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HIP JOINT IN SYSTEMIC BONE DISEASE

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Abstract

It is generally accepted that X-ray examination of pelvis together with that of spine, hand and foot are of major importance in the diagnosis of systemic bone disease.

Six cases of systemic bone disease are described which show typical X-ray findings of hip joint and pelvis (achondroplasia, spondyloepiphyseal dysplasia congenita, Morquio's disease, spondyloepiphyseal dysplasia tarda, multiple epiphyseal dysplasia tarda and dysplasia epiphysealis capitis femoris). Characteristic X-ray findings of epiphyseal disturbance is the deformity of femoral capital epiphysis and that of physeal disturbance is poorly developed iliac wing.

Difficulties of the diagnosis in mild form of epiphyseal disturbances like "Chondrodystrophischer Habitus", in precocious arthritis of trada type of epiphyseal disturbance and in dysplasia epiphysealis capitis femoris which must be always taken into considerations in the diagnosis of Legg-Perthes' disease of bilateral hip involvement were discussed.

INTRODUCTION

Attention has been paid in recent years to the hip joint in systemic bone disease. Many of the more common diseases such as congenital dislocation of hip joint and Legg-Perthes' disease might be mistaken and therefore the analysis of the hip joint X-ray findings in systemic bone disease will be of a considerable significance. Frequent occasions to examine hip X-ray are due to the following reasons: (1) It is more difficult to evaluate hip deformity clinically. (2) Congenital dislocation of hip joint was taken into considerations in the past most frequently as the cause of gait disorders among the children and examination of hip joint X-ray was always necessary. On the other hand, it is apparent that routine X-ray examination of hip must be avoided owing to the

exposure of reproductive organ to X-ray and moreover we cannot diagnose the systemic bone disease only with hip joint X-ray. Neverthless the importance of hip joint X-ray cannot be overemhapsized.

Six cases that we show are the typical systemic bone disease selected from 28 cases and characteristics of the disease are discussed. (Table 1)

TABLE 1

Rubin's classification of our cases

Spondyloepiphyseal dysplasia congenita	4
Morquio's disease	2
Pseudoachondroplastic form of Spondyloepiphyseal dysplasia congenita	1
Spondyloepiphyseal dysplasia tarda	4
Multiple epiphyseal dysplasia tarda	8
Diastrophic dwarfism	1
Achondroplasia congenita	3
Achondroplasia tarda (Hypochondroplasia)	2
Osteopetrosis tarda	1
Dysplasia epiphysealis capitis femoris	2
Total	28

CASE REPORT

Case 1: 9 years, female, Achondroplasia (Fig. 2.)

Development of iliac wing is poor. Transverse diameter of inner pelvic inlet is longer than its depth. The inner pelvic contour gives a champagne glass appearance (Fig. 1.). The sacrum in narrow and articulate low on the ilium. The capital femoral epiphysis is normal in shape. The femoral neck is short in length.

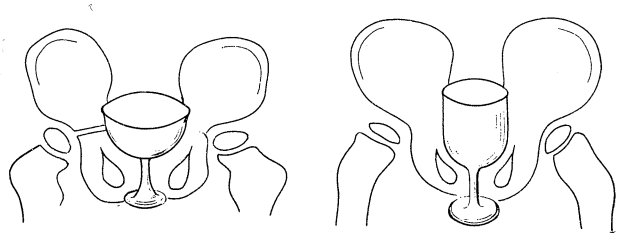

Fig. 1. The inner pelvic contour represented different types of glasses.
a. Achondroplasia: champagne
b. Spondyloepiphyseal dysplasia: wine

Case 2: 2 years, female, Spondyloepiphyseal dysplasia congenita (Fig. 3.)

