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Benign hypermobility syndrome: a case report

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Joint hypermobility is a connective tissue disorder commonly seen in childhood and adolescence. The Beighton hypermobility score is an easy and practical method to determine joint laxity and hypermobility. Some major and minor criteria based on symptoms and objective signs (arthralgia, back pain, spondylosis, spondylolysis/lystesis, dislocation/subluxation, scoliosis, soft tissue rheumatism, marfanoid habitus, skin hyperextensibility, ocular symptoms, varicose veins, hernia, uterine/rectal prolapse) have been determined for diagnosis of benign hypermobility syndrome.

A 13 year-old female presented with long-standing complaints of anterior knee pain which increased at night in particular, also pain and stiffness in her pain. She was of thin and tall phenotype. In her medical history, frequent ankle sprains were reported. In her physical examination determined dropping eyelids, myofascial tender points in the trapezius muscle, hyperextensible elbows, wrists, thumbs, metacarpophalangeal and knee joints and moisture on palmar aspect of the hands. Laboratory parameters and conventional radiographic observations were normal.

In benign hypermobility syndrome, clumsiness, motor retardation, poor coordination, recurrent joint sprains and limited school-based activities may be seen in the history and examination. Delayed diagnosis may lead to poor control of pain, limitations in home-life, schooling and daily activities. We recommend to patient to use joint protective methods, to make modifications in daily life, to avoid heavy physical activities and use pain-relief medication.

Keywords: Ehlers-Danlos syndrome type 3; joint instability; arthralgia

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БУЫНДАРДЫҢ ҚАТЕРСІЗ ГИПЕРБЕЛСЕНДІЛІГІНІҢ СИНДРОМЫ: КЛИНИКАЛЫҚ ЖАҒДАЙ

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Буындардың қосарласқан гипербелсенділігі балалар мен жеткіншектерде жиі кездеседі. Бейтон әдісі арқылы осы ауру түрін оңай және жылдам тексеруге болады. Сонымен қатар, клиникада буын гипербелсенділігін диагностикалау үшін келесі үлкен немесе кіші диагностикалық критерийлерді анықтауға жүгініп жатады, олар: буындағы ауырсыну сезімі, арқа тұсындағы ауырсыну сезімі, спондилез, спондилолистез, буын шығуы, сколиоз, көз симптомдары, көктамырлардың варикозды кеңеюі, жарықтар, жатыр мен тік ішектің пролапсы.

Бұл мақалада 13 жасар қыз баланың жағдайы берілген. Ол түнгі уақытта күшейетін тізе буынындағы ауру сезіміне шағымдалды. Дене бітімі бойынша қыз бала өте арық әрі бойы ұзын болып шықты. Ауру тарихында өкше буынының созылуы жарақаты жиі болған. Объективті қарағанда шынтақ, білезік, тізе және саусақ буындарының гипербелсенділігі анықталды. Зертханалық және рентген диагностикалық әдістердің нәтижелері қалыпты деңгейде болды.

Буындардың қатерсіз гипербелсенділігі синдромында жеткіншектерде ебедейсіздік, қозғалу қабілетінің жетілуінің баяулығы, координация бұзылысы, буындардың жиі шығуы байқалады. Диагнозды кеш анықтау буын қызметінің бұзылуы мен қозғалудың шектелуіне әкеледі. Біз науқастарға ауыр дене күшін шектеу үшін күнделікті өмірде қорғаныс әдістерін қолдануды ұсынамыз.

Маңызды сөздер: 3 типті Элерс–Данлос синдромы, қосарласқан буын гипербелсенділігі, артралгия.

ДОБРОКАЧЕСТВЕННЫЙ СИНДРОМ ГИПЕРМОБИЛЬНОСТИ СУСТАВОВ: КЛИНИЧЕСКИЙ СЛУЧАЙ

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Сочетанная гипермобильность суставов часто наблюдается в детском и подростковом возрасте. Методика Бейтона – самый легкий и практичный метод оценки гипермобильности для диагностики синдрома доброкачественной гипермобильности суставов. Также могут быть определены некоторые большие и малые критерии, основанные на таких симптомах и объективных признаках, как боль в суставах, боль в спине, спондилез, спондилолистез, вывих/подвывих, сколиоз, глазные симптомы, варикозное расширение вен, грыжи, пролапс матки/ прямой кишки.

