PROLONGED FEVER : DIAGNOSTIC DILEMMA

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GUIDE:

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HISTORY:

16 Years old, girl 4th born to non- consanguineous parents presented with

- Fever-high grade,intermittent,no chills and rigors x 1 month
- Vomiting - non bilious,non projectile - on & off 1 month
- Cough – expectoration ,mucoid consistency
- Skin rashes , hyperpigmentation
- Weight loss +

- No h/o breathlessness ,loose stools, dysuria,joint pain&swelling,night sweats.
- No previous treatment history.

PAST HISTORY:

- Primary complex at 2 years of age treated.
- At 3 years child had skin lesion with progressive subcutaneous thinning and scarring involving R UL &LL – Skin grafting was done
- Immunised up to age.

O/E:

- Temp : 100f
- PR-100/min,RR-20/min,BP-100/60.
- Pallor +, No palpable cervical lymph nodes.
- Wt: 45 Kg, ht: 146, BMI :20
- CVS – S1,S2+,No murmur
- RS-B/L AE+
- P/A :soft,Spleen tip palpable
- CNS: No focal neurological deficit.

L/E OF SKIN :

PAPULAR RASHES PRESENT OVER UL AND FACE

SKIN CHANGES :

- Atrophic skin changes,subcutaneous wasting,contractures of R fingers,previous grafting +

DD :

- TB
- EBV
- BRUCELLOSIS
- CONNECTIVE TISSUE DISORDER
- MALIGNANCY

INVESTIGATIONS:

- CBC – Anemia,leukopenia

Hb-10

Platelets-1.50

TC-1340

- LFT,RFT,Urine R/e – normal
- Urine c/s – E.coli
- USG Abdomen- Splenomegaly,multiple abdominal lymphnodes.

INFECTIONS:

- QBC-Negative
- Blood c/s ,Rpt urine c/s –no growth
- **ESR – ELEVATED. CRP-NORMAL**
- Mantoux,AFB – Negative,
- Cxr - Normal
- HIV – Negative
- **LEPTOSPIROSIS IGM , EBV – POSITIVE**
- Immunogloubin levels- Normal

AUTOIMMUNE DISEASE ??

- ANA- Negative
- RA Factor – Negative
- CPK-Normal
- C3,C4 -Normal

DERMATOLOGY :

- In view of skin rashes dermatology opinion was sought and treated as Erythema multiforme with T.Acyclovir x 5days.
- Nerve conduction test - Normal
- Skin rashes progressively increased hence biopsy was done

Histopathology report-

- Epidermis with mild hyperkeratosis
- Increased collagenisation

Features Suggestive of Morphea

MALIGNANCY???

- PS- Normocytic,normochromic,no blast cells
- **LDH- ELEVATED**
- Uric Acid- Normal
- Bone marrow Aspiration-Normal
- Bone Marrow C/s : No growth

- ECHO- Normal

- CT Abdomen – Showed Multiple Lymphnodes with few nodes showing **central hypodensity.**
- CT thorax- Normal

- Planned for laproscopic LN biopsy.
- Reexamination –peripheral nodes: small axillary nodes
- Hence axillary lymph node biopsy was done

EVALUATION OF LYMPH NODE BIOPSY :

- HPE of axillary LN showed reactive changes with foci of necrosis and scattered nuclear debris and apoptotic bodies
- Features suggestive of NECROTISING LYMPHADENITIS
- Immunohistochemical : CD3,20,68 positive

KIKUCHI'S DISEASE

- Child was started on Oral steriod –fever spikes settled, Symptomatically improved.
- Child is on regular follow up

DISCUSSION

KIKUCHI'S DISEASE :

- Kikuchi and Fujimoto first reported in Japan in 1972. Though it has been reported worldwide, is still confused with malignant lymphoma, SLE
- Self-limiting benign systemic lymphadenitis, especially involving the cervical nodes of unknown cause.

ETIOLOGY :

Unknown, EBV,HHV,HSV,Bacteria,Protozoa,Neoplastic conditions, autoimmune disorders

Ref: Kikuchi's disease;Malathi sathiyasekaran,So.sivabalan;Indian Pediatrics;vol 41(192-194);feb 17,2004

CLINICAL FEATURES :

- Female predominance (M:F 1:4)
  - Painless lymphadenopathy cervical region.-80 %,
    Single location – 83 %.

EXTRANODAL FINDINGS

- Incidence of skin involvement - 5 to 30%.
- Maculopapular lesions, morbilliform rash, nodules, urticaria and malar rash, which may resemble that of SLE.

DD:SLE/Lymphomas/TB/Infectiousmononucleosis

Ref :Kikuchi – Fujimoto disease;Xavier Bosch &Antonio;orphanet . Rare disease.2006,1:18

LAB INVESTIGATIONS :

- No specific serological markers.

IMAGING STUDIES :

- CT scan of the affected lymphnode shows hypodense centers with peripheral ring enhancement corresponding to the central necrosis.

Ref: Kikuchi's disease;Malathi sathiyasekaran,So.sivabalan;Indian Pediatrics;vol 41(192-194);feb 17,2004

BIOPSY OF THE LYMPHNODE IS DIAGNOSTIC.

The pathological features :

- Patchy or confluent area of necrosis,
- Varying amount of nuclear debris in affected area
- Aggregates of histiocytes, presence of medium - large sized transformed lymphocytes (immunoblasts) and plasmacytoid T cells
- **Absence of neutrophils and eosinophils.**

Ref: Kikuchi's disease;Malathi sathiyasekaran,So.sivabalan;Indian Pediatrics;vol 41(192-194);feb 17,2004

TREATMENT :

- Self-limited within one to four months.
- Recurrence rate of 3 to 4% has been reported
- Symptomatic treatment
- Rapid resolution with steroids.
- Regular follow- to rule out the development of SLE.

INTERESTING POINTS IN THIS CASE :

- Prolonged fever (1 month) with red herrings : UTI, EBV, Lepto,
- Axillary nodes and abdominal nodes involvement without cervical nodes
- Kikuchi disease with EBV Positive
- Associated Morphea

TAKE HOME MESSAGES :

- Kikuchi's disease should be considered as differential diagnosis in young patients with lymphadenopathy and fever of unknown origin
- **DAILY CLINICAL EXAMINATION IS MANDATORY** in chronic case- as some findings may be absent/missed initially which can be picked up in later days