

Juvenile osteoporosis in a 5-year-old girl

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Abstract

Idiopathic juvenile osteoporosis (IJO) is a term used to describe a primary osteoporosis of unknown etiology in prepubertal children. It is rarely described in the literature and treatment modalities vary with spontaneous remission also being reported at the time of puberty. We report a 5-year-old girl with IJO who had spinal deformities and was successfully treated with oral alendronate.

Key words: Alendronate, children, juvenile osteoporosis

INTRODUCTION

Idiopathic juvenile osteoporosis (IJO) is a term used to describe a primary osteoporosis of unknown etiology. It is characterized by prepubertal onset, and spontaneous remission with the onset of puberty.^[1] We describe a case of IJO in a 5-year-old girl who was successfully treated with oral alendronate and oral calcium carbonate.

CASE REPORT

A 5-year-old girl born of nonconsanguineous marriage presented with nonprogressive chest deformity since birth. A physician had done X-ray of spine that showed osteopenic bones with cod-fish appearance of spine and kyphosis without signs of rickets and had referred the patient to us. There were no fractures. Child had an elder sister of 6½ years of age who was normal. The patient had no other deformities and milestones were normal. She was on regular balanced diet. On examination, she had kyphosis and hunchback. Other systems were normal. Investigations showed serum calcium of 9.8 mg/dL, phosphorus of 4.8 mg/dL, and alkaline phosphatase of 1431 IU/L. Her serial calcium, phosphorus, and alkaline phosphatase are depicted in Table 1. Her urine calcium/creatinine was 0.46. Blood gas analysis showed no acidosis (pH 7.36, bicarbonate = 21.7) with ultrasound kidneys being normal and creatinine of 0.5 mg/dL. Bone mineral density (BMD) showed severe osteoporosis [Table 2]. She was treated with oral calcium carbonate (50 mg/kg/day) and oral alendronate (0.5 mg/kg/day) for 9 months. Her 25 hydroxy Vitamin D levels at end of 6 months of therapy were 24 ng/mL (normal = 9-37.6 ng/mL) and serum parathyroid hormones

were 27.9 pg/mL (normal = 12-95 pg/mL). Her deformity and BMD [Table 2] gradually improved and she had no adverse effects to alendronate.

DISCUSSION

Osteoporosis in pediatric population usually occurs secondary to definite causes such as Cushing's syndrome, celiac disease, immobilization, osteogenesis imperfecta, homocystinuria, rickets, and endocrinopathies.^[1] However, certain cases of obscure etiology have been identified by exclusion of the above diagnoses and labeled as IJO. Dent was the first to describe its cardinal features as an onset before puberty, multiple fractures, pain in back and extremities, and radiological evidence of osteoporotic new bone and metaphyseal compression fractures.^[2] In our patient, renal, metabolic causes, rickets, and recurrent fractures had been excluded and she had no other systemic illness. In addition, BMD was performed which was suggestive of osteoporosis. Hence, a diagnosis of IJO was formulated.

The age of onset of IJO in various reports ranges from 1 to 13 years (ours was 5 years) and no sex predilection although some reports suggest a younger onset in girls.^[1,2]

In our case, the plasma alkaline phosphatase level was raised at the time of diagnosis but quickly improved on therapy. This is in accordance with the findings of Jowsey and Johnson^[3] but contradict several others^[1,2] including a recent Indian study.^[4] There is no consensus in the literature regarding this finding and cannot be explained by the most common hypothesis for pathogenesis which points toward an abnormality of bone formation at

Table 1: Serial biochemistries

Serum biochemistries	Aug 2006	Oct 2006	Jan 2007	May 2007
Calcium (mg/dL)	9.8	10.2	9.3	9.9
Phosphorus (mg/dL)	4.8	4.6	3.9	5.4
Alkaline phosphatase (IU/L)	1431	604	748	657
Urine calcium/creatinine	0.46	0.48	0.21	0.2
Treatment	Alendronate calcium	Alendronate calcium	Alendronate calcium	Alendronate calcium

Table 2: Bone mineral density changes

Dates	BMD, g/cm ²		Z score
	Femur	Lumbar spine	
Sept 2006 (Pretreatment)	0.514	0.308	-3.4
May 2007 (Posttreatment)	0.603	0.410	-2.5

BMD: Bone mineral density

surfaces that are in direct contact with the marrow cavity and hence, a decrease in cancellous bone mass.^[5] However, detailed understanding of pathogenesis and the natural history of the disease is needed to ascertain the exact mechanism.

There is currently no consensus regarding the requirement, nature, and duration of treatment. A variety of drugs including calcitriol, bisphosphonates, fluorides, calcitonin, vitamin D, and their combinations have been used with varying success.^[6] Bisphosphonates have been proven effective in increasing effective bone mass and reducing fracture risk in adults with acceptable safety profiles. Although the natural history of the disease points to a complete remission around the age of puberty, there is a risk of major spine and long bone deformities. Since our patient had a significantly decreased BMD, spinal deformity, and radiological changes, we decided in favor of bisphosphonate therapy.

In one partially randomized, open trial in patients with OI, oral alendronate and IV pamidronate appeared equally effective in increasing BMD,^[7] hence the decision to use

oral therapy. Alendronate caused no adverse effects in our patient and led to clinical and radiological improvement. However, we conclude that bisphosphonates may be safe for use in IJO.

REFERENCES

1. Smith R. Idiopathic osteoporosis in the young. *J Bone Joint Surg Br* 1980;62-B: 417-27.
2. Dent CE. Osteoporosis in childhood. *Postgrad Med J* 1977;53:450-7.
3. Jowsey J, Johnson KA. Juvenile osteoporosis: Bone findings in seven patients. *J Pediatr* 1972;81:511-7.
4. Kulkarni ML, Keshavamurthy KS. Juvenile idiopathic osteoporosis. *Indian Pediatr* 2004;41:737-40.
5. Rausch F, Travers R, Norman ME, Taylor A, Parfitt AM, Glorieux FH. The bone formation defect in idiopathic juvenile osteoporosis is surface specific. *Bone* 2002;31:85-9.
6. Krassas GE. Idiopathic juvenile osteoporosis. *Ann N Y Acad Sci* 2000;900:409-12.
7. DiMeglio LA, Peacock M. Two year clinical trial of oral alendronate versus intravenous pamidronate in children with osteogenesis imperfect. *J Bone Miner Res* 2006;21:132-40.

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