Представлен случай 13-летней девочки с жалобами на боль в коленном суставе, которая усиливалась ночью. По конституции девочка очень худая и высокая. В анамнезе были зарегистрированы частые растяжения связок голеностопного сустава. При объективном осмотре определяется гипермобильность в локтевом, луче-запястном, коленном суставах и в кистях рук. Результаты лабораторных и рентгенологических обследовании были в пределах нормы.

При синдроме доброкачественной гипермобильности суставов, у больных подростках часто наблюдается неуклюжесть, двигательная отсталость, нарушение координации и частые вывихи. Позднее постановка диагноза может привести к ухудшению функции суставов и к ограничению движения. Мы рекомендуем пациентам в повседневной жизни использовать защитные методы, чтобы избежать тяжелых физических нагрузок.

Ключевые слова: синдром Элерса–Данлоса тип 3; сочетанная гипермобильность суставов; артралгия.

Introduction

Joint hypermobility or ligamentous laxity is a connective tissue disorder seen in childhood and adolescence, which is overlooked in the majority of cases. Benign hypermobility syndrome (BHS) which has become chronic is clinically defined as more generalised leading to functional losses. Musculoskeletal system symptoms emerge as joint pain, back pain, joint sprains, chronic pain syndromes and soft tissue rheumatism. Benign hypermobility syndrome is seen more often in females and in certain races.

The Beighton hypermobility score is an easy and practical method to determine joint laxity and hypermobility. For diagnosis of Benign Hypermobility Syndrome, some major and minor criteria have been determined based on symptoms and objective signs (arthralgia, back pain, spondylosis, spondylolysis/lystesis, dislocation/subluxation, scoliosis, soft tissue rheumatism, marfanoid phenotype, skin hyperextensibility, ocular symptoms, varicose veins, hernia, uterine/rectal prolapse). A high Beighton score by itself does not mean that an individual has a hypermobility syndrome. Other symptoms and signs need to also be present [1].

The Beighton score is calculated as follows:

- One point if while standing forward bending you can place palms on the ground with legs straight
- One point for each elbow that bends backwards
- One point for each knee that bends backwards
- One point for each thumb that touches the forearm when bent backwards
- One point for each little finger that bends backwards beyond 90 degrees [1-3].

Benign hypermobility syndrome is a disorder which must not be overlooked in the differential diagnosis of children and adolescents presenting with joint pain (especially in the knee). Differential diagnosis must be made thoroughly with other causes of anterior knee pain. The case is here presented of a classic BHS patient.

Case

A 13-year old white Turkish girl presented with complaints of knee and neck pain. Bilateral anterior knee pain, which increased at night, had been ongoing for a long time. There was also neck pain frequently involved. It was learned that there was a history of frequent ankle sprains. The patient had a tall, thin body structure. She did not do any sports and had a normal level of achievement at school. There wasn't any known disease neither in her past history nor in her family's history. She has been menstruated for 8 months regularly.

The physical examination determined drooping eyelids, and myofascial trigger points in the trapezium muscle in the neck. No skin lesion was determined. The palmar aspects of the hands were moist. Hyperextension was determined in both elbows, wrists, thumbs and metacarpophalangeal (MCP) joints and in both knee joints.

No sensitivity was determined in the patellar tendons, and the patella inferior and superior areas in both knees. There was no sensitivity or swelling over the tuberositas tibia. The patellar pivot-shift test was negative. Both knees had full and pain-free active movements. There was no knee pain on jumping or standing on tiptoe. No active arthritis findings were determined in all the joint examinations. The lumbar and hip examinations were normal and there was no sacro-iliac sensitivity.

The laboratory tests were within normal limits. No positive findings were determined which could be considered pathology in the direct radiographs of both knees.

As there was no clinically active arthritis in the physical

examination and the sedimentation and C-reactive protein values were within normal limits, juvenile chronic arthritis was discounted from the preliminary diagnosis. In addition, other causes of anterior knee pain (chondromalacia patella, patella bipartite, Osgood Schlatter disease, patellar misalignment, patellar tendinitis) were discounted with physical and radiological examination.

She evaluated by Beighton Hypermobility Score, and got 6/9 point. This score suggests our diagnosis, BHS with other symptoms like chronic knee and neck pain, with history of ankle sprains, hyperextensibility in multiple joints, myofascial soft tissue syndrome in her neck, without any family history.

