidence of dense neutrophilic infiltrate without evidence of primary leukocytoclastic vasculitis

Minor criteria:-
Pyrexia (>38°C)
- Association with an underlying hematological or visceral malignancy, inflammatory disease, or pregnancy or preceded by an upper respiratory or gastrointestinal infection or vaccination
- Excellent response to systemic corticosteroids, potassium iodide or colchicines Abnormal laboratory values (3 of the following 4):
- Erythrocyte sedimentation rate greater than 20 mm/h,
- positive C-reactive protein, >8000 leucocytes, and >70% neutrophils

Historical aspect:-
Dr Robert Douglas Sweet 1917 – 2001,
English Dermatologist
He collected a group of patients with skin problems which he later described as *Acute Febrile Neutrophilic Dermatosis*
This began life in his notes as *Gomm-Button Disease*,
names derived from his first two patients with this disease
His findings were presented to the Dermatological world in two papers published in the *British Journal of Dermatology* in 1964 and 1968



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