

Two Cases of Polymyositis associated with Liver cirrhosis

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Abstract

Polymyositis is a nonsuppurative inflammatory muscle disease of unknown etiology.

Recently, we have experienced two cases with Polymyositis associated with liver cirrhosis.

General weakness, general muscular ache(neck and back), massive hematuria and the marked elevations of CK, LDH, aldolase, and CPK were characteristic clinical findings.

They had previously liver cirrhosis. One patient died soon after admission(due to acute hyperkalemia).

One patient was female and she had liver cirrhosis related to HCV infection. The other patient was male in which alcoholic induced liver cirrhosis. He underwent hemodialysis due to acute renal failure. After hemodialysis level of CK, LDH, CPK, and aldolase decreased to normal value. Also BUN. and Cr. values decreased to normal value.

The causes of Polymyositis in cirrhosis are not known yet; However, we suggest that it might be related to the viral infection or combined with autoimmune state.

We present the cases with review of literature; and these cases were rapid progressive disease and needed emergency treatment.

Polymyositis rarely leads to acute renal failure. Two case were reported here, and certain their common features in reported cases are discussed. Given therapeutic advances, these patients should do well despite the dismal prognosis reported in the literature, but early diagnosis and prompt treatment are essential.

Key Words : Liver cirrhosis, Polymyositis, acute renal failure.

Introduction

Polymyositis^{1,2,3)} is a systemic disease and patients may develop morning stiffness, fatigue, anorexia, weight loss, and fever. Arthralgia is not uncommon, but frank synovitis is less usual. Raynaud's phenomenon is sometimes present. Muscle pain and tenderness are present in about half of patients. Periorbital edema may occur. The neurologic portion of the physical examination is normal except for motor function.

Typically adult-onset polymyositis begins insidiously over 3 to 6 months with no identifiable precipitating event. Only very rarely is the onset abrupt and associated with clinically evident rhabdomyolysis^{3,4,5)} and myoglobinuria.^{3,4,5)} The muscle weakness initially affects the muscles of the shoulder and pelvic girdles, with the latter slightly more common. Distal weakness is uncommon initially but can develop over time in severe cases. Weakness of neck muscles, particularly the flexors, occurs in about half of the patients. We encountered two cases of polymyositis associated with liver cirrhosis who had acute renal failure.

One patient expired. The other patient was survived and polymyositis improved after prednisolone therapy along with hemodialysis.

Case report

One case : a 70-year-old female patient was admitted via the ER for general weakness for 2 days. She felt well until 2 days before admission, when she began having difficulty

walking and dressing. She denied either engaging in strenuous exercise or using alcohol.

On physical examination, her body temperature was 37°C, respiratory rate was 20 breaths per minute, and blood pressure was 110/70 mmHg. there was 1+ pitting edema bilaterally up to middle thigh, but there was no respiratory distress. Results of cranial nerve test were normal,

lower extremity deep-tendon reflexes were absent, and there was normal muscle tone without acropathy. Muscle strength was 3/5 in the proximal and 3/5 in the distal upper extremities and 3/5 in the remainder of the lower extremities.

She has diagnosed HCV induced liver cirrhosis at 1990. After that time she admitted 5th times for α -INF and supportive therapy. Recently she visited for general weakness at May. 25 1998,

But she had normal S-GOT, LDH. After that time she had been do well.

On admission urine output was normal, urine tests for myoglobin were negative but others, urinalysis were protein 3+, erythrocyte 3+. Laboratory tests yielded the following values(Table-1). Chest view was normal and EKG was also normal Her EMG(Fig-1) finding showed reduced interference pattern on contraction in right vastus medialis and right biceps brachii long head muscle and discrete activity on maximal contraction on both gastrocnemius medialis, both vastus lateralis and right tibialis anterior muscles which were not the completely characteristic of polymyositis. Muscle biopsy could not be performed.

Table-1 Showed laboratory data on admission and two days after.

