

Case Report

Macro CK and Idiopathic Focal Myositis: An Atypical Case of False Rbdomyolysis Due to Convergence of Two Infrequent Benign Conditions

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Abstract

We present an 18 years old man who developed a tender and swollen pectoral mass since the morning. After an initial diagnostic approach he was treated as a sport related rbdomyolysis however levels of creatinine phosphokinase were disproportionate high to the muscle mass involved. An ultrasonography study suggested an inflammatory local myositis as well the report of an electrophysiology approach. Levels of CPK decreased after treatment with prednisone but never got normal. An electrophoresis analysis of the CPK molecule demonstrated a Macro-CPK. A brief discussion about both uncommon clinical features is also included.

Keywords: Macro-CPK; Local inflammatory myositis; Rbdomyolysis

Case Presentation

An eighteen years old male presented to our clinic due to swelling and pain over the right pectoral area of his chest.

Patient was a non professional basketball player used to play or train three times a week. Last training took place a week before. He did not have any personal or family history of autoimmune diseases. There were no registries of previous medical consultations after he completed his pediatric regular follow up four years ago.

That morning he woke up and notice pain and swelling over the right pectoral. The night before, he was asymptomatic. The chest pain was present continuously but it was greatly increase by moving the right arm away from the chest or during an external rotation movement. He denied use of drugs of abuse or any kind of over the counter prescription.

Physical examination demonstrated normal constants and no fever. Patient had an athletic constitution. The right pectoral was tender and swollen without skin lesions, local heat or erythema (Figure 1). All movements involving major pectoral muscle were painful and limited. There were no other remarkable findings in the physical exam.

Lab tests were as follows: Red and white blood cells and platelets were normal. The erythro sedimentation rate was slightly high (25mm per hour). C-reactive protein was normal as such the liver profile. Creatin phosphokinase (CPK) levels were severely high (>60000U/L, normal range up to 240U/L according to our lab). Creatinine levels and urine analysis were completely normal.

A musculoskeletal ultrasonography performed immediately showed enlargement of the height of the major pectoral muscle and a hypoechoic fiber pattern predominantly seen into the external portion of the muscle compared to the left major pectoral.

Patient was treated as a case of sport related rbdomyolysis with

regular biochemistry controls of creatinine, acute phase reactants and CPK. After 24 hours CPK levels reduced almost 5%. Creatinine levels and urine analysis were always normal as well as liver profile. An electrophysiology study showed profuse denervation potentials with motor unit action potentials of short duration and reduced amplitude from the same portion of the right major pectoral. Considering the diagnosis of focal idiopathic myositis, patient started on prednisone 1mg/Kg/day. Two weeks after, chest appearance returned to normal and tender was completely relief. However, a month after starting prednisone CPK levels was still over 6000 U/L without any symptoms. An electrophoresis CPK study was ordered and it demonstrated a Macro-CPK IgG I. Patient did not accept to undergo a muscle biopsy due to the absence of symptoms.

Discussion

This is an extraordinary clinical case where two different but organ related infrequent processes took place into the same patient.

The Idiopathic Focal Myositis (IFM) is a benign rare condition related to striated muscle seen most frequently in young male adults



Figure 1: Aspect of the chest of the patient on the first day. Arrow points the lateral portion of the right major pectoral.

but seen at any age [1]. It often involves distal parts of lower limbs however it has been described in non peripheral areas [1-4]. There are no universally recognized diagnostic criteria for IFM however it can be suspected on patients with a focal swollen and tender muscle not necessarily related to an overexertion, denervation electrophysiological pattern and an image proof of muscle edema and fat infiltration [1,2]. General therapeutic recommendation is to start prednisone 1mg/Kg/day or equivalent [1,2,5]. A few patients with not enough good clinical response have shown benefit from treatments with methotrexate or even local therapies, when the size of the lesion allows it [6].

By the other hand, macro-CPK is a generally non pathological condition where the circulant CPK molecule is attached to a pre-protein modifying its molecular weight without having a significant change in its regular functioning [7-9] its prevalence in global population is less than 0.5% [10]. The major problem with is kind of CK isoenzyme is the effect over the regular methods of biochemistry measures. The cuantification of CPK in patients with macro-CPK always is higher than normal range. On special situations, like sports injuries, levels could be even ten or more folds the normal. By the contrary, the real mass of the isoenzyme is not as high and patients could misdiagnose of severe rabdomyolisis [9].

Our case report seems to summarize both infrequent circumstances. Our patient carried a macro-CPK since probably he was born and it was never had been identified. Also, he developed a focal myositis of the lateral portion of the major pectoral. Presence of signs of enlargement and muscle edema, demonstrated by ultrasonography, the electrophysiology findings and the good response to prednisone suggest an IFM, however since patient did not accept to undergo a biopsy the diagnosis is just highly probable and not definite. The major problem with this case is that the status of macro-CPK was responsible of the high amount of CPK measured by conventional lab methods. Common cases of rabdomyolisis linked to sport injuries or

overexertion can show high levels of CPK with other major clinical features as choluric urine and renal failure. Conventional therapy is fluid support. Our case featured with a relative small volume of muscle affected and an extremely disproportionate level of CPK without any choluria or renal impairment. The combination of IFM and macro-CPK easily explain this phenomenon.

References

1. Auerbach A, Fanburg-Smith JC, Wang G, Rushing EJ. Focal myositis: a clinicopathologic study of 115 cases of an intramuscular mass-like reactive process. *Am J Surg Pathol.* 2009; 33: 1016-1024.
2. Heffner RR, Armbrustmacher VW, Earle KM. Focal myositis. *Cancer.* 1977; 40: 301-306.
3. Flaisler F, Blin D, Asencio G, Lopez FM, Combe B. Focal myositis: a localized form of polymyositis? *J Rheumatol.* 1993; 20: 1414-1416.
4. Prop S, van Vuurden D, van der Kuip M, van der Voorn JP, Plötz FB. [A boy with cervical focal myositis]. *Ned Tijdschr Geneesk.* 2014; 158: A6935.
5. Garcia-Consuegra J, Morales C, Gonzalez J, Merino R. Relapsing focal myositis: a case report. *Clin Exp Rheumatol.* 1995; 13: 395-397.
6. Mitrovic J, Prka Z, Zic R, Marusic S, Morovic-Vergles J. Focal myositis of lower extremity responsive to botulinum A toxin. *Clin Neuropharmacol.* 2014; 37: 55-57.
7. Etienne E, Hanser AM, Woehl-Kremer B, Mohseni-Zadeh M, Blaison G, Martinot M. [Macroenzymes: macro-ASAT and macro-CPK. Two cases and literature review]. *Rev Med Interne.* 2009; 30: 963-969.
8. Axinte CI, Alexa T, Cracana I, Alexa ID. Macro-creatine kinase syndrome as an underdiagnosed cause of ck-mb increase in the absence of myocardial infarction: two case reports. *Rev Med Chir Soc Med Nat Iasi.* 2012; 116: 1033-1038.
9. Liu CY, Lai YC, Wu YC, Tzeng CH, Lee SD. Macroenzyme creatine kinase in the era of modern laboratory medicine. See comment in PubMed Commons below *J Chin Med Assoc.* 2010; 73: 35-39.
10. Lee KN, Csako G, Bernhardt P, Elin RJ. Relevance of macro creatine kinase type 1 and type 2 isoenzymes to laboratory and clinical data. *Clin Chem.* 1994; 40: 1278-1283.