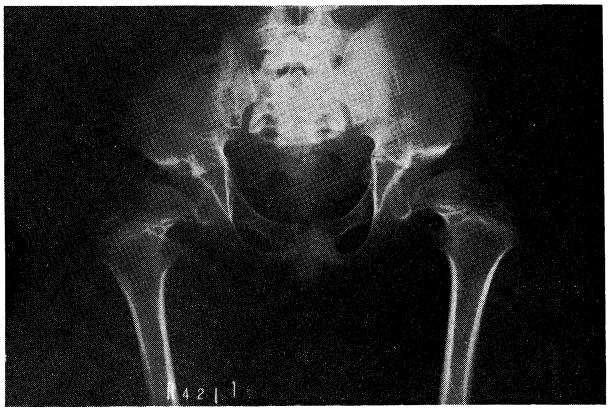

Fig. 2. Case 1: 9 y. female, achondroplasia.
Poorly developed iliac wing, champagne glass appearance of inner pelvic contour. Narrow sacrum articulating low on the ilium.

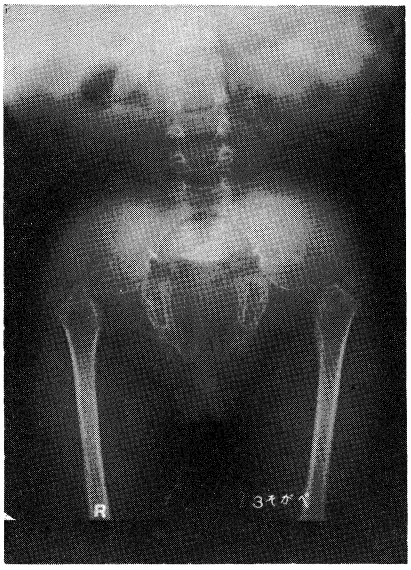

Fig. 3. Case 2: 2 y. female, spondyloepiphyseal dysplasia congenita.
Delayed appearance of capital femoral epiphysis and coxa vara. Poorly developed iliac wing.

Delayed appearance of the capital femoral epiphysis and coxa vara mimicks the dislocated hip joint and arthrographic examination will be necessary. Poorly developed iliac wing and small acetabular angle are also characteristic.

Case 3: 9 years, female, Morquio's disease (Fig. 4.)

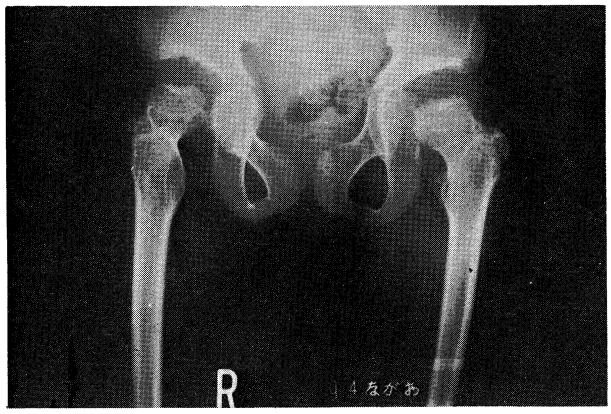

Fig. 4. Case 3: 9 y. female, Morquio's disease.
Irregular capital femoral epiphysis. Flaring of iliac crest with acetabular protrusion, wine glass appearance of inner pelvic contour.

The capital femoral epiphysis is irregular in its contour. The femoral neck shows coxa valga. Flaring of iliac crest with acetabular protrusion gives a wine glass appearance of the inner pelvic contour (Fig. 1.). To make the diagnosis of Morquio's disease the increase of the urinary excretion of keratosulphate must be ascertained in this case. But the X-ray findings can give us fairly reliable information about the differential diagnosis from spondyloepiphyseal dysplasia congenita.

Case 4: 27 years, male, Spondyloepiphyseal dysplasia tarda (Fig. 5.)

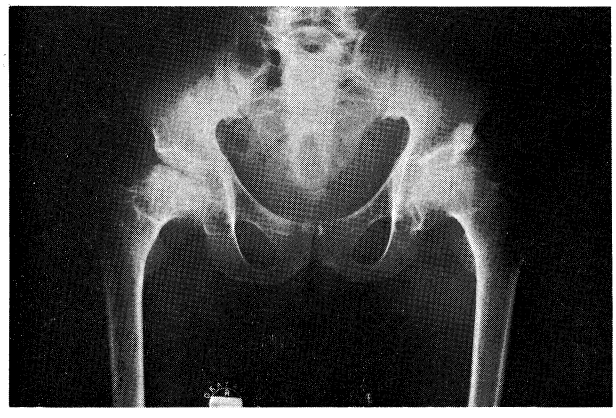

Fig. 5. Case 4: 27 y. male, spndyloepiphyseal dysplasia tarda.
Normal pelvis. Deformity of femoral head (precocious arthritis).