	LDH IU/L	CPK IU/L	S-GOT IU/L	Aldolase SU/L	Cr mg/dl	BUN mg/dl	Na/K meq/dl
98.8.5	2094	1600 ↑	1925	30	1.0	12	132/4.8
98.8.6	2000		2100		3.2	56	122/8.2

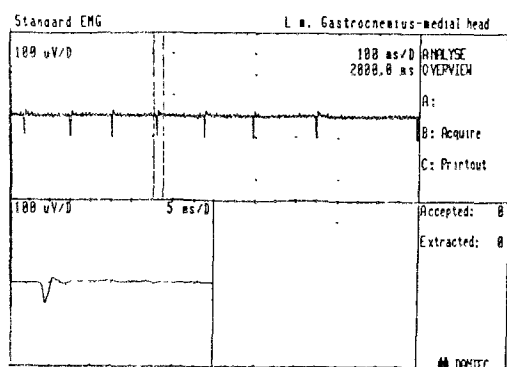


Fig-1. EMG showed reduced interference pattern on contraction on both gastrocnemius medialis.

Follow up EKG was normal. But on urinalysis, massive hematuria was occurred. We treated prednisolone 60 mg and IV globulin 2 days after admission. On 2day of admission her urine out put decreased 400ml/day. When we sampled BUN, creatinine, and sodium, potassium. she had respiratory difficult We checked EKG three times and the first two were normal and last EKG was acute hyperkalemia.

The other case was a 62-year-old man who admitted via the OPD for general weakness for 2 days.

He felt well until 2 days before admittance, when he began having difficulties in walking and dressing. He denied either engaging in strenuous exercise or using alcohol.

On physical examination, his temperature was 36.5°C, respiratory rate was 20 breaths per minute, and blood pressure was 140/90

mmHg. There was 1+ pitting edema bilaterally up to middle thigh, but there was no respiratory distress. Results of cranial nerve test were normal, deep-tendon reflexes were decreased in the lower extremity, However there was normal muscle tone without acropathy. Muscle strength was 3~4/5 in the proximal and 4/5 in the distal upper extremities and 2~3/5 in the proximal and 4/5 in the distal of the lower extremities.

He has diagnosed alcohol induced liver cirrhosis in 1994. On admission, he had normal urine output, urine tests for myoglobin were negative and other results of urinalysis were protein negative 2 RBC cast. Laboratory tests yielded the following values (Table-2). Autoantibody were negative (ANA, Double strand DNA Ab, Anti microsomal Ab, Antimitochondrial Ab)

Chest view was normal and myocardial ischemia was seen on EKG.

On his EMG (Fig-2), there was positive sharp waves in right tibialis anterior and left gastrocnemius medialis muscles and wide spread fibrillations with marked myopathic changes on electromyogram, which are characteristic of acute severe myopathy. After 8 days, a deltoid muscle biopsy (Fig-3) revealed several necrotic fibers but we could not find endomysial inflammatory cells, which suggests that treated with prednisolone previously. However It was characteristic of myopathy.

The patient was vigorously rehydrated and

Table-2 Showed laboratory data on admission and the days after.

1998	08.01	08.04	08.10	08.15	08.22	09.02
U/A SG/P/S/ERY.			1.005/-/-/++	1.010/-/-/++	1.010/-/-/++	1.010/-/-/++
BUN/Cr mg/dl	21/1.1	72/4.4	109/6.9	46/3.1	24/1.2	17/1.0
Na/K meq/dl	130/3.9	128/5.7	118/5.9	132/4.1	130/3.5	141/12.6
S-GOT IU/L		1654	137	20	27	24
LDH IU/L		3986	3091	1409	1233	437
Aldolase SU/L		30 ↑				3
CPK IU/L		1600 ↑		1082	68	57
RA Neg IU/L		+				

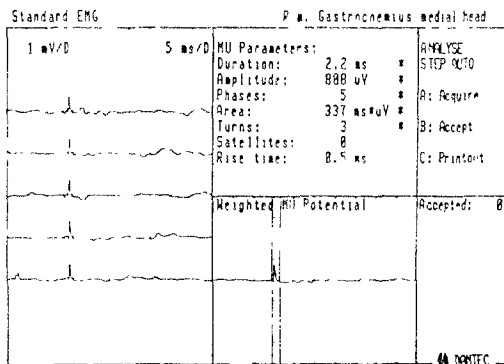


Fig-2 EMG showed positive sharp waves gastrocnemius and wide spread fibrillations.

