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An interesting case report of polymyositis- presenting as quadriparesis with dysphagia and subcutaneous edema

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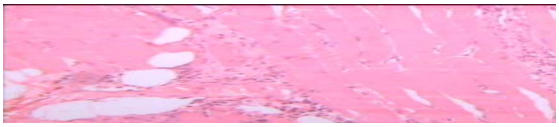
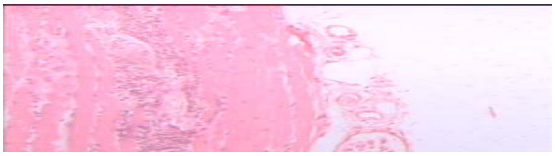
Abstract : A 15 year old boy was presented with complaints of weakness and swelling of all four limbs, fever, dysphagia and dysarthria for 10 days. Examination showed nonpitting subcutaneous edema of limbs, minimal tenderness in limbs, weakness was more in proximal than distal, lingual plus guttural dysarthria ,preserved gag reflex.CPK and LDH both were raised 100 fold.ANA weakly positive,anti dsDNA,antiJO,anti U1RNP ,anti CCP were negative.EMG, muscle biopsy and MRI muscle all showed polymyositis pictures. Patient was started on steroid and responded well within3weeks. We report a rare presentation of polymyositis with dysphagia ,dysarthria and diffuse non pitting subcutaneous edema.

Keyword :polymyositis,dysphagia,dysarthria,quadriparesis, subcutaneous edema

Muscle biopsy showing infiltrates.





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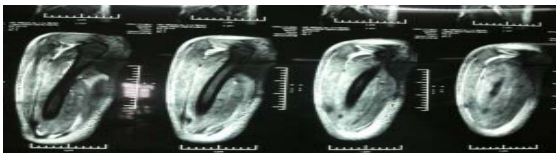
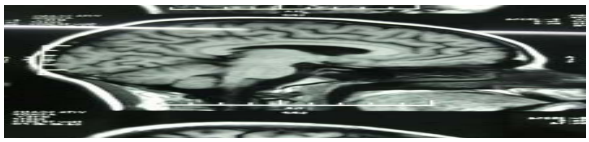


MRI THIGH-Shows signal hyperintensities in quadriceps muscle and calf muscles



MRI BRAIN-Normal which was done to rule out central demyelination.

Introduction:

Polymyositis itself is a rare disease, its presentation as quadriparesis with dysphagia,dysarhria, subcutaneous edema makes it a much rarer form. As because its very near differential diagnosis- AIDP a different disease their separation is must.

Case report:

A 15 yr old school going boy without any comorbid illness was presented with complaints of fever of low grade for 1 month, swelling of limbs, weakness of all four limbs proximal>distal, difficulty in swallowing and difficulty in speaking for 10 days.

No h/o trauma No history of oliguria, abdominal distension, facial puffiness, No history of skin rash, photosensitivity, hair fall, joint pain, easy fatiguability, oral ulcer or malar rash No h/o diplopia/decreased sensation over face/deviation of angle of mouth/vertigo/nasal regurgitation/chewing 0Past and family history were unremarkable.0 No h/o any drug intake or toxin ex-posure

EXAMINATION:

Higher mental function- normal

Nasal speech present, slurring of speech present,inability to pronounce ‘gna’ and’tha’ syllables. Skin and general examination normal except for diffuse nonpitting edema present in all 4 limbs,

All cranial nerve function test normal, **gag reflex present**,minimal bilateral sagging of soft palate,uvla in midline,palatal movements normal. Motor system**;tone;**normal, **power,**proximal 3/5 and distal 4/5 neck muscle 3/5, **DTR normally present**,plantar b/l flexor Sensory and cerebellar system function tests were normal.

INVESTIGATIONS:

CBC;TC;10900c/ml,DC;P63%L32%E5%,Hb;12g/dl,PCV;38%,ESR40mm/hour,platelet 1.38lakh/mcl,RBC;3.5million/mcl peripheral smear ;normal counts,microcytic hypochromic anemia, Serial renal function normal Liver function test normal except AST 234IU/L, ALP 548IU/L Urine myoglobin negative CPK 470IU/L and LDH

1906IU/L

With above history,examination and investigations patient was started with iv methylprednisolone 500mg for five days in view of neck muscle and bulbar muscle weakness and changed over to oral steroids @1mg/kg/day and continued for 2 weeks then tapper over 8 weeks. Meanwhile the following investigations were done, I.ANA-weakly positive

dsDNA-negative

antiJO negative

antiU1RNP negative-done to rule out associated connective tissue disorders II.antiCCP negative RA factor negative-Done to rule out associated rheumatoid arthritis III.Thyroid peroxidase antibody negative Thyroid function test normal- Done to rule out associated autoimmune thyroiditis/evidence for autoimmunity USG abdomen- normal study High frequency USG limbs showed subcutaneous edema As patient presented with fever,fever profile was done to look for other causes of fever HBsAg/anti HCV/HIV/ IgM dengue/chickungunya/MSAT/widal/ negative Blood and urine culture no growth

ELECTROMYOGRAPHY; spontaneous insertional fibrillation,low amplitude short duration polyphasic potential suggestive of myositis MUSCLE BIOPSY(Quadriceps) Biopsy showed muscle bundle infiltrates with necrosis and regeneration suggestive of polymyositis.

