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A CASE OF JUVENILE DERMATOMYOSITIS WITH PERITONEAL CALCIFICATION

NISHAD RAVEEDRAN
Department of General Medicine,
MADURAI MEDICAL COLLEGE AND HOSPITAL

Abstract :
Juvenile Dermatomyositis is a multisystem disease characterized by acute and chronic nonsuppurative inflammation of skin and striated muscle. Calcinosis is one of the hallmark squeal of juvenile dermatomyositis, and despite recent progress in the therapy of these disorders, dystrophic calcification still occurs in approximately one-third of patients. Ectopic calcification in JDM is thought to develop through a dystrophic mechanism. The sites most frequently affected are the elbows, knees, digits and extremities, although it may occur virtually anywhere over the body. This complication can cause significant debility with severe pain, skin ulceration, muscle atrophy, joint contracture, and acro osteolysis. Although various clinical patterns of calcification have been described peritoneal calcification is not reported so far. We report a case of Juvenile Dermatomyositis with extensive visceral peritoneal calcification.

Keyword : Juvenile Dermatomyositis,Calcinosis Case Report A 14 year old girl was admitted to our hospital with complaints of abdominal pain of 1 week duration.It was dull aching in nature,mainly felt in the epigastrium.No history of radiation of pain,no relation with food intake.History of early satity was present.No history of vomiting,constipation,hemetemesis or malena.No history of fever,jaundice.She is a known case of JUVENILE DERMATOMYOSITIS diagnosed 2 years ago in our hospital . Then she presented with 3 month h/o alopecia, violaceous papules over the dorsum of hands and elbow, pain both UL & LL and muscle weakness in the form of difficulty in standing from sitting position and difficulty in climbing stairs.On examination: Heliotropic rashes was present, Gotron papules was present, Other general examination findings were normal.Higher Mental functions,Cranial nerves were normal.Motor System:Bulk was normal bilaterally,Tone was normal bilaterally,Power : Grade5/5 both upperlimbs; grade 4/5 bilaterally in hip flexors and adductors.Distal muscles grade 5/5 bilaterally.Deep Tendon Reflexes were normal.Plantar: flexor plantar response.Other systemic examinations were normal.Routine

investigations were normal.EMG: Myopathic pattern with polyphasic spikes s/o inflammatory myopathy.Serum CPK:1356IU/L.Skin Biopsy:Perivascular lymphocytic infiltration in upper and lower dermis compatible with the clinical diagnosis of JDM.She was started on steroids, improved symptomatically .Her muscle weakness improved and skin lesions started disappearing and CPK levels normalized and discharged. Then she lost follow up and discontinued treatment.No other significant past history

PHYSICAL EXAMINATION:

Patient is conscious &oriented, Ill built & ill nourished, Pallor was present, No icterus ,No cyanosis, No clubbing ,No pedal oedema, No lymphadenopathy. Cutaneous calcifications were present,No other skin lesions were present.

Vitals :stable

Abdomen:Diffuse Mass palpable,8×6 cm,Epigastrium & left hypochondrium,Hard,Non tender,Irregular surface,Illdefined margins,Moves with respiration.No other palpable organomegaly. No free fluid clinically.

Other systemic examinations including spinomotor system were normal.At present there was no weakness of lower limbs.

Investigations

Hemoglobin -11g,TotalCount -7500,DifferentialCount –P65 L30 E2 M3,RBC -3.89 lakhs ,Platelet Count – 4.46 Lakhs ,PCV – 32%,ESR – 25mm/1st hour,Perepheral Smear – hypochromic microcytic anaemia RBS–36mg/dl,B.urea-29mg/dl,S.creatinine-0.5mg/dl,S.Sodium-139meq/dl,S.Potassium - 4.4meq/dl,Routine urine -normal study,

Liver Function Test: normal

S.Calcium:9.5gm/dl

S.Phosphorus:3.6 gm/dl

Mantoux test:negative

S.ALP:86U/L

S.PTH:40pg/ml

USG Neck:Thyroid normal.no mass.

