Esetismertetés

Aquired non-inflammatory and non-traumatic hypoplasia-dysplasia of the femoral neck

Report of two cases

Richard Nicol, Marzena Wisniewska, Kazimierz Kozlowski

Recently DiFazio et al reported four children with remarkable femoral neck deformity who had extracorporeal membrane oxygenation (ECMO) therapy. We report two patients with similar femoral neck changes who did not have ECMO treatment but have had complicated, strenuous first few weeks after birth. The first patient had been operated on twice for diaphragm repair at the age of 18 hours and 1 month. The second patient had cardiac surgery at the age of two months, with one hour of extracorporeal cardiorespiratory bypass. We agree with DiFazio et al and believe that four of their patients and both of ours have femoral neck growth cartilage injury secondary to hypoxia, vascular disturbance and coagulopathy.

femoral neck, coxa vara, coxa valga, hypoxia, coagulopathy

Szerzett nem gyulladásos, nem traumás combnyak-hypoplasia-dysplasia

DeFazio és munkatársai négy gyermek esetét ismertették a közelmúltban, akik extracorporalis membránoxigenátor kezelésben részesültek, és a femurnyak deformitásának kialakulását figyelték meg esetükben. Jelen közleményben két további olyan betegről számolunk be, akiknél hasonló elváltozátozás alakult ki, noha extracorporalis membrán-oxigenátor kezelésben nem részesültek, de életük első heteiben súlyos állapotban voltak. Az első betegnél kétszer végeztek rekeszizom-rekontsrukciót 18 órás és egyhónapos korban. A másik betegnél szívműtétet hajtottak végre kéthónapos korban, s a műtét alatt az extracorporalis keringés időtartama egy óra volt. Egyetértünk DiFazioval és munkatársaival, akik úgy gondolják, hogy mind a négy betegüknél – hasonlóan a mi betegeinkhez – a megfigyelt femurnyak-elváltozást a hypoxiás porckárosodás okozta, amely keringési zavar és coagulopathia következményeként jött létre.

femurnyak, coxa vara, coxa valga, hypoxia, coagulopathia

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Retrospective diagnosis of the cause of a skeletal abnormality may be difficult, frequently uncertain and often impossible.

In 2003 *DiFazio* et al[1] identified a remarkable pattern of proximal femoral growth arrest with coxa vara in four children with a history of neonatal treatment with ECMO (extracorporeal membrane oxygenation). They presumed that hypoxia, vascular disturbance and coagulopathy in the femoral neck vessels resulting in coxa vara were a complication of ECMO.

We describe two patients who had a history of strenuous postnatal period but without ECMO therapy. The presenting clinical symptoms were abnormal gait and discrepancy in leg length in the first few years of life. Radiographic examination

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documented isolated proximal femoral growth arrest with coxa vara or coxa valga.

We believe that the same basic process – hypoxia, vascular disturbance and coagulopathy – is the cause of the deformities in DiFazio's[1] patients and in ours as well.

Case report

Patient I.

The boy was born at term after an unremarkable pregnancy weighing 3.4 kg. Hernia on the left side of the diaphragm and ductus arteriosus were diagnosed and a repair of the former was performed at the age of 18 hours, with presented cyanosis. No ECMO was required. A re-operation of the hernia was performed a month later. He received a few doses of corticosteroid to enhance left lung growth. He had no evidence of neonatal sepsis. At the age of two he had an exploration for undescended testis but the testis was found to be absent on that side. He had the repair of the other undescended testis at the age of seven. Ophthalmologic examination revealed small angle strabismus on left side, which has led to dense amblyopia. No lower limb abnormalities were noted at birth. At preschool age a mild progressive limp was observed while walking, and later discrepancy in leg length was also noted.

The child was presented at the Orthopaedic Clinic at the age of eight, his height and weight were at around 25th centile and his head circumference at the 90th centile. There were no dysmorphic features. There was a mild discrepancy in lengths of the lower limbs, the right being 1 ½ cm longer and the right calf and thigh being slightly larger. There was a significant limitation to hip abduction in flexion, this being 30 degrees.

