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Original Research Paper

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LEGG-CALVE-PERTHES DISEASE: A MUST KNOW FOR PEDIATRICIANS

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ABSTRACT

Legg-Calve-Perthes disease is a type of avascular necrosis of femoral head seen predominantly in male children especially between 4 to 10 years of age. This is a rare entity with an incidence varying between 0.4 in east India to 4.4 per 1,00,000 in west India. Therefore, it is fairly rare for a paediatrician to encounter this condition. It must be kept as a differential in a child with abnormal gait or limping gait and should not be missed. Its treatment in earlier stages is promising. We present a case of a nine year old male child with Legg-Calve-Perthes disease.

KEYWORDS :

INTRODUCTION

Perthes' disease is an idiopathic osteonecrosis of a developmental hip in childhood. The process is self limited, renders the femoral head and hip joint deformed with a limited range of motion and future risk for secondary arthiritic changes. The child's age and extent of head involvement determine the prognosis.[1]

CASE REPORT

A 9 year old male child presented with history of limping gait for the last two months. He also complained of pain at lateral aspect of upper one third of right thigh. There was no history of weakness in the limb. No history of fever. No significant past history. On Examination, his general physical examination, had an abnormal gait. His vitals, central nervous system examination was normal. On musculoskeletal examination, abduction and internal rotation at the right hip was painful. His X-Ray of right hip joint confirmed the diagnosis of Perthes disease showing flattened and sclerotic epiphysis in stage 3/4. The child was then referred to an orthopaedician.

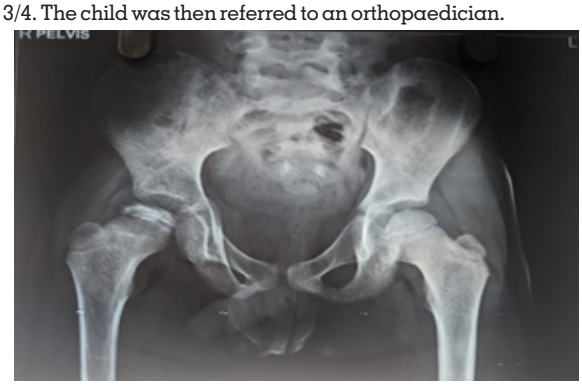

Fig 1 An Xray Radiograph Of The Patient Showing Sclerotic Femoral Epiphysis.

DISCUSSION

Perthes disease generates a susceptibility of the femoral head to change shape due to forces acting across the joint. These shape changes alter the way the joint moves, which can cause lifelong pain, functional limitations and accelerate the development of osteoarthiritis. The precise etiology of Perthes disease is not known, however it is clear that a vascular insult is the final precipitating episode leading to disease onset.[2]There are few studies that support familial predisposition. The clinical presentation may be insidious with pain and limp in an otherwise healthy child. The child may present with knee pain, stiffness in hip, groin, thigh or knee. The involvement is frequently unilateral. Perthes disease is self-limiting in that the blood supply to the femoral epiphysis restores over a period of two to four years. During this period, the necrotic avascular epiphysis fragments and is then resorbed and completely replaced by new bone. This sequence of healing can be identified clearly on plain radiographs and on the basis of these radiographic appearances the disease can be quite reliably divided into avascular necrosis, fragmentation, regeneration and healed stages. The stages of avascular necrosis, fragmentation and regeneration can be further divided into early and late parts of the respective stage. In many children, the epiphysis heals well with the femoral head remaining spherical; in these cases, the affected hip should then function normally throughout life. However, in some cases the femoral head is irreversibly deformed during the healing process resulting in premature degenerative arthritis sets in these hips. Treatment focuses on the prevention of femoral head collapse, restoring the range of motion and improving the functional recovery (absence of pain, amount of usual daily activity and sport related activity) of the children. There is no consensus for the best management approach in paediatric orthopaedic community.[3]

Early treatment during the disease aims to prevent the femoral head deformity since it has been shown that if the femoral head remains spherical there is a very good chance of avoiding degenerative arthritis in adult life. This is achieved by containment which attempts to keep the anterior and lateral region of the avascular epiphysis within the acetabulum. thereby preventing the femoral head deformity caused by stress transmitted across the acetabular rim. The are surgical and non-surgical methods to achieve containment. Treatment to achieve containment is not indicated after the disease has progressed to the late fragmentation stage. Treatment may be directed towards reduction of forces across the hip hopefully to delay the onset of arthritis or treatment may be primarily to alleviate pain. Of late, there has been a great deal of interest in the use of bisphosphonates for treatment of avascular necrosis of the femoral head due to causes other than Perthes disease.[4]In our case the femoral head was deformed and thus it was too late for containment procedures.

CONCLUSION

Since the aim of Perthes treatment is to prevent femoral head deformity and thereby retain the sphericity of the femoral head, it follows that treatment needs to be instituted before irreversible femoral head deformity occurs. Studies on the natural history of Perthes disease suggest that if untreated, femoral head deformity tends to occur either in the latter part of the fragmentation stage or very early in the regeneration

stage.[5] This implies that the timing of containment (irrespective of the containment method) is crucial to success; containment should be undertaken before the latter stage of fragmentation. This fact makes it very important for this disease to be diagnosed at the earliest and not to be missed as the child may present to paediatrician initially with a vague pain in the limb.

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