OGD :Thickened mucosal folds in stomach

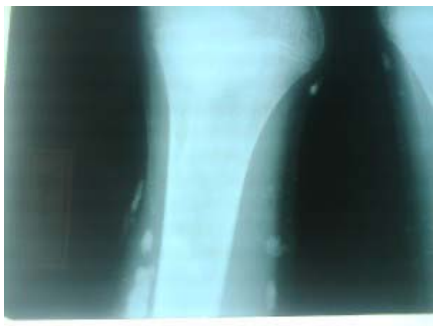
Biopsy: Nonspecific inflammatory changes

USG Abdomen

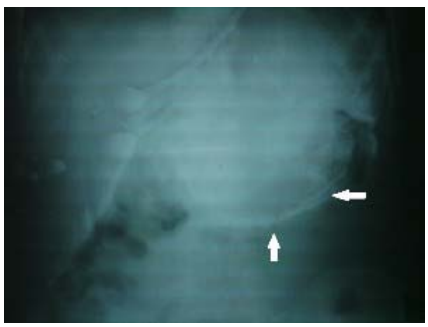
Liver – 10.8cm normal echoes,GB – normal,Pancreas – not separately seen,Spleen – 7.2cm, normal echoes,Kidneys – Normal,Dense linear hyper

echoic focus with shadow noted in the epigastrium ,No e/o free fluid

Imp - calcific pancreatic mass

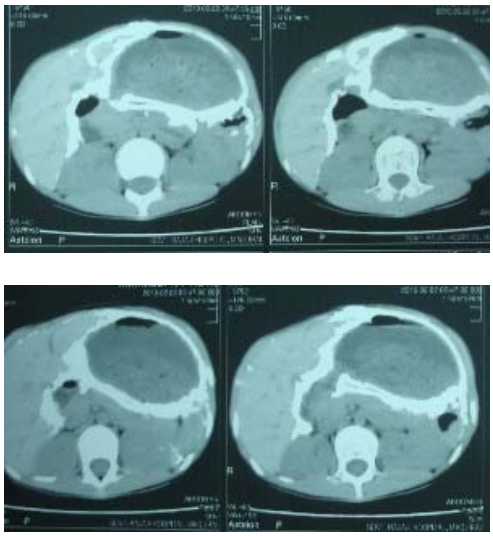


X Ray Right hand showing soft tissue calcification.



X Ray Adomen showing calcification along stomach wall

CT ABDOMEN



CT Abdomen showing extensive calcification involving visceral peritoneal lining of stomach,falciform ligament,undersurface of liver.All other intra abdominal organs normal.No intra abdominal lymphadenopathy.No evidence of free fluid.

A final diagnosis of Juvenile Dermatomyositis with soft tissue and peritoneal calcification was made.Patient was started on Steroids and Hydroxychloroquine and she is on follow up.

Discussion

Juvenile Dermatomyositis is a multisystem autoimmune disorder of unknown etiology.This results from inflammation of small vessels of muscles and skin and also other organ systems.This is characterized by muscle weakness due to muscle inflammation and a characteristic rash accompanying, or more often preceding muscle weakness. It accounts for 85% of pediatric inflammatory myopathy with an incidence of 2-3/million in children. JDM presents as progressive and symmetric muscle weakness. Initially it predominantly involves proximal muscles and as disease advances distal muscles also are involved. Ocular muscles are spared even in advanced untreated cases. If ocular muscles are involved diagnosis of inflammatory myopathy should be questioned.(10) The skin lesions in JDM includes(10) 1.Heliotrope rash: bluish purple discoloration of upper eye lid with edema 2.Gottrons sign:erythema of the knuckles with raised scaly eruptions 3.Flat red raised rash on the upper trunk and face Erythematous rash can also occur on other body surface area. Diagnostic criteria for JDM are currently still based on those established by Bohan and Peter in 1975 , which include a characteristic skin eruption, symmetrical proximal muscle weakness, elevated muscle enzymes, pathological muscle histology, and myopathic electromyographic changes. The presence of 3 of these criteria characterizes definite JDM, whereas the prevalence of 2 criteria makes the diagnosis probable.(7)Our patient fits in to the diagnosis of definite JDM as she has symmetrical proximal muscle weakness,skin eruptions,elevated CPK, typicalelectromyographic changes.

