Case Report

Developmental dysplasia of the hip: A special pathology

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Abstract

Developmental dysplasia of the hip (DDH) is one of the most common congenital disorders in childhood. Its diverse pathological changes require different treatments and result in different outcomes. Although many studies have been conducted on DDH, some special pathology is still unrecognized. We here presented a rare case of a one-year and eleven-month old girl with DDH; a half-free intra-articular osteocartilaginous tissue was found in her right hip joint. X-ray, computer assisted tomography (CT) and magnetic resonance imaging (MRI) were performed to evaluate the pathological changes. MRI revealed some positive findings. The patient experienced open reduction and histopathological examination of the small tissue. Through gross anatomy it is a half-free intra-articular osteocartilaginous tissue, which can fully match a fossa observed at the femoral head. Histopathological examination found that the tissue was composed of collagenous fiber and cartilage-like tissue. Interestingly, we found the expression of type I collagen according to immunohistochemical analysis, which indicated that the cartilage-like tissue was formed due to laceration of the articular cartilage. This kind of disorder should be included as one of the pathologies of DDH. The most possible origin of this tissue is the femoral head which we speculate may have been fractured before.

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Introduction

Development dysplasia of the hip (DDH), together with congenital talipes equinovarus and polydactyly/syndactyly, was known as the most common three congenital disorders in childhood. DDH was initially named as congenital dysplasia of the hip (CDH) because of the inaccurate cognition that DDH was caused by primary dysplasia of the acetabulum and laxity of the ligament at the peripheral joint. With deeper studies on the etiology, the nomenclature of this disorder was changed by the Pediatric Orthopedic Society of North American from CDH to DDH in 1992; obviously the latter can better describe the complexity of the condition and the spectrum of the associated anatomical and clinical abnormalities.

Because the therapy and prognosis of DDH are significantly related with the pathologies, it is vital to clarify the histopathological changes. Moreover DDH is a progressive disease, so the histopathological changes alter with age even for the same person. Based on the level of severity, DDH is divided into three types: simple aplasia of the acetabulum, subluxation of the femoral head and dislocation of the femoral head. Most of the cases can recover after appropriate management at early stage. Up to date, there are amount of studies focusing on the pathologic changes of DDH. The anatomic abnormalities mainly include skeletal abnormalities and soft tissue changes. In the presented case, we report a special kind of skeletal abnormalities of DDH.

Case report

A one-year and eleven-month girl was taken to us by her parents for unsteady walking for four months. The patient was born by caesarean section at 40 weeks gestation because of cephalopelvic disproportion. The infant was 4600 g and 51 cm at birth. There was neither consanguineous marriage in her pedigree nor medication, thalidomide or etretinate for example, was used during pregnancy. The symptom of unsteady walking becomes more and more apparent with age, tending to easy tumble.

CT and MRI findings

At admission, the trendelenburg test was positive and allis test negative. Anteroposterior radiography was conducted, which
showed moderately shallow, inclined and saucer-like acetabula of both sides. Femoral head ossification centers were observed at the upper and outer aspects of the acetabulum. The bones of the upper femur looked normal (Fig. 1). On computer assisted tomography (CT) images, the anteversion angle and neck-shaft angle of the right femoral head were respectively 40.0° and 149.0°. The corresponding data for the left side were 40.7° and 138.1° (Fig. 2). MRI was further performed, which showed discontinuous edge of the cartilage of the right femoral head, different from the opposite side (Fig. 3).

Surgical treatment

Open reduction with varus, derotation and shortening osteotomy of the femur and acetabuloplasty were firstly performed on the left side. During operation, the ligamentum teres was observed slightly hypertrophied and elongated, disconnected from the femoral head. The transverse acetabular ligament was thick. Then the abnormal joint laxity capsule was opened, inside of which a mass of areolar tissue was found and removed. In addition, the ligament teres was cut.

Nine months later, the right hip received the same operation. Different from the left side, an obstruction consists of cartilaginoid tissue and areolar tissue was observed at the ligaments teres, near the femoral head (Fig. 4). At the same time, there is an apparent fossa on the right femoral head. Coincidentally, we can put the cartilaginoid tissue into the fossa though they did not match perfectly (Fig. 5). Then, this obstruction was removed. Postoperatively, anteroposterior radiography was performed, which showed that both of the femoral heads were well covered.

Histopathology

HE staining and immunohistochemical staining were performed. The first antibody was anti-collagen I antibody and all steps were carried out in a moist chamber. Cartilage cells and collagenous fiber were found by HE staining (Fig. 6). According to the section of immunohistochemical staining, type I collagen was expressed at the half-free intra-articular osteocartilaginous tissue (Fig. 7). This indicates that cartilage injury happened before.

