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RESEARCH ARTICLE

LUPUS PANNICULITIS IN A 12 YEAR OLD BOY: A CASE REPORT.

Romita Bachaspatimayum, Y. Imlongchaba, Anup Goswami and Angila Dorjee.

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Abstract

Introduction: Lupus panniculitis, also known as Lupus profundus or subcutaneous Lupus erythematosus (LE), was first described by Kaposi in the year 1869. It is a small subset of all cutaneous LE which represents 2-3% of this group. It usually occurs in 3rd – 4th decade but children can also develop lupus panniculitis. There is female preponderance with female: male ratio ranging from 2:1 to 4:1.[1]

Case report: A 12 year old boy presented with multiple painful , mildly erythematous to bluish coloured nodules over the face and upper arms, sparing the distal upper limbs for last six months. Cutaneous examination revealed multiple, mildly-tender, indurated erythematous to bluish coloured nodules over face & upper arms. ANA- 2.5 & anti-ds DNA- 200 IU/ml. Histopathological examination revealed mild pericapillary lymphocytic infiltration in dermis with deep subcutaneous fat showing moderatelymphohistiocytic cell infiltration with focal areas of necrosis and occasional ill-formed granulomas which was consistent with granulomatous panniculitis. Direct immunofluorescence (DIF) showed- deposits of IgG, IgM, IgA and C3 along the dermo-epidermal junction.

Introduction:-

Lupus panniculitis also known as lupus erythematous panniculitis (LEP) or lupus profundus was first clinically described by Kaposi in 1869.[1] Lupus panniculitis is characterized by tender subcutaneous nodules and plaques that may arise in crops. A history of trauma can sometimes be elicited. Lesions tend to develop on the face, upper arms, hips and trunk. The lack of involvement of the distal extremities is noteworthy.[2] LEP may develop in association with discoid lupus erythematosus (DLE) or systemic lupus erythematosus (SLE) or may occur as an isolated phenomenon.[3] It usually occurs in adults where median age of onset is 30-40 years, but can also occur in children. There is female preponderance (M:F = 2:1 to 4:1).[1] Characteristic microscopic finding include deep lymphocytic infiltration in the fat lobules and in the septa. Lymphoid aggregates, nodules & germinal centers also known as *follicular centers*, are common. The dermis can have just a superficial and deep perivascular lymphocytic infiltrate with plasma cells or can have all of the changes of lesions of discoid lupus erythematosus.[4] Here we present a classical case of lupus panniculitis in a young boy.

Case report: A 12 years old boy attended our out patient department (OPD) of Dermatology in January 2015 with multiple, painful swellings over face, neck and upper limbs since last 6 months. It first occurred on face, then subsequently similar lesions erupted on neck and upper arms. There was no history of any preceding injury, any

Corresponding Author:- Anup Goswami, Post Graduate Trainee
Address:- Department of Dermatology, Venereology & Leprology, Regional Institute of Medical Sciences, Imphal, Manipur, India.

drug intake or application of any medications. There was history of photosensitivity and atopy. There was no any history of fever, cough, dyspnea, myalgia, fatigue, gastro-intestinal symptoms, Raynaud's phenomenon or joint pain. On cutaneous examination multiple, around 20 in number, tender, well- defined, mildly erythematous to bluish nodules and plaques with size ranging from 0.5cm x 0.5cm to 1.5cm x 1.5cm were present over face (Figure-1), neck and upper arms (Figure-2).

