PERSISTENT ELEVATION OF TRANSAMINASES: AN IGNORED INDICATOR OF POLYMYOSITIS

Transaminazların sürekli yüksekliği: Polymyositis'in gözden kaçan bir göstergesi

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Summary: *The infants or children with prolonged elevation of alanine aminotransferase (ALT) and aspartate aminotransferase (AST) are usually thought to have liver disease. A 12-year-old boy presented with the symptoms of fatigue and malaise The physical examination was normal except for height and weight. Laboratory studies revealed the abnormalities of "liver function tests". The patient underwent several investigations in order to elucidate the elevation of levels of serum transaminases and polymyositis was diagnosed. This case indicates the necessity of appropriate work-up for occult myopathy, contrary to widespread belief that the source of elevated transaminases is the liver.*

Key Words: *Aminotransferases, Child, Polymyositis*

Özet: *Çocuklarda uzun süreli alanine aminotransferase (ALT) ve aspartate aminotransferase (AST) yüksekliğinin genellikle karaciğere bağlı olduğu düşünülür. Oniki yaşında bir erkek çocuğu halsizlik ve bitkinlik belirtileri, boy ve kilo dışında normal fizik muayene bulguları ile başvurdu. Laboratuvar çalışmaları karaciğer fonksiyon testlerinin anormal olduğunu gösterdi. Transaminaz seviyelerinin yüksekliğini açıklamaya yönelik bir çok araştırma yapıldı ve polymyositis tanısı kondu. Bu olgu, transaminazlardaki yüksekliğin kaynağının karaciğer olduğu şeklindeki yaygın inanışın aksine, gizli miyopati için uygun araştırmanın yapılması gerektiğini göstermektedir.*

Anahtar Kelimeler: *Aminotransferazlar, Çocuk, Polimyozit*

Patients with polymyositis usually present with a wide variety of symptoms, signs and laboratory abnormalities that can mimic other disorders (1). Although childhood polymyositis is quite rare, early diagnosis and treatment are very important since it is a potentially fatal disease (2,3). We present a case of polymyositis with elevated serum transaminase levels initially presumed to be of liver in origin.

CASE REPORT

A 12-year-old boy presented with the symptoms of fatigue and malaise. The result of the physical examination was normal, but his height and weight were below the third percentile. Laboratory studies revealed the abnormalities of liver function tests (AST 192 U/L and ALT 140 U/L). Complete blood count, erythrocyte sedimentation rate, blood glucose, kidney function tests and albumin level were normal.

In order to elucidate the elevation of transaminase levels, the patient experienced several investigations. Hepatitis B virus markers, hepatitis A virus IgM, Epstein Barr virus viral capside antigen IgM, Cytomegalovirus IgM, Herpes simplex virus IgM, toxoplasma IgM levels and anti-HIV were studied; the results were normal.

Serum copper, ceruloplasmine and immunoglobuline levels were within normal limits and protein electrophoresis revealed a normal result. Direct antiglobulin test, antinuclear antibody and anti-dsDNA antibody were negative.

Since the diagnostic work-up failed to disclose the

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etiologic agent(s) which caused elevation of serum transaminase concentrations, the patient was re-examined carefully in order to identify new findings. Physical re-examination revealed symmetric weakness of proximal limb-girdle muscles of the lower extremities. Myopathy was suspected, and serum creatine kinase (CK) and lactate dehydrogenase (LDH) concentrations were measured; the results were markedly high (CK 7520 U/L, LDH 2220 U/L).

Electromyography demonstrated short duration polyphasic motor unit potentials and fibrillations reflecting the denervation potentials. Nerve conduction studies were normal. Muscle biopsy disclosed fiber necrosis and inflammatory infiltrates (lympocytes, plasma cells and macrophages) around the necrotic muscle fibers (Fig. 1).

Serum muscle enzyme concentrations decreased and returned to normal, and muscle weakness improved with oral prednisolone administration until the end of the first month after diagnosis.

DISCUSSION

Inflammation of muscles occurs in a number of diseases according to diverse etiologic factors. Infectious agents include viruses (retroviruses, influenza viruses A and B, adenoviruses, coxsackieviruses, Epstein-Barr viruses etc.), bacteria, fungi and parasites (4,5).

However, the most commonly encountered childhood inflammatory myopathic conditions are idiopathic and include the dermatomyositis-polymyositis complex. The annual incidence of dermatomyositis-polymyositis is about 3-5 cases per 1 million children (4). Childhood polymyositis is a rare connective tissue disease and distinguished from dermatomyositis by the absence of characteristic rash.

The diagnosis of polymyositis is often delayed because the symptoms are usually vague and nonspecific (e.g., easily suffering from fatigue and malaise) as those seen in several diseases. Serum transaminase levels are usually determined through the routine biochemical analysis in the evaluation when physical examination fail to show abnormal finding(s). The elevation of transaminase concentration is usually interpreted as evidence of liver disease and diagnostic but expensive studies are performed in order to elucidate the etiologic factor in a long period of time. Actually, these elevated results may indicate the presence of rare disorders (e.g., polymyositis, dermatomyositis) and lead to misinterpretation as in our case.

Schwarz et al. (6) reported four children with prolonged elevation of serum transaminase concentrations thought to be secondary to liver disease but all had the diagnosis of myopathy at the end of diagnostic work-up.

Another aspect of childhood-onset dermatomyositis-polymyositis is its relationship with malignancy. Although it is controversial (2,3,7,8), the number of children with association or independent occurrences of these two disorders are increasing (9,10). In addition, the presence of life-threatening complications makes this disease more serious and early diagnosis becomes more important.

In our particular experience, we conclude that serum gamma-glutamyl transpeptidase, a specific enzyme for liver, CK and LDH levels should be measured before proceeding to expensive and sophisticated laboratory investigations in patients with elevated transaminase concentrations whose physical examinations are normal.

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