Discussion

According to Loder RT’s study, the incidence of DDH differs significantly among races due to different geographic location, from 0.06 among Africans to 76.1 among native Americans in every 1000 live births. DDH prefers to attack the left-side. As we know, the therapy of DDH aims to obtain and maintain a persistent stability of the hip joint. So it is important to reconstruct the joint and wipe out any factors that may induce future instability. This is closely associated with the pathologic changes following DDH. Therefore the pathology of DDH is a dominant factor deciding the progress and treatment protocols. Skeletal abnormalities consist of abnormalities of the acetabulum, the femoral head, the femoral neck and the pelvis. The morphology of the acetabulum is deep and a typical shape of ball-socket at embryo, which turns shallow at birth. In most cases, the acetabular bone becomes deeper with age and finally achieves full coverage of the femoral head. In this way, the optimum acetabular containment ability is formed. Nevertheless, some acetabula become shallow and inclined as infant grow up, even worse, inner-convex happens. In addition, the abnormal thickening of the acetabulum, acetabular anteversion and the aforementioned disorders make the acetabulum unable to cover the femoral head. Acetabular index (AI) is routinely used to evaluate the coverage. There are studies reporting the formation of secondary acetabulum during the progress of DDH. Fisher et al used cartilaginous acetabular index (CAI, different from BAI) to evaluate the acetabular coverage of femoral head in a retrospective study. They employed MRI to identify the cartilaginous changes in DDH patient. Their study indicates that the incomplete coverage of the acetabular cartilage is a significant factor for DDH, other than bone aspect. That is to say, the patient can achieve a good prognosis when he/she had adequate coverage by acetabular cartilage even with a bone deficiency.

After dislocation, the epiphysis of the femoral head may grow slowly and result in anatomic abnormality with time. Aseptic necrosis of the femoral head following DDH is not uncommon. An extremely rare condition of double-head of the femur has been reported as a complication of DDH. The femoral neck becomes thick and short in child with DDH. As a result, the anteversion angle and the neck-shaft angle of the femoral head change, which disturbs the spatial correspondence and force transmission between the acetabulum and the femoral head. Furthermore, mutual stimulation between the acetabulum and the femoral head decreased and finally hip development is delayed.

When unilateral DDH happens, the pelvis inclines and the spine shows compensatory curve; when DDH attacks both sides, the patient may present a waddling gait as a result of vertical pelvis, increased lumbar lordosis, and hip kyphosis. Change of soft tissue involves the glenoid labrum, articular capsule, ligaments and fat. The glenoid labrum locates at the posterosuperior direction of the labrum acetabulum, and often adheres to articular capsule or the ligament of the femoral head. When the hip was dislocated, the articular capsule would be stretched to accommodate the dislocated hip and thus becomes loose. There can be an impression when the iliopsoas passes through the front of the capsule, which will obstruct the reduction of dislocation when it forms an hourglass constriction. The inversion of the glenoid labrum and the capsule can also obstruct the reduction of dislocation. The ligamentum teres becomes elongated and hypertrophied in most dislocated cases, which is also an obstacle of reduction due to space-occupy. In addition, there are some reports demonstrated that when the acetabulum was filled with areolar tissue permeated with fat, the dislocated hip cannot be restored simply by closed reduction.

Fig. 1. X-ray of the upper femur.
It is extremely rare that there is a half-free intra-articular osteocartilaginous tissue in the dislocated hip joint in DDH, as our case. Up until now, none such cases have been reported. There are three similar cases reported by Simons et al. In their report, all patients have a cartilage-covered intra-articular obstruction, present respectively on the anterior, superior, and posterosuperior aspects of the wall of the acetabulum. The morphology of femoral head is normal. They suggested that the intra-articular osteocartilaginous obstruction possibly arose from the acetabulum and the triradiate cartilage and assumed four possible explanations for the source of the intra-articular tissue: all of them were combined with hypertrophy of adjacent tissues around the hip joint.

But in our case, we cannot find any extra abnormality through imageological examinations except for discontinued edge of the cartilage of the right femoral head at MRI image. Histologically, the intra-articular osteocartilaginous tissue is at a half-free state in the hip joint and connects the cracked ligamentum teres. Some scholars have found that type I collagen is seldom or never expressed at articular cartilage. But other researchers found type I collagen in articular cartilage after cartilage injury.

Fig. 2. CT of the hip joint.

Fig. 3. MRI of the hip joint shows discontinuous edge of the cartilage of the right femoral head (white arrow).
Fig. 4. The osteocartilaginous tissue.

Fig. 5. The fossa on the right femoral head.

Fig. 6. HE staining shows cartilage cells and collagenous fiber.

Fig. 7. Immunohistochemical staining reveals that type I collagen was expressed.
Histopathological results of our patient demonstrate a previous cartilage injury. In addition, there was a fossa which can roughly match the osteocartilaginous tissue. We think that the redundant tissue in the joint arises from the femoral head. The child did not have any history of trauma, and thus we speculate that the right femoral head has been fractured by the oblivious violence at delivering or during pregnancy or even during his growth. It can also be caused by the spontaneously tractive force during the progress of DDH.

As a new spectrum of pathology of DDH, a half-free intra-articular osteocartilaginous tissue originated from the femoral head should be classified to the morphology abnormalities of DDH. And we need to do more studies to identify the force that causes femoral head fracture in DDH.

Fund

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Appendix A. Supplementary data

Supplementary data related to this article can be found at https://doi.org/10.1016/j.cjtee.2018.02.001.

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