CASE REPORT

Precocious Puberty and Pineal Cyst – An Uncommon Association

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We present a five year old boy with central precocious puberty and pineal gland cyst on neuroimaging. This association is uncommon and highlights the role of pineal gland in puberty.

Key Words: *Pineal gland cyst, Central precocious puberty, Pineal tumors.*

Pineal gland secretes melatonin whose regulatory role on puberty is hypothesized but ill defined. Evidence exists that disorders of the hypothalamic-pituitary-gonadal axis are linked to altered plasma melatonin profile, demonstrating the role of the pineal gland in reproductive endocrinology(1).

Pineal gland lesions present mostly as tumors and rarely as cystic lesions. They affect children and adolescents and the clinical presentation is usually prolonged history of head ache and vomiting. Germ cell tumors are the commonest tumors of pineal gland and the elevated human chorionic gonadotropin results in sexual precocity in these patients. Pineal gland cysts are usually asymptomatic and are rarely reported to be associated with central precocious puberty(2). We present an interesting case of a young boy with true sexual precocity and an associated pineal cyst to highlight the role of pineal gland in pubertal sexual maturation.

CASE REPORT

A 5 year old boy was brought by parents with history of rapid increase of height, enlargement of the external genitalia, and appearance of pubic hair, voice change and presence of acneiform lesions over face for last two years. The boy was the last sibling amongst four (2 girls and 2 boys), born out of a third degree consanguineous marriage with all other siblings reportedly normal. The birth weight was 2.8 kg and his motor and mental milestones were normal. The parents denied history of headache, vomiting, seizure episodes, loss of consciousness and any other feature to suggest CNS illness. They also denied any family history of precocious puberty or exposure to medications by the child.

Anthropometric examination revealed: height 125 cm (>95th centile), weight 23 kg (>95th centile), upper/lower segment ratio 1.2 and arm span of 122 cm. Pubertal assessment by Tanner grading revealed G3P3A2 with bilateral scrotal testes of volume 8-10 mL and stretched penile length of 7 cm. Detailed clinical examination revealed acne over face, with no evidence of neurocutaneous markers, midline defects, goiter or evidence of systemic disease. Neurological examination revea-led normal visual fields and fundi with no clinically detectable abnormality.

Estimated bone age was 12 years by the Greulich-Pyle method. His hormonal evaluation revealed a basal LH (1.1 IU/L) and FSH (0.7 IU/L) and a high level of serum testosterone (4.3 nmol/L). Other hormonal evaluation was normal. T1- and T2-weighted and FLAIR magnetic resonance imaging

(MRI) images showed a 1.1 × 0.8 × 0.7 cm cyst in the pineal region. The content of the pineal cyst was homogeneous and isointense relative to cerebrospinal fluid. The wall of the cyst was slightly thickened with focal irregularity. Based on history, findings of precocious puberty and pubertal hormonal profile, we started the patient on depot preparation of GnRH-analogue (Inj Leuprolide 3.75 mg subcutaneous every 4 weeks). In view of asymptomatic pineal gland cystic lesion, no neurosurgical intervention was planned immediately and the patient is kept under regular follow up.

DISCUSSION

Our patient, a case of true precocious puberty had an associated pineal cyst. Literature search revealed only three similar case reports earlier(2,3). Pineal cysts are benign, asymptomatic and are detected incidentally with a population prevalence of 1-4%. Our case had typical features of true precocious puberty without neurological symptoms or signs attributable to pineal cyst. Hence, the pineal cyst was considered as an associated finding and the boy was started on medical therapy with Leupro-lide. Pineal cysts of less than one cm and asymptomatic should be followed with serial neuroimaging and symptomatic large cysts are subjected for surgical resection. There is no role of chemotherapy or radiotherapy for benign pineal cysts(4). In our case, surgical resection was not contemplated in view of small size and asymptomatic presentation.

The pineal gland influences human reproductive function at hypothalamic-pituitary level, by inhibition of the gonadotropin releasing hormone (GnRH) pulse and also at the gonadal level(2,5). Pineal gland germ cell tumors cause sexual precocity by release of human chorionic gonadotropin but the mechanism of precocity is unknown with other lesions. Data from animal models and human studies suggest that there is removal of the gonadotropin inhibition by melatonin leading to precocity or stimulation of hypothalamo-pituitary-gonadal axis via a secretory product analogous to GnRH. Other proposed mechanisms are loss of inhibitory effect of the pineal gland on gonadotropin release. The contributing factors are size of the cyst and alterations in the functional capacity of the gland due to cystic nature of pineal gland(6).

To conclude, we present an unusual association between precocious puberty related to pineal cyst. The underlying etiological association is unclear and this case exemplifies the enigmatic role of the pineal gland in puberty.

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