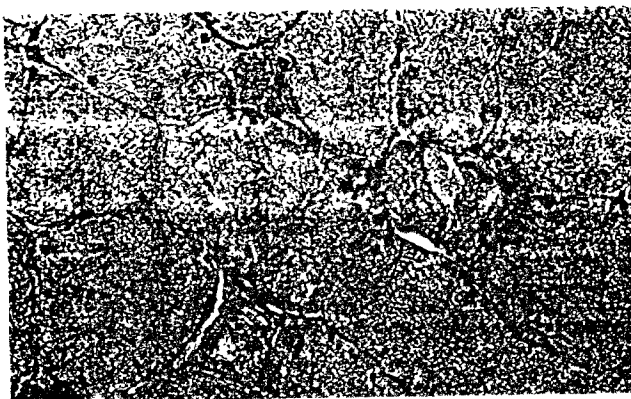


Fig-3 deltoid muscle biopsy revealed several necrotic fibers but is not seen endomysial inflammatory cell

We used 60 mg of prednisolone every 8 hours.

He had hemodialysis four times. After then, he had normal BUN, Cr. The patient's strength gradually improved and also the renal function. He was discharged walking by himself.

Discussion

Polymyositis is an inflammatory myopathy in which skeletal muscle is damaged diffusely by a perivascular and/or interstitial infiltration of inflammatory cells which are predominantly lymphocytes. The etiology of the disease is unknown but probably involves genetic factors, viral infection of muscles, and autoimmune^{7,8,9)} mechanisms. Polymyositis may develop at any age and in either sex. It represents as a proximal symmetrical muscle with pain and tenderness. One-third of cases are associated with connective-tissue disorders and one-tenth with a malignancy.¹⁴⁾ A diagnosis is based in the typical clinical picture, a typical electromyogram, elevation of serum creatine kinase, and a diagnostic muscle biopsy. When muscle destruction is acute and extensive, myoglobin can be found in the

urine.

There have been very few reported cases of rhabdomyolysis occurring in patients with polymyositis resulting in acute renal failure.^{5,6)} In rhabdomyolysis,^{4,5)} widespread muscle injury releases large quantity of myoglobin into the circulation. Myoglobin is freely filtered and can be detected in the glomerular filtration. Endocytotic proximal tubular reabsorption is overwhelmed and high concentrations are reached in the urine. However, not all patients with myoglobinuria will develop renal failure. There is no clear relationship between the levels of myoglobinuria and the development of renal failure. Myoglobin itself is relatively nontoxic, but becomes highly toxic in the presence of acidosis, dehydration, or both. At acid pH, ferrihaemate dissociates from myoglobin and depresses effective tubular transport without a decrease in oxygen utilization. Also myoglobin enhances vasoconstriction by inhibiting the production of relaxing factor. Therefore it has been proposed that renal damage could result from an imbalance between reduced oxygen supply and continued oxygen demand.

Diagnosis can be difficult as myoglobin is often not detected in the urine, which is probably related to markedly reduced GFR and oliguria that occurs in the full syndrome of rhabdomyolysis.

These cases showed that liver cirrhosis induced polymyositis. One case was female. She died suddenly after 2 days admission. She had hepatitis C induced liver cirrhosis. The other case was alcohol induced liver cirrhosis. There have been very few reported cases of polymyositis occurring in patients with liver cirrhosis^{10 11 12 13)} and also

there have been very few cases of acute renal failure^{4,5,6)} occurring in patients with polymyositis.

Although, the characteristic triad, myoglobinuria due to rhabdomyolysis, high CPK level, myoglobinuria, and pigmented casts in the urine could not be satisfied with two cases,

they had high CPK level and erythrocytes were found in the urine sediment but could not detect myoglobinuria.