The pelvis is normal. Coxa vara and femoral head deformity is observed. Surface of the femoral head is irregular. Platyspondyly (flattening of vertebral body) is another characteristic figure.

Case 5: 19 years. male, Multiple epiphyseal dysplasia tarda (Fig. 6.)

The pelvis is normal. Acetabulum developed poorly. The femoral head deformed severely. He has no platyspondyly.

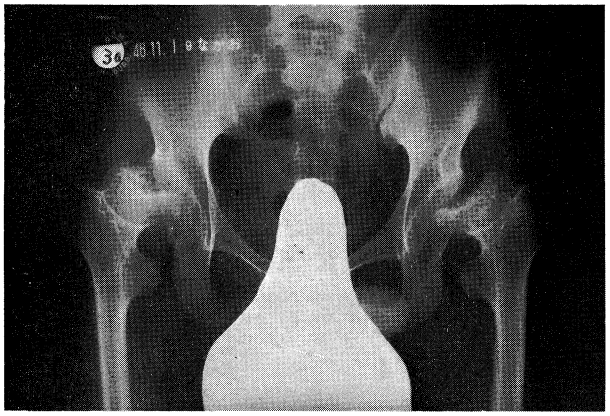

Fig. 6. Case 5: 19 y. male, multiple epiphyseal dysplasia tarda. Normal pelvis. Deformed femoral head.

Case 6: 4 years, male, Dysplasia epiphysealis capitis femoris (Fig. 7.)

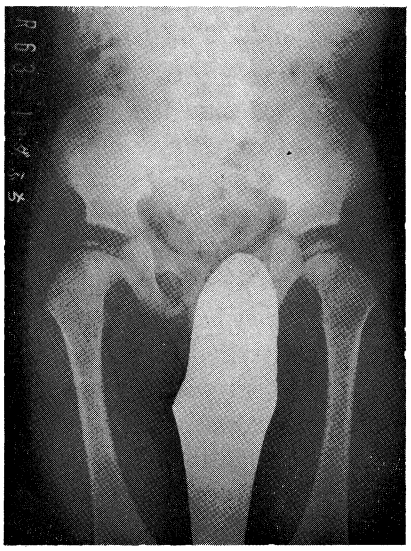

Fig. 7. Case 6: 4 y. male, dyspla epiphysealis capitis femoris. Deformed capital femoral epiphysis without metaphyseal change of both hip joint.

The capital femoral epiphysis is deformed. There is no metaphyseal change. Most important difference from Legg-Perthes' disease is good prognosis without rest deformity.

DISCUSSION

There exists a considerable literature on the X-ray analysis of bone in systemic bone disease, but little work has been done to the X-ray analysis of hip joint in detail.[1,6] X-ray changes of hip joint are observed frequently together with that of spine, hand and foot. Rubin[5] has explained the fundamental differences of the hip involvement in systemic bone disease in his clear-cut classification. Mau's classification is not clear-cut, in other words not practical, but the borderline cases could be understood more easily by his systemic analysis. Pseudoachondroplastic form in Rubin's classification might correspond to the intermediate form[5] of "Dysostosis epiphysarea" and "Dysostosis metaphysarea" in Mau's[3] classification. The word "Dysostosis" should be understood as "dysplasia" (disturbance in form) in Rubin's literature.[5] We use "epiphyseal disturbance" instead of "Dysostosis epiphysarea", "physeal disturbance" instead of "Dysostosis metaphysarea" in the following paragraph. Mild form among the tarda types by Rubin[5] could be regarded as the intermediate type between normal and pathologic constitution by Mau.[3] It is necessary not only to diagnose systemic bone disease but also to differentiate epiphyseal and physeal disturbance in the examination of mild type or intermediate type. Shortening of the limb, especially that of humerus and femur is characteristic feature in physeal disturbance, wheras the striking findings in epiphyseal disturbance are the changes in hip joint and spine.[3] X-ray findings of the hand and spine could be decisive in the diagnosis. As for the findings of pelvis the longitudinal shortening of ilia owing to undergrowth at the iliac base is characteristic of physeal disturbance together with the squared-off appearance of poorly developed iliac wing[5]. Sharp sacrosciatic notch gives an impression of champagne glass appearance (Fig. 1.) in inner contour. In epiphyseal disturbance the pelvis is sometimes normal or the flaring of iliac crests, opening of the sacrosciatic notches and acetabular portrusion are sometimes prominent. The inner contour of the pelvis shows a wine glass appearance (Fig. 1.) in this case. The capital femoral epiphysis in epiphyseal disturbance is irregular in shape. Its shape is round and its surface is smooth unless it is deformed secondarily after growth. (Table 2)