The skeletal survey documented significant shortening of the femoral necks with varus deformity. The femoral neck growth cartilage was narrowed and had an irregular outline *(Figure 1)*. The changes were more marked on the left side. The remaining skeletal survey was normal.

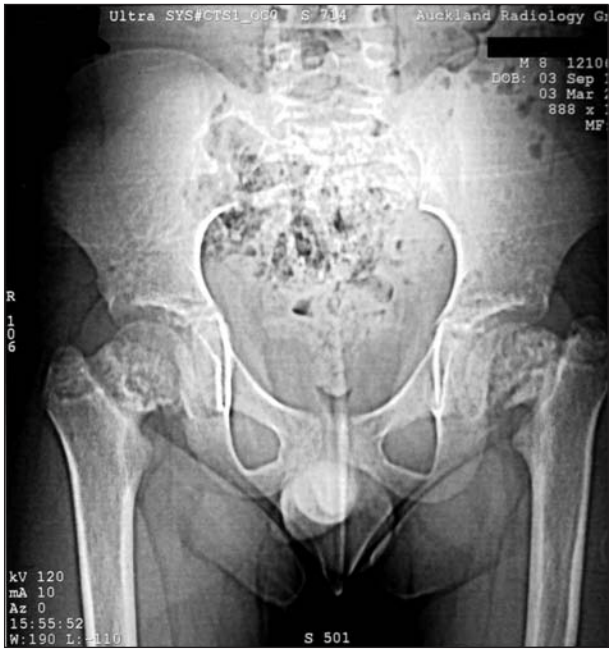

Figure 1. **Patient 1. Eight year old boy. Bilateral coxa vara more outstanding on the left side.** Hypoplastic/dysplastic femoral necks. The bony structure of both femoral necks and capital femoral epiphysis is abnormal but the femoral heads are well formed

Patient II.

The four year-three month-old girl was referred for consultation due to right lower limb shortening. She was born from a second twin pregnancy of non related parents. The father was 27 and the mother 26 years old at the time of delivery. The family history was uneventful. Birth occurred by cesarean section at 38 weeks of the gestation. Birth weight was 2100 g, length of 46 cm, head circumference of 32 cm. Apgar was 8-9. Because of recurrent respiratory distress due to multpile congenital heart disease (ASD, VSD, TI), complicated with recurrent pneumonia cardiac operation was carried out at the age of two months. The procedure with extracorporeal cardiorespiratory bypass lasted about one hour. She had no evidence of neonatal sepsis and no ECMO was utilized. At the time of examination her height was 96 cm (3 lst centile), weight 13.5 kg (3-10 centile). She showed a slightly dysmorphic face – narrow palpebral fissures, epicanthus and broad nasal bridge. She had leg length discrepancy of 2 cm (left > right). Her developmental milestones were minimally delayed as compared with the twin sister who developed normally. The routine blood and urine examinations, the karyotype 46XX and the brain MR were all normal.

The skeletal survey documented bilateral, severe

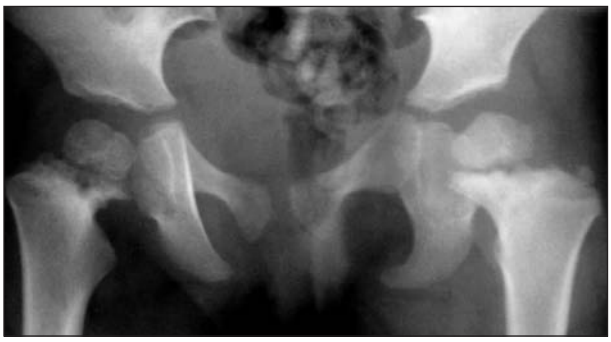

Figure 2. **Patient 2. Four year and three month-old girl. Coxa valga on the left side.** Hypoplastic/dysplastic femoral necks. The capital femoral epiphyses are grossly normal. Note the similarity between the left femoral neck of the first patient and the right femoral neck of the second

shortening of the femoral necks. The femoral neck growth cartilage was narrowed and irregular in outline. There was coxa valga on the left side. The femoral capital epiphyses and hip joint spaces were normal *(Figure 2)*. No other skeletal abnormality could be seen.

