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Joint Hypermobility Syndrome - Travails of A Young Boy

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Abstract
Hypermobility of joints was earlier considered a mild or trivial condition but recently it is reported to be a hereditary connective tissue disorder with multisystem involvement and serious morbidities. This condition can cause widespread chronic pain, dysmotility of the gastrointestinal tract, anxiety, etc. We have discussed a case of hypermobility syndrome. A normal male child, aged 25, with complaints of joint pain for 3 months. We have discussed the clinical features and management of the entity. In our case, the Beighton score was 6/9. The symptoms of arthralgia involving multiple joints (>4) were present for more than 3 months duration. To make a diagnosis of joint hypermobility syndrome our patient had two major criteria. Among the general population, approximately 3% are believed to be having joint hypermobility but due to lack of awareness, many cases are being missed and diagnosis is being delayed.

1. Introduction

The term "hypermobility syndrome" was first used by Kirke et al to describe the occurrence of rheumatic symptoms in otherwise healthy people in whom generalised joint laxity is the only abnormality identified. There is about a 4 – 13% prevalence of generalised hypermobility in the general population in the absence of any systemic disease. Higher laxity of joints is usually seen in children compared to other age groups and this laxity diminishes gradually once adolescence or adulthood is attained[1,2].

2. Case Report

A 25-year-old developmentally normal male child, presented with complaints of joint pain for 3 months, involving the shoulder, knees, and ankle. He presented to us with pain in the bilateral knee joint. He had myalgia especially over the thigh and calf muscles with difficulty in attaining squatting posture because of pain. The child had difficulty sitting in class for long hours (history of not attending school for 3 months). The child was treated with intravenous gamma globulin (IVIG) for suspicion of Kawasaki disease at 2 years. The follow-up echocardiogram (ECHO) was normal.

Initially, the child was managed with non-steroidal anti-inflammatory drugs (NSAIDs). Baseline investigations were normal. Given persistent painful joints, he was started on aspirin and referred to a rheumatologist. The rheumatologist's opinion was sort and he was started on cholecalciferol for 8 weeks. He was evaluated for SLE, anti-nuclear antibody by IFA– negative, DNA (double-stranded) antibody - negative, CRP-<1 (<5mg/L), ASO-54.4 (<200IU/L), CPK–135 (31-152 U/L), RA-IgM -< 10 (non-reactive <14).

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Neurologist consultation, suspecting polymyositis, nerve conduction study, and electromyogram was done which was normal. After psychiatric consultation to rule out somatoform disorder, he has been prescribed Clonazepam for 2 weeks because of anxiety. Orthopaedician opinion was obtained which was normal. He was on calcium, vitamin D supplements, multivitamins, and paracetamol (SOS). General examination indicated that the child was alert, well-built, and well-hydrated. The head-to-toe examination was normal and vitals were stable. Anthropometry was normal for age and no abnormality was detected upon systemic examination. Local examination of joints revealed no warmth and no tenderness, but there was a restriction of movement of both knee joints on flexion. Other joints were normal.

On Further Examination, thumb opposition is seen towards the flexor aspect of the forearm (right, left), and the knee is hyperextended beyond 90 degrees. Also, the elbow is hyper-extended beyond 90 degrees and the metacarpophalangeal joint is dorsiflexed passively to 90 degrees. Forward flexion of the body with hands lying flat on the floor and knees in the extended position is seen.

In our case, the Beighton score was 6/9. The symptoms of arthralgia involving multiple joints (>4) were present for more than 3 months duration. To make a diagnosis of joint hypermobility syndrome our patient had two major criteria.

3. Discussion

Joint hypermobility syndrome (JHS) is a disorder of connective tissue where symptoms of the musculoskeletal system are seen even in the absence of any systemic rheumatological abnormality. Joint hypermobility appears to be related to the race and sex of an individual. It usually decreases as individual ages. It is seen commonly in individuals who are healthy and without any specific complaints. JHS manifests primarily with multiple joint pains as well as joint hypermobility. To attain an accurate diagnosis of the disease, a detailed clinical history and clinical examination of the patient are needed and the disease can be confirmed by applying the Modified Beighton score and Brighton Criteria while laboratory investigations help to distinguish JHS from other diseases[1-5].

According to the patient's clinical presentation, treatment should be personalised. Pain management is to be given. Proper rest, physiotherapy, occupational therapy, activity modification, and proper physical training improve the stability and strength of muscles as well as joint proprioception. This kind of training helps in avoiding injuries and also overcoming difficulties in daily life[6-9].

Proper health education is to be given to the patients as well as to the family members of the patients suffering from JHS regarding the nature of the disease and measures to be taken to overcome the disease.

4. Conclusion

One of the common causes of unexplained joint pain is hypermobility syndrome but it is frequently misdiagnosed in primary care settings. Recognition of generalised joint hypermobility is very difficult and less than 10% of cases are being recognised or recognised among the patients who were referred to general physicians and rheumatologists. In a child who has unexplained body aches and pains, recognition of generalised hypermobility syndrome helps in the prevention of over prescription of investigations as well as drugs. The primary diagnosis of the disease helps in early and effective pain control which ultimately improves the quality of life of the patients by decreasing disruptions in their physical activities at school, work, etc. When the child attains adolescence, due to the development of resistance of periarticular tissue, symptoms usually resolve. Hence in such children, restriction of physical activity is usually not advisable unless there is the involvement of the child in strenuous activities like dance, sports, or gymnastics.

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