For the trigger points in the muscles around the neck, after blockage with 1% lidocaine, stretching exercises were applied. The patient benefitted from this treatment. The patient was given a program of strengthening, stretching and proprioception exercises for the muscles around the knee and ankle to be repeated ten times per week. With recommendations for pain relief, joint protection, modifications to daily activities, and avoidance of heavy physical activities, the patient was discharged.

Discussion

BHS is known as Ehler-Danlos syndrome Type 3. In comparison with other types of Ehler-Danlos syndrome, this is benign in character. It is a connective tissue disease characterised by skin hyperextensibility, joint hypermobility, chronic pain, increased tissue fragility, ease of injury and delayed wound healing as a result of atrophic scars. Disability due to severe pain in the joints may lead to a reduction in quality of life. In addition, it may accompany various connective tissue diseases such as Marfan Syndrome and Ehlers-Danlos Syndrome. The etiopathogenesis of the disease is not fully known. Collagen abnormalities have been determined in histological and electron microscope studies [2].

BHS is not a rare disorder, hypermobility that is not associated with systemic disease occurs 4% to 13% of the population. In BHS, there may be a history of clumsiness, motor retardation, motor co-ordination disorders, overuse injuries such as frequent sprains and failure in some school-based activities. Although there are probably predisposing factors to the development of osteoarthritis in BHS, it has not been proven. Generally delays in diagnosis are caused by poor pain control and restrictions in home or school and daily activities [3,4].

Although this syndrome often presents with chronic pain (especially anterior knee pain) there may be various other symptoms with joint limitations associated with subluxation in the joint. In these cases, physiotherapy is useful [5].

The vast majority of children presenting at paediatric outpatient clinics with complaints of pain have conditions which are non-inflammatory in origin. Causes are primarily mechanical and in recent years, hypermobility or joint laxity has been determined at an increasing frequency. Several studies have stated that hypermobility is related to specific symptom complexes. However this can not be verified as not all hypermobile individuals are symptomatic or they do not develop musculoskeletal disorders later in life. It must be considered here that psychological and psychosocial factors play a role [6].

Various studies have stated that BHS leads to a tendency to osteopenia. In a study of 25 premenopausal females with benign hypermobility, the total femoral and trochanteric bone mineral densities were measured and lower t and z scores were determined compared to a control group. Hypermobility increases the risk of low bone mineral density 1.8 fold. In another study of females with benign hypermobility, when ultrasound and tomography

measurements were examined, lower t scores together with a weaker bone structure and bone strength were determined [7,8].

For anxiety, depression and increased somatosensorial complaints related to chronic pain and inadequate definition of the disease in BHS patients, psychological support should be provided [9].

Anterior knee pain in childhood and adolescence is a frequently encountered complaint. Together with BHS in the differential diagnosis, patellar misalignment, patellar tendinitis, patella bipartite, chondromalacia patella, and Osgood Schlatter disease, should be considered. It is useful to question whether or not the patient participates in challenging sports which require jumping movements. Patellar physical and radiological examination will facilitate differentiation [7-9].

In children, the site of the patellar tendon attachment to the tibia is part of the tibial apophysis. In some children, inflammation may develop at the patellar tendon attachment site after activities such as walking, running and jumping and this is known as Osgood-Schlatter disease. It is associated with overuse and disappears over time. It is classified as an osteochondrosis. There may be pain, swelling and sensitivity over the tuberositas tibia and pain increases with knee extension against resistance.

Diagnosis can be made with simple radiological tests. In the treatment, patellar band, ice packs and non-steroid anti-inflammatory drugs are used with recommendations to abstain from activities which force the knee into flexion (such as sitting cross-legged) and from challenging sporting activities [7-9].

BHS is a connective tissue disorder, often seen in females in childhood and adolescence, characterised by joint hyperlaxity, which is not yet fully understood and the majority of cases are overlooked. It is characterised by chronic joint pain, back pain, soft tissue rheumatism, posture disorders and subluxations or dislocations in the joints. Besides the musculoskeletal system, findings related to other organ involvement may be seen (eyes, skin, heart, genitourinary). Knowledge of diagnosis and simple precautions can be effective in reducing morbidity and costs. Therefore, BHS should be kept in mind for early diagnosis of paediatric patients presenting with musculoskeletal complaints. Treatment includes patient education, modification of activities, correcting lifestyle, stretching and strengthening exercises effective for the joint and manipulation. Although often seen, this is a disease which can be said to be often overlooked and which lowers quality of life.

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