The diagnostic criteria of benign monomelic amyotrophy

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To the Editor,

I read the article entitled "Benign monomelic amyotrophy in a 7-year-old girl with proximal upper limb involvement: case report" with interest[1]. I think that some of the diagnostic criteria for monomelic amyotrophy (MMA) suggested by the authors should be reviewed again.

The authors stated that no familial patient has been presented to date. However, Gücüyener et al.[2] reported two young siblings with MMA. They proposed that MMA, at least in that family, was inherited as an autosomal recessive trait. Nalini et al.[3] also reported a familial case.

The authors also stated another diagnostic criterion - that diagnosis of MMA is based on exclusion of causative local pathology and cord compression by magnetic resonance imaging.

When we consider the Hirayama disease, a contradiction arises from the point of this criterion. According to De Freitas and Nascimento[4], when MMA is restricted to the distal aspect of the upper limb, it is known as Hirayama disease. Hirayama disease has been called many different names: MMA, juvenile muscular atrophy of distal upper extremity, juvenile asymmetric segmental spinal muscular atrophy, segmental muscular atrophy of distal upper extremity with juvenile onset, and benign focal amyotrophy[5]. The 17th reference of Yılmaz's[1] article was reported by Hirayama et al. in 1959[6]. This article is also the first article about the disease now known as Hirayama disease[6]. Therefore, we may conclude that MMA and Hirayama disease are considered as the same disease by authors. On the other hand, the radiological investigations of Hirayama disease proved compressive flattening of the lower cervical cord due to forward displacement of the cervical dural sac and spinal cord induced by neck flexion. According to the Hirayama hypothesis, neck flexion causes tightening of the dura and intramedullary microcirculatory compromise with resultant ischemic nerve cell damage[6].

Nevertheless, there are some differences between the reported case and Hirayama disease: 1- The reported case had weakness on proximal muscles of the right upper extremity and scapula. Hirayama disease is characterized by muscular weakness and wasting in the distal upper extremity (the forearm and hand)[6]. Proximal muscle weakness and atrophy in Hirayama disease are extremely rare[7]. 2- The electromyography (EMG) showed abnormalities suggestive of neurogenic changes in 98.4% of 245 cases with Hirayama disease[8]. However, the EMG of the reported case was normal.

Key words: benign monomelic amyotrophy, diagnostic criteria, Hirayama disease.

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