

CASE REPORT

UNUSUAL PRESENTATION OF POLYMYOSITIS WITH RHABDOMYOLYSIS IN A CHILD

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Abstract

Acute onset of rhabdomyolysis is an uncommon presentation of polymyositis in children. A 4 years old female child presented with acute onset difficulty in swallowing. She developed acute rhabdomyolysis and weakness of all extremities and trunk. MRI revealed polymyositis which was confirmed by muscle biopsy. She was treated with intravenous fluids, intravenous immunoglobulin (IVIG) and steroids. She recovered in next 14 days.

Introduction

Polymyositis is a disease of muscles featuring inflammation of muscle fibres. The muscle weakness begins from muscles close to the trunk over a period of weeks or months. (1) Rarely do they complain of muscle pain, breathing difficulty and difficulty in swallowing. Polymyositis is uncommon in children. (2,3) Presentation as acute rhabdomyolysis with severe muscle pain is even rarer in polymyositis. (4) Diagnosis in such situations is difficult and timely management is life-saving. We present a 4 years old female child with acute onset difficulty in swallowing followed by acute rhabdomyolysis and weakness of all extremities and trunk due to polymyositis. She had complete recovery with intravenous immunoglobulin (IVIG) and steroids.

Case Report

A 4 years old female child presented with acute onset breathlessness while playing indoors. There was no history of any insect bite, rash, foreign body aspiration, injury or drug ingestion. Mother also noted that the child had difficulty in swallowing and had taken her to a local hospital where she was treated with oxygen and salbutamol nebulization. However, she did not improve and hence was referred to our hospital. On admission she was afebrile, tachypneic with no intercostal or subcostal retractions, pulse rate was 150/min, respiratory rate was 45/min, and blood pressure was 100/70mm of Hg. Systemic examination was normal. She maintained a saturation of 96% in room air. Biochemical evaluation showed a sodium of 130 mEq/L, potassium of 4.2 mEq/L and bicarbonate of 17 mEq/L. Her urea was 45mg/dl and creatinine was 0.9mg/dl. Hemoglobin was 15.6 gm/dl, total white cell count 18000 cells /cumm (polymorph 90%, lymphocyte 8% and eosinophil 2%), platelet count 4,30,000/cumm. Total bilirubin was 1.1 mg/dl, SGPT was 1319 IU/l. Her urine output was 1.5 ml/kg/hour. At 6 hours of hospital stay, she developed red colored urine and urine revealed 2+ albumin, few RBC's with hyaline casts and protein: creatinine ratio of 0.4. Urine was negative for myoglobin and hemoglobin. Urine colour returned to normal in the next 6 hours. Repeat urea and creatinine were normal. Chest X-ray, ECG, Ultrasonogram (USG) of the abdomen and echocardiography were normal. On day 2 of hospital stay she developed severe myalgia over the extremities

predominantly the shoulders and hips. Her CPK was 1,00,000 IU/L. In view of difficulty in swallowing and throat pain, throat swab was sent which was negative for diphtheria. Repeat CPK was 1,52,000 IU/L, LDH was 6240 IU/L and ESR was 30 mm at one hour. She was started on forced alkaline diuresis for a brief duration. Her renal parameters and serum electrolytes were within normal limits. CPK continued to increase to 1,99,000 IU/L. Neurological examination revealed conscious child with normal cranial nerves, hypotonia of all four limbs, and power of 4/5 over the gluteal and hamstring muscles, absent deep tendon reflexes and a flexor plantar response. Snake envenomation was suspected and she received full dose of antsnake venom on the 2nd day of admission. She had head lag and was unable to get up from bed on day 3 of hospital stay. MRI muscles showed symmetric hyper intensities involving ileopsoas, sartorius, obturator, rectus femoris, gluteus maximus, medius, minimus & adductors on diffusion weighted images. Child did not have any rash suggestive of dermatomyositis. Muscle biopsy was done on the 4th day of hospital stay, from the right gluteus maximus muscle. She was given Inj methylprednisolone 30mg/kg/day for three days. Since she did not show any clinical improvement she was given intravenous immunoglobulin (IVIG) 400g/kg/day for 5 days and by day 7 of hospital stay started to have clinical improvement. Her CPK decreased to 10,100 IU/L by the 8th day of hospital stay. Her muscle biopsy histology by light microscopy showed preserved architecture, acute necrosis, myophagocytosis, interstitial inflammation, regenerating fibers and edema suggestive of acute rhabdomyolysis and polymyositis. Nerve conduction study done in the second week of illness was normal. Autoimmune antibody work up revealed ANA 6.76 units (Normal <20), Anti - Jo antibodies 0.59 U/ml (normal < 3), C3 102.44 mg/dL (normal - 90-120) and C4 18.09 mg/dL (normal 10-40). Child was discharged on oral steroids at a dose of 1 mg/kg/day and was walking without any residual defect on follow up after one month. She is on follow up with the rheumatologist regarding tapering dose of steroids. Her CPK values are within normal limits.