The subset of patients with liver cirrhosis induced polymyositis had symptoms of dramatic muscle weakness and acute renal failure. In two cases acute renal failure occurred at the onset of disease, and the severity of symptoms led patients to seek prompt medical attention.

Reference

1. Harrison's : PRINCIPLES of INTERNAL MEDICINE, 14th ed, U.S of America, McGraw-Hill, 1998, 1896~1901.
2. Kelly. Harris. Ruddy. Sledge : Textbook of Rheumatology. fourth ed. 1993, 11 : 1164~1165
3. Raymond. D. Adams. Maurice Victor. Allan H. Ropper.; Principles of Neurol. 6th ed,
4. Case Report : Polymyositis-Induced Myoglobinuric Acute Renal Failure. Am J Med Sci 312(2) : 85~87, 1996
5. Case Report; Polymyositis : a cause of acute renal failure. Nephrology Dialysis Transplantation. 11 : 699~701, 1996
6. Lim-R;Parker-JL : Apparent haematuria : an early sign in polymyositis ;Br-Med-J-Clin-Res-ed. 296(6638) : 1725~6, 1988 Jun 18

7. Robert W. McMurry and Keith Elbourn e : Hepatitis C Virus Infection and Autoimmunity. *Seminars in Arthritis and Rheumatism*, Vol 26, pp 689~701, No 4(February),1997
8. Horsmans Y, Geubel AP : Symptomatic myopathy in hepatitis C infection without interferon therapy.[letter; comment]. *Lancet* 345 : 1236, 1995
9. Ueno Y, Kondo K, Kidodoro N, et al : Hepatitis C infection and polymyositis. [letter;comment]. *Lancet*, 346 : 319~320, 1995
10. Polymyositis Associated with Asymptomatic Primary Biliary Cirrhosis. *Eur Neurol.* 33:51-53, 1993
11. Jong Ho Ahn : A case of polymyositis in a patients with primary biliary cirrhosis treated with D-penicillamine; The korean journal of internal medicine. Vol 8 No 1. january. 1993
12. Boki-KA; Polymyositis Associated with Primary Biliary Cirrhosis. *ClinRheumatol.* 14(3) : 375~8. 1995 May
13. Sang-Young Han; Clinical study of Polymyositis-like syndrome associated with Liver cirrhosis. *Dong A University Journal of medicine.* 49~59, 1994
14. Sattar-MA; Polymyositis and hepatocellular carcinoma. *Clin-Rheumatol.* 7(4) : 538~42, 1998 Dec

국문초록

다발성 근염은 원인이 알려져 있지 않은 비화농성 염증성 근육질환이다. 최근에 저자는 2예의 간경화에 병발된 다발성 근염을 경험하였다.

다발성 근염의 일반적인 임상양상은 전신적 피로감 전신적 근육통 다량의 혈뇨 그리고 CK, LDH, aldolase 와 CPK의 급격한 상승을 볼 수 있다.

두환자는 이전에 간경화로 치료를 받고 있었으며 한 환자는 입원후 곧 사망하였다. 사인은 급성고칼륨혈증으로 추정되며 환자는 HCV 간염에 병발된 간경화였다. 나머지 환자는 알콜에 의한 간경화 환자이다. 환자는 급성신부전으로 혈액투석을 받았으며 CK, LDH, CPK, 그리고 aldolase 모두가 정상수치로 돌아왔으며 BUN, Cr.도 정상수치로 돌아왔다.

간경화에 병발된 다발성 근염 원인은 아직알려져 있지 않으나 바이러스성 감염이나 자가면역과 연관된 가능성으로 인식되고 있다.

저자들은 2예의 증례보고와 문헌고찰을 보고하는 바이며 이러한 예들은 급진적으로 진행되는 병이므로 응급치료를 요하며 다발성 근염은 급성신부전으로 잘 진행되지 않는다

그러나 본 증례의 경우 모두급성신부전으로 진행이 되었다.

이러한 환자들은 예후가 좋지않으나 치료가 발달함에 따라 조기진단과 즉각적인 치료를 하여 예후가 좋아질 수 있다.