TABLE 2

X-ray changes of hip joint in systemic bone disease

	Epiphyseal change	Aceta-bulum	Ilium	Coxa vara	Others
Spondyloepiphyseal dysplasia congenita	moderate severe	protrusion	flared	frequent	platyspondylies (tongue-shaped)
Pseudoachondroplastic form of spondyloepiphyseal dysplasia congenita	moderate severe		flared		dwarf, platyspondylies, brachydactylies
Spondyloepiphyseal dysplasia tarda	moderate severe				precocious arthritis
Multiple epiphyseal dysplasia congenita	stippled		stippled		contructure, may be fatal
Pseudoachondroplastic form of multiple epiphyseal dysplasia	moderate severe				dwarf, brachydactylies
Multiple epiphyseal dysplasia tarda	small and flat				precocious arthritis
Diastrophic dwarfism	severe	dysplasia			hip dislocation, contructure
Achondroplasia			squared off		dwarf, short thick tubular bone
Morquio's disease	severe	enlarged	flared		platyspondylies (pear-shaped) mucopoly-saccharidosis IV

Good prognosis of Legg-Perthes' disease with localized epiphyseal change is generally accepted. Factors such as hormonal conditions, mechanical overloading, circulatory disturbances and most important of all, constitutional or dysostotic factor represented by skeletal age should be taken into considerations in evaluating its prognosis. Mau[3] suggested that early bilateral onset with microepiphysis, relatively large major trochanter and delayed skeletal age is characteristic findings of Legg-Perthes' disease with dysostotic character. It is probably the same disease as Kraup has reported under the name of dysplasia epiphysealis capitis femoris in its good prognosis. (Fig. 6)

Properties characterize the disease are summarized by Meyer[4] as follows :

1) This disease is observed below the age of 4 years and mainly among the boys.
2) Patient has no symtoms or mild complaint.
3) Bilateral affection of hip joint is frequent.

4) The ossification of capital femoral epiphysis is delayed and the epiphysis does not appear until about 2 years of age, when at last it appears the small epiphyseal nucleus will be of a pathological appearance.
5) It takes about 3 years to return to normal femoral epiphysis and the natural history of this disease is shorter than that of Legg-Perthes' disease.

The presence of dwarfism, deformities of thorax and shortening of the limb are helpful to diagnose the systemic bone disease, especially in physeal disturbance. But it is very difficult to diagnose the mild form of epiphyseal disturbance like "chondrodystrophischer Habitus" (achondroplastic constitution). One can assume that the deformity of mild type is influenced secondarily by the mechanical factors. Tarda type (Rubin) characteristic in its delayed onset of the symptoms would be classified in this category.

Osteoarthritic changes of the hip joint in systemic bone disease are seen frequently together with spine, hand and foot deformities. Tarda type must be always ruled out in diagnosing the osteoarthritis of hip. Multiple or bilateral joint lesion, relatively younger age, small statue and familial appearance call our attention to systemic bone disease and X-ray examination of the spine, especially thoraco-lumbar region, pelvis, hand and foot are necessary. Taking the dysostotic factor into consideration in the diagnosis of osteoarthritis of hip joint might be of practical importance not only to decide the diagnosis but also to evaluate the prognosis.

Acknowledgement

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