Neurological face of familial mediterranean fever

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INTRODUCTION

Familial Mediterranean fever (FMF) is an autosomal recessive, auto-inflammatory disease. It is characterized by recurrent fever and inflammation of serous membranes. Although it occurs worldwide, it is most frequent in the population of the Mediterranean region. Familial Mediterranean fever is a multisystemic disease, and fever and abdominal pain are the main complaints of patients. Different neurological signs and symptoms have been reported in children with FMF in different case series. The exact pathophysiology of the neurological manifestation of FMF is unclear. The present study aimed to search for neurological signs and symptoms in pediatric FMF patients. We further aimed to search relationship between genetic mutation type and neurological signs and symptoms.

MATERIALS and METHOD

Medical records database from 2010 to 2020 was screened retrospectively. In total, 625 children with familial Mediterranean fever were included in the study. Neurological symptoms and associated factors were searched.

RESULTS

A total of 625 pediatric FMF patients (320 girls and 305 boys) were included in the study. The female to male ratio was calculated as 1.04. The mean age at the onset of FMF symptoms was 5.12 ± 3.51 years. Nearly half of children (44.7%) were diagnosed at an age between 5 and 10 years, and the mean age at diagnosis of FMF was calculated as 7.27 ± 3.9 years. Medical records showed that genetic analysis had been performed on 610 children, and the most common mutation type was the M694V mutation (51.5%). Medical records of patients showed that the co-existing disease was present in 49.9% of children. The neurological symptoms were present in 142 (23.5%) patients. The results revealed that parenteral consanguinity and family history of FMF does not affect the frequency of neurologic symptoms. However, in children diagnosed younger than 5 years old, neurologic manifestations are statistically less frequent than others ($P < .05$). Headache was the most common symptom, and it was accompanied by secondary other neurological disorders in 10 patients. The type of syncope was vasovagal in all cases. The origin of vertigo was central for 4 children with stroke and cerebral vasculitis. Fifty patients suffered from myalgia. However, none of them were diagnosed with protracted febrile myalgia. During follow-up, different neurologic diseases were diagnosed in 40 FMF patients and epilepsy was the most frequent disease. Epilepsy was the most frequent neurologic disease. Except for 4 children, epileptic seizures were idiopathic in 10 patients. The seizures were focal in symptomatic epilepsies. The magnetic resonance imaging (MRI) revealed pathologic neuroimaging findings in 14 patients. Acute ischemic infarct in the brain region supplied by the left middle cerebral artery was detected in stroke cases. The 2 cases with cerebral vasculitis were diagnosed by MRI findings. In terms of genetic mutations on the FMF gene, the frequency of neurological symptoms was higher in patients with E148Q mutation. However, M694V mutation was more frequent in patients who have no neurologic symptoms. The results showed that the frequency and duration of FMF attacks and compliance with colchicine treatment do not affect neurologic symptom frequency ($P > .05$). It was found that the coexistence of juvenile idiopathic arthritis is a risk factor for neurologic manifestations ($P < .05$).

CONCLUSION

Neurologic manifestations are more common in FMF patients than in the general population. This could affect the prognosis of pediatric FMF cases. As a result, during routine follow-up visits, clinicians must be suspicious of any neurologic symptoms. Prospective, case-control research could support clarifying the pathophysiology of FMF neurologic signs and symptoms.