Figure:1(a)

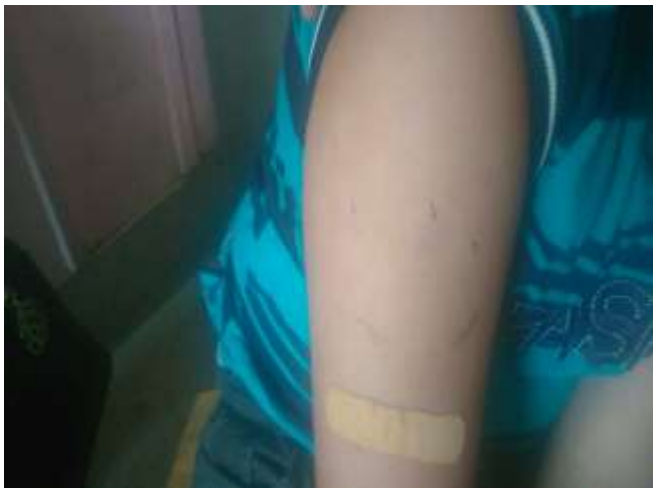

Figure:1(b)

Figure: 1(a) shows ill-defined erythematous to bluish-grey plaque over right side of cheek, figure: 1(b) shows ill defined bluish-grey nodule over extensor aspect of right arm.

Patient was diagnosed as a case of lupus panniculitis. He was started on oral antibiotic, antihistaminic, topical steroid, moisturizer cream and advised him for baseline investigations, serological investigations, skin biopsy and Immunohistochemical study. Routine investigations -including complete blood count (CBC), random blood sugar (RBS), liver function tests(LFT), kidney function tests (KFT), thyroid profile, bleeding time (BT), clotting time (CT), activated partial thromboplastin time (aPTT), prothrombin time (PT), urine analysis, chest x-ray (PA view) , ANA, Anti-ds DNA ECG, ultrasound abdomen, skin biopsy and Direct immunoflurescence test (DIF).

There was lymphocytosis in differential blood count, serum alkaline phosphatase (ALP) was high in LFT, ANA and anti-ds DNA both were elevated.

Histopathologic section showed mild pericapillary lymphocytic infiltration in dermis, lymphohistiocytic cell infiltration with focal areas of necrosis, ill-formed granulomas which was consistent with granulomatous panniculitis (Figure-2). DIF showed deposits of IgG, IgM, IgA and C3 along the dermo-epidermal junction.

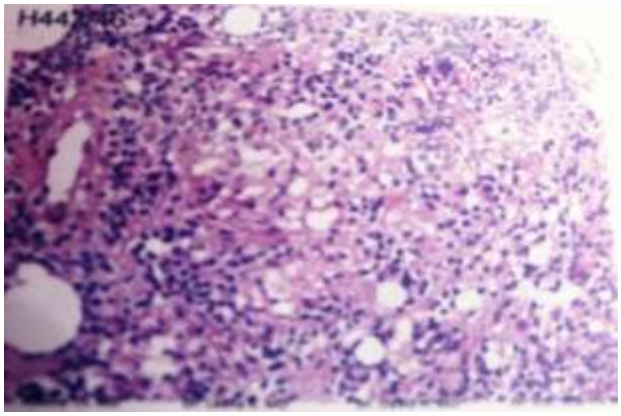

Figure 2:- HPE (H&E-10X) shows mild pericacapillary infiltration in dermis, lymphohistiocytic cell infiltration with focal areas of necrosis, ill-formed granulomas.

Hydroxychloroquine 200 mg (HCQs) orally twice daily and systemic corticosteroid (prednisolone 20 mg) once daily were started with symptomatic improvement & reduction of size & induration after 3 weeks. Oral steroid was gradually tapered to 10 mg/day and oral HCQs to 200 mg once daily. By 6 weeks all lesion reduced except 2 lesions over the cheeks which was resolved with 3 doses of Triamcinolone intra lesional injection (TAIL) 10 mg given for 3 weeks. The patient is on regular follow up with no recurrence of lesions till last follow up.

