Volume : 2 | Issue : 7 | July 2013 • ISSN No 2277 - 8160

Research Paper

Medical Science

Precocious Puberty due to Hypothalamic Hamartoma: A case presentation

Dr Shilpa S Bhat	Assistant Professor, SKNMC & GH, Narhe, Pune.
Dr Arti Shirsath	Assistant Professor, SKNMC & GH, Narhe, Pune.
Dr Pushplata Naphade	Professor and Head of Department of OBGY, SKNMC & GH, Narhe, Pune.

KEYWORDS: Precocious puberty, Hypothalamic Hamartoma

Introduction: Pubertal changes before age of 8 years (menarche before age of 10 years) is called Precocious puberty. It is five times more common in girls than in boys.

Most common form is idiopathic or constitutional; but this must be a diagnosis of exclusion with prolonged follow up in an effort to detect slowly developing lesions of brain, ovary and adrenal glands.

A girl of age 3 and half year old presented in pediatric OPD with her mother.

Mother noticed increase in her height ,enlargement of breast size and appearance of axillary hairs. After investigation MRI showed Hypothalamic Hamartoma.

Treatment started with GnRH agonist 3.75mg im monthly and psychological counseling. After 6 months of treatment at present breast and axillary hair changes regressed.Treatment has to be continued till average age of puberty.

Case Presentation: Three and half year old girl was brought by her mother to OPD. Mother noticed increase in her height than that of her elder son. She also noticed enlargement of breast size and appearance of axillary hairs. On examination her height-114cm and weight 18kg (more than 97th percentile for that age).Breast tanners stage3, Axillary hairs tanners stage 2. Investigations-Heamogram- PBS- ,ESR- ,URINR Routine and Micro- ,BSL Random,TFT-within normal range.

X-ray wrist AP and lateral- and X ray Knee AP and lateral –Bone age 6-7 years.USG Abdomen and pelvis NAD; Fundoscopy –NAD; MRI Brain plain and contrast-Hypothalamic Hamartoma of size 9x9x7mm.

Treatment started with GnRH agonist inj.3.75mg monthly IM. GnRH agonist injections have to be continued till average age of puberty. Counseling should include need of continuation of treatment. These kids are over expected by teachers and friends .These kids can be wrongly labeled as mentally retarded. Parents counseled that these are physiological changes but appeared earlier. It will not affect her future fertility nor the age of menopause.



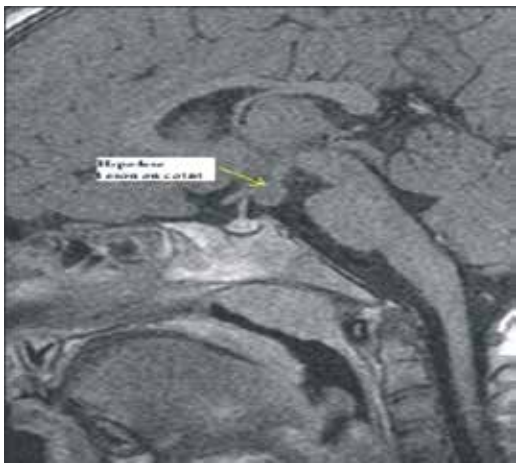
Presented as---







Bone age 6-7years



MRI Brain contrast- Hypothalamic



Hamartoma After Treatment of 6 montths

Discussion: At puberty, maturation of the hypothalamic-pituitary-gonadal axis results in the profound physical, sexual, and emotional changes that transform a girl into a woman and a boy into a man. Difficult under normal circumstances, puberty out of synchrony with friends and classmates has the potential for significant emotional distress. If one accepts the mean ±2.5 SDs as encompassing the normal range, puberty is precocious if changes occur in a girl before 8 years (or possibly 7 years if a more recent study is correct) and in a boy before 9 years. Precocious puberty requires a careful history, physical examination, laboratory evaluation, and imaging studies to search for an underlying cause; however, in most children, especially girls, no abnormal cause is identified. Treatment is available for most cases of gonadotropin-dependent and gonadotropin-independent forms of precocious puberty.

Although children with precocious puberty undergo early physical and sexual development, their emotional and intellectual development is commensurate with their chronologic age. They and their parents require special counseling to help them adjust to these changes. Several factors seem to influence the time of initiation of puberty, including genetic factors, geographic location, exposure to light, and general health and nutrition. In family studies, the onset of the mother's puberty seems more important than the father's puberty; the daughters and sons of early-maturing mothers are early to mature.[4] In girls, sexual precocity usually begins with breast development, although pubic hair growth or vaginal bleeding may be the initial manifestation. Other signs of estrogen secretion include nipple enlargement and pigmentation, thickening of the labia minora, a change in the vaginal mucosa from shiny red to dull purple, and vaginal mucus production. Some emotional lability and cyclic mood swings may be present. Percentage of body fat composition increases, and a female body contour develops. Precocious puberty is divided most easily into gonadotropin-dependent and gonadotropin-independent sexual precocity. Gonadotropin-dependent precocity is almost always pituitary-mediated and so is also termed *central precocious puberty*. Gonadotropin-independent precocity, usually a result of a gonadal or adrenal disorder, is also termed *peripheral precocious puberty*. Precocious puberty is 3–5 times more common in girls than in boys