Discussion

Juvenile dermatomyositis (JDM) and Juvenile polymyositis (JPM) are rare autoimmune disorders in children. JDM is the more common of the two disorders accounting for approximately 85 percent of idiopathic inflammatory myopathies of childhood. (5) JPM accounts for only 3 to 6 percent of cases. (2, 3) The classification of idiopathic inflammatory myopathies developed by Bohan and Peter in 1975 still is being used despite different classifications using the new additional information in the recent years. The diagnostic criterion based on Bohan and Peter(6) criteria include the following : Muscle involvement - as symmetrical and progressive proximal muscle weakness (dysphagia and respiratory

involvement), muscle biopsy with necrosis of type I and II fibers, phagocytosis, regeneration with basophilia, large vesicular sarcolemmal nuclei, prominent nucleoli, atrophy in a perifascicular distribution, variation in fiber size, inflammatory exudates, often perivascular, elevation of muscle enzymes particularly creatine phosphokinase, often aldolase, AST, lactate dehydrogenase and electromyogram with short, small, polyphasic potentials and complete recruitment.

Our patient had an acute onset of breathlessness and dysphagia mimicking a foreign body aspiration or envenomation or poisoning. Muscle weakness was predominantly over the extremities to begin with. Severe muscle pain with rhabdomyolysis is rarely reported in JPM in literature. (4) The muscle weakness is predominantly in the trunk including the neck flexors, limb girdle and trunk muscles. Distal muscles are involved in severe cases. Pharyngeal, hypopharyngeal and palatal muscles can be involved in 25% of children. Risk of aspiration may be high in such cases. (7) Arthralgia is common in polymyositis, however if it presents with severe arthritis then one has to look for overlap syndromes. Clinical picture with weakness of muscles with elevated enzymes should suggest the possible diagnosis of polymyositis. Skin involvement favors the diagnosis of more common dermatomyositis. Interstitial pneumonitis, cardiac involvement and GI involvement can be associated with polymyositis and the presence of various antibodies can give a clue for the diagnosis. Electromyogram can help to diagnose polymyositis as well as rule out other close differential diagnosis. MRI can show the sites of muscle inflammation and this can be used to identify the sites of muscle biopsy. Muscle biopsy is confirmatory in its histopathology. Presentation with such acute onset and with rhabdomyolysis is rare in juvenile polymyositis. Rhabdomyolysis can precede the development of polymyositis by years. Isolated presentation as rhabdomyolysis need to be followed for occurrence of muscle weakness. (4)

Muscular dystrophy can mimic the presentation as polymyositis and literature has shown similar studies where the children diagnosed as polymyositis have been finally diagnosed as muscular dystrophy. (8) Viral myositis with rhabdomyolysis can mimic the diagnosis in children. Management of polymyositis is by corticosteroids. In acute presentation intravenous preparations can be used followed by oral medication as used in this child. (9,10) If unresponsive to steroids, IVIG, immunosuppressive agents like methotrexate can be tried. Overlap myositis that is associated with conditions like scleroderma, systemic lupus erythematosus, juvenile immune arthritis, Kawasaki disease have a relatively milder muscle disease with a favorable response. Outcome can be good with recovery and no further recurrence (monocyclic), some continue to have disease activity as chronic polymyositis and some may have remission followed by recurrence later. (11)

Conclusion

Acute presentation of polymyositis with rhabdomyolysis is rare in children. MRI of the muscles may provide a valuable clue for diagnosis. Intravenous steroids and IVIG may be helpful in the management of the acute phase.

References :

1. Dalakas MC, Hohlfeld R. Polymyositis and dermatomyositis. *Lancet*. 2003; 362: 971-982
2. McCann LJ, Juggins AD, Maillard SM, Wedderburn LR, Davidson JE, Murray KJ, et al. The Juvenile Dermatomyositis National Registry and Repository (UK and Ireland)--clinical characteristics of children recruited within the first 5 yr. *Rheumatology (Oxford)* 2006; 45:1255-1260.
3. Symmons DP, Sils JA, Davis SM. The incidence of juvenile dermatomyositis: results from a nation-wide study. *Br J Rheumatol* 1995; 34:732-736
4. Tabarki B, Thabet F, Selmi H, Yacoub M, Mokni M, Korbi S, et al. Acute rhabdomyolysis disclosing polymyositis in children. *Arch Pediatr*. 2001; 8: 51-54
5. Rider LG, Miller FW. Deciphering the clinical presentations, pathogenesis, and treatment of the idiopathic inflammatory myopathies. *JAMA*. 2011; 12; 305: 183-190
6. Bohan A, Peter JB. Polymyositis and dermatomyositis . *N Engl J Med* 1975;292(7):34-7
7. Compeyrot-Lacassagne S, Feldman BM Inflammatory myopathies in children. *Pediatr Clin North Am*. 2005; 52: 493-520, vi-vii.
8. D'Arcy CE, Ryan MM, McLean CA. Juvenile polymyositis or paediatric muscular dystrophy a detailed re-analysis of 13 cases. *Histopathology*. 2009; 55: 452-462
9. Murthy JM. Drug treatment of polymyositis and dermatomyositis. *Neurol India*. 2010; 58: 3-5
10. Mastaglia FL. Inflammatory muscle diseases. *Neurol India*. 2008; 56: 263-270
11. Huber AM, Lang B, LeBlanc CM, Birdi N, Bolaria RK, Malleson P, et al. Medium- and long-term functional outcomes in a multicenter cohort of children with juvenile dermatomyositis. *Arthritis Rheum*. 2000; 43: 541-549.

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**DOI: 10.7199/ped.
oncall.2014.15**



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