Discussion

The patients of *DiFazio* et al[1] required ECMO because of persistent hypoxia and respiratory distress. Subsequently they were doing well. None of their patients had evidence of neonatal sepsis. In one patient hip abnormality was recognised at the age of eight months during evaluation of tight heel cords. In the remaining patients hip malformation was detected at the age of 17 months, three years and seven years because of refusal to walk, pain, limp and leg length discrepancy.

ECMO needs heparinization of the circuit because of tendency to clot formation[2, 3]. As well as *DiFazio et al*[1] we also could not demonstrate the involvement of the metaphyseal cartilage in other parts of the skeleton, so we may presume that the proximal femoral metaphyseal cartilage with its particularly vulnerable blood supply is most susceptible to occlusion following hypoxia, vascular disturbance and clot formation[4].

Our first patient's postnatal course was complicated with two surgical procedures because of recurrent diaphragmatic hernia. The other patient had a strenuous postnatal course complicated by recurrent pneumonia and cardiac operation with extracorporeal cardiorespiratory bypass lasting one hour. No clotting abnormalities were noted before, during and after the operation.

The first patient presented with a limp for a number of years with the left limb shorter by 2 cm. It is surprising that he did not complain of hip pain on the severely affected left side. The limp of the second patient was noted at the age of three. She also did not complain of pain inspite of severe metaphyseal involvement on the right side.

Radiographs of our patients show severe shortening and unusual deformity of the femoral necks. The growth cartilage is narrowed and of irregular outline. Bilateral coxa vara can be seen in our first patient, and coxa valga on the left side in the second one. The left femoral neck of the first patient and the right of the second is deformed to such an extent that a fracture through the femoral neck is highly probable in case of stress or even of mild trauma.

The differential diagnosis is with other hip disorders characterised by femoral neck shortening with subsequent coxa vara or valga. The remaining skeletal survey was normal in both cases. Although *DiFazio*[1] did not perform the skeletal surveys in his patients, there were no clinical signs or symptoms suggesting other metaphyses and growth cartilage being affected. Similar radiographic appearances may be present in osteomyelitis. No clinical signs or symptoms of osteomyelitis were detected neither in *DiFazio's*[1] series nor in our patients. Neonatal osteomyelitis is often with slight or no signs of toxemia or pyrexia and of systemic disturbance[5]. However such severe femoral neck lesion without hip joint cartilage and epiphyseal changes would be unusual indeed. Moreover, bilateral severe asymptomatic osteomyelitis of the proximal femoral metaphyses without any other inflammatory locus would be an atypical finding[6, 7]. The lack of history of trauma, and the absence of any traumatic lesion in the skeletal survey excludes posttraumatic coxa vara or valga[8].

Idiopathic coxa vara is characterized by femoral neck angle close to 90 degrees and the femoral metaphysis is only slightly irregular. The femoral neck may be affected in severe Perthes disease but the femoral head is predominantly affected in this case. Finally, localised osteochondritis have been described in many parts of the skeleton but not as an isolated anomaly of the femoral neck[9].

Conclusion

The most likely explanation of the hip changes in our patients is – as suspected by *DiFazio* et al[1] – damage to the proximal femoral growth cartilage due to hypoxia, vascular damage and coagulopathy. These insults to growth cartilage of the femoral neck present later in life as limp, pain and shortening of the leg. Radiological examinations reveal short hypoplastic/dysplastic femoral neck, distorted growth cartilage with irregular outlines and deformity like coxa vara or coxa valga. The possibility of femoral neck fracture after a fall or minor trauma should be considered. This type of change may be expected in newborns with severe postnatal respiratory distress requiring intensive care therapy, particularly extracorporeal cardiopulmonary bypass.

The history of our two cases implies that proximal femoral growth cartilage arrest may develop after a severe hypoxic incident in the neonatal period. It also widens the spectrum of hip disturbances, which may develop in patients with a stressful first few weeks of life.

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