Investigations-A careful history and physical examination are important in directing the laboratory evaluation. A search for exogenous estrogens (or androgens) should include not only questions regarding birth control pills, but also estrogen-containing cosmetics and 'health foods'. Symptoms and signs of hypothyroidism, possibly associated with galactorrhea, suggest this disorder as the cause of sexual precocity. Irregular café-au-lait spots and orthopedic problems or bone cysts on radiologic examination point toward McCune-Albright syndrome, whereas round café-au-lait spots, axillary freckling, and skin neurofibromas point toward von Recklinghausen's disease. Neurologic and ophthalmologic abnormalities should raise suspicion of a cerebral abnormality associated with precocious puberty. Attempts have been made to identify useful clinical and hormonal predictors of gonadotropin-dependent precocious puberty that identify girls at risk for an underlying central nervous system abnormality as the cause of their precocious puberty. A retrospective cohort study of 197 girls showed that 6% had an identifiable central nervous system abnormality. Of the 11 girls with an abnormality, only 3 were older than age 6 years. Overall, there was a 1:5 chance of finding a central nervous system abnormality in a girl younger than age 6 and a 1:50 chance of finding an abnormality in a girl older than age 6.

Treatment-Aims

To maximize adult height

Regression and arrest of pubertal changes.

Supression of menses

Psychological support.

Children with precocious puberty are at risk for psychological consequences of their early sexual development and reduced adult stature from their accelerated skeletal maturation. GnRH agonists, whose continuous stimulus results in initial stimulation followed by down-regulation of the gonadotropin receptor, have proved to be relatively safe and effective treatment for children with gonadotropin-dependent precocious puberty. Six commercial GnRH agonists, differing slightly in amino acid structure and metabolic clearance and relative potency, have been used to treat central precocious puberty .Side effects of GnRH agonist treatment may include local reactions such as erythema, induration, and wheal and sterile abscess formation; negative effects on growth velocity; and transient headaches, hot flushes, and withdrawal bleeding.Patients must be monitored during therapy to allow correct adjustment of drug dosage, including assessment of Tanner stage, growth rate, bone age advancement, and hormone production. Signs of puberty, such as breast development, genital maturation, and pubic hair growth, should fail to progress and may regress. Growth velocity should return to normal, and the rate of bone age advancement should slow to normal or less than normal. Some children experience a marked decrease in growth velocity on GnRH agonist therapy; preliminary studies suggest that the addition of growth hormone treatment improves growth velocity and predicted adult height. Most hypothalamic hamartomas grow slowly and stop enlarging when the central nervous system completes its growth at 8–12 years of age. Unless these tumors cause vision impairment, hydrocephalus, or focal neurologic abnormalities, they are best followed carefully with serial MRI studies. When hamartomas are complicated by gelastic seizures, neurosurgical excision may be considered in cases refractory to anticonvulsant therapy.

Careful consideration must be given to the management of psychosocial problems in all children with precocious puberty. As mentioned previously, these children have intellectual, behavioral, and psychosexual maturation in keeping with their chronologic age, not their physical or pubertal age. They do not have early sexual activity. Parents, teachers, and peers may have unrealistic expectations of their intellectual and athletic abilities, and these children may even be labeled inappropriately as developmentally delayed. A careful explanation of these considerations must be given to parents. Children should be counseled that their secondary sexual characteristics are normal albeit early. If the child is bright, advancement in school may be possible with special tutoring and may prove beneficial. Children with precocious puberty may place stress on the marital or family relationship, and in these situations formal psychological counseling may be indicated.

REFERENCES

1 Tanner JM: Growth at Adolescence. 2nd ed.. Oxford, Blackwell Scientific Publishers, 1962 | 2 Marshall WA, Tanner JM: Variations in pattern of pubertal changes in girls. Arch Dis Child 44:291, 1969 | 3 Marshall WA, Tanner JM: Variations in the pattern of pubertal changes in boys. Arch Dis Child 43:13, 1970 | 4 Garn SM, Sullivan TV: Are parent-child resemblances in growth sex-limited? Am J Dis Child 141:127, 1977 | 5 Zacharias L, Wurtman RJ, Schutzoff M: Sexual maturation in contemporary American girls. Am J Obstet Gynecol 108:833, 1970 | 6 Zachmann M, Prader A, Kind HP, et al: Testicular volume during adolescence: Cross-sectional and longitudinal studies. Helv Paediatr Acta 29:61, 1974 | 7 Herman-Giddens ME, Slora EJ, Wasserman RC, et al: Secondary sexual characteristics and menses in young girls seen in office practice: A study from the Pediatric Research in Office Settings Network. Pediatrics 99:505, 1997 | 8 Lee PA, Kulin HE, Guo SS: Age of puberty among girls and the diagnosis of precocious puberty. Pediatrics 107:1493, 2001