Calcinosis occurs in up to 30% of patients with JDM, although current prevalence ranges from 10 – 50% . The calcification is dystrophic, which by definition occurs at sites of injured tissue with simultaneously generally normal serum calcium and phosphorous levels . The sites most frequently affected are the elbows, knees, digits and extremities, although it may occur virtually anywhere over the body . The onset of calcinosis is most often 1 – 3 years after illness onset, but has been reported to occur from the time of illness onset to as long as 20 years later and now it is considered as a marker of inadequate treatment.

Ectopic calcification in JDM is thought to develop through a dystrophic mechanism, whereby damaged muscle releases mitochondrial calcium

into matrix vesicles, which then promote mineralization (8). Histological study of the lesions shows hydroxyapatite accumulation rather than bone (9).

Four subtypes of calcifications has been noted in JDM(5)

1.Calcinosis circuscripta
2.Tumoral calcinosis
3.Calcinosis along facial planes
4.All over the body surface

But the visceral peritoneal calcification as seen in this patient has not been reported.

Marked regression of calcinosis, as well as retardation or progression following treatment for 2 – 10 months with hydroxychloroquine, intravenous immunoglobulin, cyclosporin, and most recently with infliximab has been observed in a limited number of reported cases . Other agents which are tried includes Methotrexate ,Azathoprime and recently Diltiazem. Overall, the effectiveness of a particular agent appears to depend on its success in controlling the underlying inflammatory process, and many agents are potentially partially beneficial.

Calcinosis is associated with a variable natural history and a number of sequelae in children with JDM. Over an unpredictable period of time,spontaneous regression through reabsorption or extrusion of the calcific material may occur .Calcinosis may be more likely to improve in subjects with nactive disease, those who engage in physical activity, those with superficial plaques or nodules rather than those with deeper or more extensive deposits, and those who undergo an aggressive therapeutic regimen for JDM.(1),(5).

Diffuse peritoneal calcifications may be seen due to a number of causes.Usually the sheet like calcification as seen in our case is mostly seen in benign lesions while nodular calcification is seen with malignancy(4). Among malignancies ovarian carcinoma is most commonly associated with peritoneal calcification.Very rarely it is also associated with colonic and gastric carcinoma and peritoneal mesothelioma.These malignant calcification is also mostly associated with lymphnode calcification(4).Some malignancies like squamous cell lung carcinoma,malignant melanoma of skin and renal cell carcinoma can induce paraneoplastic hyperparathyroidismand hypercalcemia.They can produce sheet like peritoneal calcification(12).In our patient CT Abdomen showed normal ovaries,colon,kidneys,stomach.There was no intra abdominal lymphadenopathy/ascitis ruling out the possibility of malignancy.Her Serum Calcium and Parathormone levels werenormal ruling out hypercalcemia of malignancy.It can also be seen as a sequel to tuberculous peritonitis, *Armillifer armillatus infection (6).Other causes of peritoneal calcification are rupture of primary hydatid cyst in the peritoneal cavity, multiple phleboliths in haemangiomas, and sclerosing-peritonitis following long-term peritoneal dialysis(6).*

Among connective tissue disorders peritoneal calcification is reported in CREST syndrome(6).Our patient does'nt have history of Reynaud's phenomenon and skin thickening and no other features of CREST Syndrome. In our patients we have excluded almost all these causes. Her S.Calcium level is normal suggesting that it is a form of dystrophic rather than metastatic calcifications.The serum parathyroid hormone,S.Calcium and Phosphorous levels were normal ruling out the possibility of hyperparathyroidism. Moreover our patient is a proved case of JDM. Eventhough soft

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