CASE REPORTS

IDIOPATHIC JUVENILE OSTEOPOROSIS - A RARE CAUSE OF OSTEOPOROSIS REPORTED IN A FIVE YEAR OLD BOY

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Abstract

Idiopathic Juvenile osteoporosis (IJO) is a rare primary bone demineralization disorder that presents in childhood. Because of the difficulty in diagnosis, it is unfamiliar to most pediatricians and there is a long list of differential diagnosis. We report a five year old boy who presented with generalized osteoporosis. Diagnosis of IJO was made by excluding other common causes of childhood osteoporosis.

Key words: Childhood osteoporosis, idiopathic Juvenile osteoporosis.

Introduction

Osteoporosis in childhood is uncommon and may be secondary to a spectrum of diverse conditions. When such causes have been excluded, the cause is either a congenital disease (osteogenesis imperfecta) or a disease of unknown etiology called as IJO (1). IJO is characterized by prepubertal onset and spontaneous remission with progression of puberty. This case illustrates the occurrence of IJO which was considered after excluding other causes of childhood osteoporosis.

Case Report

A 5-year-old male child born to a non consanguineous married couple presented with complaints of pain in back and lower limb and inability to sit up from bed, which progress to inability to stand and walk since 1 year. On examination, weight was 13.8 kg, height was 118cm. There was reduced muscle bulk of both lower limbs. The tone and deep tendon reflexes were normal. Examination of the other systems was normal. X-ray of the spine and limbs showed generalized osteoporosis (marked reduction in the normal trabecular pattern of bones). Lateral radiograph of spine showed a “cod fish” appearance (fig. 1) with biconcave vertebral bodies

Fig 1: X-ray of the spine showing “cod fish” appearance (biconcave vertebral bodies with denser end plates and increased intervertebral space)
osteoporosis, we arrived at a diagnosis of IJO.

The exact pathogenesis of this disorder is not known but available evidence points towards disturbed bone remodeling which predominantly affects surfaces that are in contact with the marrow cavity and results in a very low bone formation rate and decreased cancellous bone volume. (5) The mean age of onset is 7 years (range 1-13 years) with no sex difference. The main presenting symptoms are long bone fractures, pain in back and difficulty or inability in walking. (6) Typical radiograph shows compression of vertebral bodies and metaphysis of long bones. Bone mineral density shows strikingly low values. In majority of cases, the disease remits spontaneously during or after puberty. Although spontaneous remission is the rule, restricting activities of the currently affected children is necessary to protect from permanent deformities of the spine and long bones. Many drugs like calcitriol, biphosphonates (7), fluorides and calcitonin have been used with equivocal results (4). Due to insufficient experience, no treatment can be advocated at the moment other than activity restriction till natural remission.

References