QUIZ

Case -1

A 32 years old female presented with painful erythematous swelling of the interphalangeal joints of both hands of 4 years duration. She was also noticed to have brownish-yellowish firm papules and plaques on the extensor surfaces of hands and forearms with involvement of the face, scalp and lower limbs, both ears has multiple firm painful nodules. The skin lesions were

mildly puritic. She was also found to have nail dystrophy. The patient was also admitted twice last year for pericardial effusion and shortness of breathing. Laboratory test shows normal Rheumatoid factor and normal ESR, abnormality of serum lipid. Histopathology of skin nodule shows heavy infiltration by mononuclear cells predominantly histocytes, lymphocytes and eosinophils with few multinucleated giant cells, the cells were PAS positive and contain Lipids.

What is your diagnosis?

Answer:

Multicentric Reticulohistocytosis :

This is a rare histocytic proliferative disease in which joints, skin and mucous membranes are affected. The arthropathy usually precedes nodular skin involvement and mucosal infiltration. The pathogenesis is unknown. It is a reactive histocytosis to unknown Antigens, tuberculosis has been implicated in some cases.

The disease usually affects women with male to female ratio of 1:3. It is a disease of middle age. 60% presents with polyarthritis of hands or other joints which is usually mutilating polyarthritis. Skin lesions are firm yellowish-brown papules involving the hands, face, scalp with characteristic coral beaded like lesions on both ears and around nail folds resulting in nail dystrophy.

50% has mucosal involvement of the mouth and sclera.

30% has abnormality of serum lipids.

Involvement of bone marrow, lymphnodes, hearts and pericardium, lungs, bone, liver and GIT may occur.

Around 20% of patient have been found to have an associated internal malignancy commonest are gastric, ovarian, breasts, uterine carcinomas, myeloma and myelodysplasia.

The prognosis is good if there is no associated malignancy. Fatal cardiac involvement may occur with widespread systemic involvement, morbidity is due to a crippling arthropathy and scarred skin. Treatment with systemic steroids combined with Azathioprine give good result as well as the use of cyclophosphamide.

Case -2

A 7 years old boy presented with Fever and Symptoms of upper respiratory tract infection, associated with maculopapular rash with no skin infiltration. On examination the child was found to have hepatosplenomegaly with moderate lymphnode enlargement. Laboratory tests show anemia, thrombocytopenia, raised liver enzymes and hyperbilirubin, elevation of triglyceride and very low-density lipoproteins and hypofibrinogenaemia . Histopathology of liver biopsy shows a diffuse infiltration with lymphocytes and mature histocytes, which exhibit active phagocytosis of erythrocytes and leukocytes.

What is you diagnosis?

Answer:

Familial haemophagocytic lymphohistocytosis

This is a rare reactive histocytosis in which there is widespread infiltration of multiple organs with lymphocytes and mature histocytes showing prominent phagocytosis.

The incidence is 1.2 per million with more preponderance in boys. Around 75% of cases are familial with an autosomal recessive inheritance.

Fever is usually the first sign with a transient nonspecific maculopapular rash at time of high fever, patient also has hepatospleenomegaly with lymphadenopathy.

Laboratory tests show anemia, thrombocytopenia, raised liver enzymes and elevation of blood lipids. They also have hypercytokinaemia with elevated circulating IFN-8, TNF-2, IL-6 during disease activity. Soluble CD8 also elevated suggesting a role of cytotoxic cell in the pathogenesis.

The prognosis is poor, 96% die within 12 months.

Initial treatment is with splenectomy, exchange transfusion and chemotherapy including vinblastine and intrathecal methotrexate or etoposide.

Bone marrow transplantation regarded now as the most effective treatment.

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NOTES FOR CONTRIBUTORS

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