Discussion:-

Lupus panniculitis, also called *lupus profundus*, is a relatively rare abnormality of connective tissue. It may occur as a separate disease or coexist with SLE or discoid lupus erythematosus (DLE). Lupus panniculitis occurs twice more frequently as a separate disease than in association with SLE or DLE. On the other hand, 1–3% of patients suffering from SLE and up to 10% of those suffering from DLE develop lupus panniculitis.[5]

The pathogenesis of lupus panniculitis is quite similar to that of other types of LE. T lymphocytes and macrophages comprises the major infiltrates in lupus panniculitis . In Childhood onset and widespread distribution.Immunohistochemical analysis has shown an interferon-driven Th1- biased immune response in active lesions of lupus panniculitis; this may result in recruitment of cytotoxic CXCR3-positive lymphocytes.[1]

Lupus panniculitis is characterized by tender subcutaneous nodules and plaques. Typically the lesions tend to develop over face, upper arms, hip and trunk sparing the distal extremities. Changes in the overlying skin range from a light pink color to those of chronic cutaneous (discoid) LE. Lupus panniculitis often occurs prior to other manifestations of LE and in the absence of other autoimmune connective tissue diseases. There is a closer relationship of lupus panniculitis to other forms of chronic cutaneous LE (e.g. discoid LE) than to systemic LE. In approximately one third of patients, coexistent discoid lesions are found whereas only 10 to 15% patients meet the diagnostic criteria for systemic LE.[1]

The microscopy reveals a deep lymphocytic infiltration in the fat lobules and in the septa. Lymphoid aggregates, nodules & germinal centers also known as *follicular centers*, are common. The dermis can have superficial and deep perivascular lymphocytic infiltrate with plasma cells or can have all of the changes of lesions of discoid lupus erythematosus. A distinctive feature is the so-called "hyaline necrosis" of the fat can be present. Blood vessels are infiltrated by lymphoid cells and can have restriction of their lumen diameter.[4]

The most characteristic DIF finding in cutaneous LE is antibody deposition at the dermal–epidermal junction and around hair follicles. These deposits are composed primarily of IgG and/or IgM, although IgA can also occasionally be seen.[6] In our patient we found ill-defined granulomas, pericapillary lymphocyte infiltration in dermis and deep subcutaneous fat, focal areas of necrosis found on subcutaneous fat in HPE. DIF showed IgG, IgM, IgA and C3 along the dermo-epidermal junction.

Antimalarials are the mainstay of treatment and they confer promising response in majority of the patients. If a patient doesn't response to single anti-malarial, a second one can be added to increase the response. Chloroquine or hydroxychloroquine (200 mg once or twice a day) is the first choice. When monotherapy is ineffective then combination of antimalarials (for example, hydroxychloroquine 200 mg and quinacrine 100 mg daily) can be used . In chronic and relapsing nature of the disease, treatment may be required for several years.[1]

Topical corticosteroids (CS) effectively reduce inflammatory symptoms in all types of cutaneous lupus erythematosus (CLE). low-mid-potency CS (e.g., methylprednisolone) should be used on the face, midpotency CS (e.g., triamcinolone acetonide, betamethasone valerate) on the trunk and extremities, and high-potency CS (e.g., clobetasol) on the palms and soles where skin is thickest. Intralesional therapy with 2.5 to 10mg/mL triamcinolone solution may be of use in patients with localized DLE refractory to other treatment.[7] Our patient responded well with HCQs, oral corticosteroids except for the face lesions wchich were responded with TAIL 10 mg.

In refractory cases, treatment options include thalidomide, methotrexate, mycophenolatemofetil, cyclosporine and intravenous cyclophosphamide. McArdle et al. reported the use of Rituximab, which is a chimeric murine human monoclonal antibody to CD20 that induces depletion of mature B cells in vivo, to induce skin improvement in one adult with lupus panniculitis.[8]

Conclusion:-

Lupus panniculitis is a rare and challenging condition which can be often misdiagnosed. Here in the reported case LEP occurred as an isolated phenomenon without associated with DLE or SLE. Patient responded well to oral HCQs and oral corticosteroids except the face lesions which were resolved after intralesionaltriamcimnolone (TAIL) without any scarring.

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