CEREBRAL GIGANTISM (SOTOS' SYNDROME)

(A Case Report)

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MJAFI 1996; 52 : 200-201

KEY WORDS : Cerebral gigantism; Sotos' syndrome.

Introduction

First described by Sotos et al in 1964, cerebral gigantism or Sotos' syndrome is a disorder of growth characterized by increased rate of growth in the early years with advanced height, weight and bone age [1]. Patients have coarse, acromegaloïd features with a large, dolichocephalic skull, antimongoloid slant of eyes and prominent jaw. Mental subnormality, clumsiness, behavioural problems and scholastic backwardness are also associated. Hormonal studies are normal. The disease is non-progressive and most patients stabilize by 5-7 years [1,2]. Only 4 cases have been reported in Indian literature [3,4]. We report a case of cerebral gigantism which has been followed up from 2 years of age to 7 years of age.

Case report

A 7-year-old male child, born of a non-consanguineous marriage, with no significant perinatal history and birth weight of 3.8 kg, first presented at 2 years of age with increased physical growth and delay in motor and mental milestones noticed since early infancy. At that time his height was 104 cm and weight 24 kg. Head circumference was 56 cm. He had coarse acromegaloïd features with a dolichocephalic skull, anti-mongoloid slant of eyes, prominent mandible and thick lips. The palate was high-arched and he had 20 teeth. The hands and feet were large but did not show distal tapering or tufting. His development quotient was 1½ years. He had started sitting at 11 months and walking without support at 20 months. His speech was bisyllabic vowels and consonants. He had not achieved bowel and bladder control. He was hyperactive and clumsy and had frequent temper tantrums. Radiogram of skull showed increased density of calvarium with normal sella and sutures. The bone age was 6.4 years. CT scan of brain, glucose tolerance test and thyroid hormone levels were normal. Basal growth hormone level was 4.5 ng/mL. EEG did not show any epileptic focus.

At 5 years of age, his height was 119 cm and weight 28 kg. Head circumference was 57.5 cm. A repeat growth hormone estimation was normal. He had poor speech and was attending a

special school. His IQ was 65. Bone age was 7.1 years. His hyperactivity had decreased but he showed behavioural immaturity and had difficulty in mixing with peer group. He was sometimes violent and aggressive at home but showed good capability to memorize and was progressing satisfactorily at school.

At 7 years of age, his height was 130 cm (90th percentile NCHS) and weight 34 kg (90th percentile NCHS). Head circumference was 57.5 cm. Bone age was 7.4 years. He had poor expression of language but was speaking bilingually (Bengali and English). He was clumsy, had a shuffling gait and was accident-prone. He still had some difficulty in mixing with peer group. However, due to his good memory power his school performance was satisfactory in academics but poor in sports activities.

Discussion

Sotos' syndrome is characterized by large birth size and increased growth in the first few years of life. This is associated with advanced height, weight, and bone age, with macrosomia, frontal bossing, dolichocephalic skull, hypertelorism, prominent mandible and anti-mongoloid slant of eyes [2,5,6]. There is delay in neuropsychomotor development during infancy. The psychological abnormalities are behavioural disorders, tantrums, hyperactivity, destructiveness, social withdrawal, and eating and sleeping difficulties. There is no specific language impairment [7]. Linear growth stabilizes by four to five years of age.

The etiology of this syndrome is still unknown. Certain chromosomal abnormalities have been detected including translocations [8]. Autosomal dominant, recessive and familial cases have described [2,5]. Endocrinal studies are normal [2,5] though glucose intolerance has been seen in 14 per cent of cases [6]. Studies done to detect any abnormalities in the pituitary and hypothalamus have

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been unremarkable [9]. There is an increased tendency to tumours reported by some authors [10]. Our patient showed typical facial features and growth characteristic. The good improvement in psychosocial behaviour and scholastic abilities suggest that a hopeful prognosis should be given at the time of diagnosis.

REFERENCES

1. Sotos JF, Dodge PR, Muirhead D, Crawford JD, Talbot NB. Cerebral Gigantism in childhood. A syndrome of excessively rapid growth and acromegalic features and non-progressive neurological disorder. *N Engl J Med* 1964; 271: 109-16.
2. Cole TR, Hughes HIE. Sotos' syndrome. A study of the diagnostic criteria and natural history. *J Med Genet* 1994; 3: 20-32.
3. Udani RH, Wagle AK, Shah AB, Pal PM. Cerebral Gigantism (Sotos' Syndrome) *Indian Pediatr* 1988; 25: 392-4.
4. Menon PSN, Batra CM, Misra NK, Koyana R. Cerebral Gigantism (Sotos' Syndrome) *Indian Pediatr* 1991; 28: 291-5.
5. Wit JM, Beemer FA, Berth FG. Cerebral Gigantism (Sotos' Syndrome). Compiled Data of 22 cases. Analysis of clinical features, growth the plasma somatomedins. *Eur J Pediatr* 1985; 144: 131.
6. Dodge PR, Holmes SJ, Sotos JF. Cerebral Gigantism. *Dev Med Child Neurol* 1983; 25: 248-52.
7. Enegam JK, Cole TR, Kingwell E, Smith ML, Smith M, Sytarenios G. Language and behaviour in children with Sotos' Syndrome. *J Am Acad Child Adolesc Psychiatry* 1994; 33: 1307-15.
8. Schander-Stompel CT, Fryns JP, Hamers GG. Sotos' Syndrome and de novo balanced autosomal translocation. *Clin Genet* 1990; 37: 226-9.
9. Whitaker MD, Scheithauer BW, Hayles AB, Okazaki W. The hypothalamus and pituitary in Cerebral Gigantism. A clinico-pathological and immunocytochemical study. *Am J Dis Child* 1985; 139: 679-82.
10. Hersh JI, Cole TR, Bloom AS, Bertolone SJ, Hughes HIE. Risk of malignancy in Sotos' Syndromes. *J Pediatr* 1992; 120: 572-4.