Echocardiogram-normal, no cardiomyopathy HRCT done to R/O ILD, as ILD usually associated with other autoimmune polymyositis and it was normal

MRI OF THIGHS

Done to rule out microabscess and cysticercae Shows signal hyperintensities in quadriceps muscle and calf muscles

MRI brain -Normal which was done to rule out central demyelination. In the course of stay in hospital patient showed significant improvement in the form of increase in motor power –proximal from 3/5 to 4+/5 and distal 4/5 to 5/5,dysphagia and dysarthria- reduced, edema completely subsided,muscle enzymes gradually reduced to LDH 320 IU/L within three weeks.

DISCUSSION:

PM occurs one per 100,000 a year(6) Polymyositis is an idiopathic inflammatory myopathy that causes proximal muscle weakness with elevated muscle enzymes ,characteristic EMG finding,muscle biopsy findings.(2) Bohan and Peter Classification(2)(5)

1. Primary idiopathic PM
2. Primary idiopathic dermatomyositis with characteristic skin changes
3. PM/DM with malignancy
4. Childhood PM/DM
5. PM/DM with connective tissue diseases
6. Inclusion body myositis
7. Miscellaneous

Polymyositis is a subacute inflammatory myopathy does not have extraocular and facial muscle weakness,family history,myotoxic drug intake,endocrine disorder,muscle dystrophy,inclusion body myositis,skin rash(1).It occurs due to aberrant immune response or autoimmune response of T cells(3). In PM cellular infiltrate is predominantly within the fascicles.(4).Il 1 and TNF alpha are increased.CD8/MHC I is characteristic association with PM. CD8 infiltrates the normal muscle fiber then with macrophages that makes the muscle necrosed.(1) Autoimmune etiology supported by antihistidyltransfer RNA synthetase or anti JO association with PM.

Connective diseases that can be associated with PM are SLE,RA,sjogren,mixed connective tissue disorders.

Autoimmune diseases that can be associated are crohn's disease,hashimoto thyroiditis,psoriasis etc…

Clinical presentation(2) Symmetric weakness(50-60%)(4) proximal muscles more involved Dysphagia(30%)once esophageal and pharyngeal straited muscles are involved, Cardiac/renal/lung involvement(10%)(4) Diagnosed by clinlcal,laboratory results (antiJO) and usually confirmed by muscle biopsy and EMG.muscle biopsy becomes critical to diagnose(metabolic myopathies,genetic testing) and to exclude other causes of muscle weakness.EMG to rule out neurologic causes.MRI of muscle become routine(4)as it is non invasive and can access a larger area than muscle biopsy,can be a guide to choose site for biopsy.

In our case there is unusual association of esophageal and bulbar weakness and subcutaneous edema(3) which takes us to a close differential diagnosis of viral myositis(4) in which case it closely mimics PM both clinical,lab and image vice can be differentiated by following, -no rhabdomyolysis and acute kidney injury in this patient,which is common in viral myostis -muscle biopsy show inflammatory infiltrates,but in viral myositis it will be absent -PM responded only with steroids wherein viral myositis steroids role is controversial -EMG supportive of PM Next near diagnosis will be AIDP with bulbar palsy,ruled out in this patient by following

-clinically, preserved deep tendon reflexs

-preserved gag reflex

-MRI brain no signs of demyelination

-EMG supportive of PM

Polymyositis itself a rare disease its unusual presentation with dysphagia,dysarthria and subcutaneous edema makes it a much rarer disease.

CONCLUSION:

Polymyositis eventhough a rare disease as it is potentially a treatable condition with prompt and early diagnosis,even when its unusual presentation like bulbar weakness and subcutaneous edema.

References;

1.Harrison textbook of internal medicine 18 th edition chat388

2.emedicine.medscape.com/article/335925 /feb 2013

3.Grandroundsjournal.comarticles/gr070007/2007/vol7/ polymyositis as subcutaneous edema

4.uptodate/article/inflammatory myopathies/2013

5.bohan A peter JB.polymyositis and dermatomyositis(part 1&2)N Engl Med 6.american rheumatology association/ diseases and conditions/inflammatory myopathies