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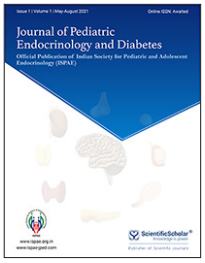

Case Report

Rare presentation of pituitary stalk lipoma as central precocious puberty in a girl

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ABSTRACT

Central precocious puberty (CPP) in girls is most commonly idiopathic. Here, we present a case of a 5-year 3-month-old child who presented with CPP with a stalk lipoma on magnetic resonance imaging. On examination, the sexual maturity rating was A1P1B (Rt B1, Lt B2), respectively. The hormonal evaluation revealed pubertal gonadotropin and E2 levels, respectively, with pubertal ultrasound (USG) parameters. She was started on gonadotropin-releasing hormone agonist therapy and is on regular follow-up. This case highlights stalk lipoma as a cause of CPP.

Keywords: Precocious puberty, Pituitary stalk lipoma, Organic cause of CPP

INTRODUCTION

Central precocious puberty (CPP) is defined as breast enlargement in females before the age of 8 years and as testicular enlargement ≥4 mL in males before the age of 9 years.[1-3] Magnetic resonance imaging (MRI) of the pituitary and hypothalamus is one of the first-line investigations to rule out organic lesions as a cause of precocious puberty. Organic causes such as hypothalamic hamartoma and germinoma are more common in males, while, in females, the cause is usually idiopathic.[4] Organic lesions are more frequent in children who manifest with puberty before the age of 4 years. Lipomas of the central nervous system are extremely rare, accounting for <1% of intracranial neoplasms.[5] They are located in the midline, in the interhemispheric fissure, or in the superior cerebellar cistern. Most are asymptomatic and incidentally detected. They may be associated with other midline anomalies such as corpus callosum abnormalities and vascular anomalies.[6] Here, we present an unusual case of pituitary stalk lipoma presenting as CPP in a 5-year 3-month-old girl.

CASE PRESENTATION

A 5-year 3-month-old female child was admitted for evaluation of premature development of the left breast noticed over 1 year. There was no history of vaginal bleeding or a noticeable growth spurt or appearance of axillary or pubic hair according to parents. There was no history of headache, visual disturbances, head trauma, intracranial lesion, radiation, suggestive drug intake, or use of any topical estrogen cream. There was no history of weight gain, lethargy, or poor concentration. She has average scholastic performance. Parents had attained puberty at the appropriate age. The child was a first-order child, born by vaginal delivery with a birth weight of

2.5 kg, birth length unavailable, out of non-consanguineous marriage, cried at birth with no history of prolonged jaundice, or intensive care unit stay.

On examination, her height and weight were 107 cm (25^{th}–50^{th} centile on IAP growth charts) and 15 kg (10^{th}–25^{th} centile on IAP growth charts), respectively, and her sexual maturity rating was breast staging Tanner 1 for the right and Tanner 2 for the left, respectively, with Tanner 1 for axillary and pubic hair [Figure 1a and b]. There was no available baseline height measurement to calculate growth velocity considering her height centiles in view of precocity. Her mid-parental height was 144.5 cm (<3^{rd} centile).

The routine investigations including complete blood counts, liver/renal function tests, lipid profile, glycemic status, calcium profile, and thyroid function tests were normal. Basal luteinizing hormone (LH), follicle-stimulating hormone (FSH), and 17β estradiol levels were 7 mIU/mL (female: basal 0.8-26 mIU/mL, ovulatory 25-57 mIU/mL, post-menopausal 40-104 mIU/mL, pre-pubertal <0.3 mIU/mL, adult male: 4-18 mIU/mL), 12 mIU/mL (female: basal 1.4-9.6 mIU/mL, ovulatory 2.3-21 mIU/mL post-menopausal 34-96 mIU/mL, male: 3-18 mIU/mL), and 27.5 pg/mL (pre-pubertal <9 pg/mL), respectively. Her bone age was 7 years as per Greulich and Pyle's radiological atlas. Her predicted height was 147.4 cm which was within her target height range (147.2 ± 5 cm).

Ultrasound (USG) pelvis revealed uterine volume of 7.3 cc (pre-pubertal uterine volume <1.8 cc) , right ovary 2.5 cc, and left ovary 2.4 cc (pre-pubertal ovarian volume <1.6 cc) , respectively, with uterine length 5.4 cm (pre-pubertal uterine length <3.5 cm) and endometrial thickness of 2 mm [Figures 2, 3a and b]. MRI pituitary and hypothalamus revealed a smoothly marginated thickening of the infundibulum (measuring 4.7 mm in thickness) with T1 and T2 hyperintensity with an anterior pituitary height of 4 mm and a visible posterior pituitary bright spot [Figure 4]. The hyperintense lesion in the pituitary stalk was hypointense on fat suppression MRI [Figure 5], suggestive of lipoma. Her insulin-like growth factor 1, 8 am serum cortisol, and serum prolactin in dilution were 64 ng/mL (50th–75th centile for age), 233 nmol/L (8.48 µg/dL; normal range 8 am Cortisol 137.95-634.57 nmol/L; 5-23 µg/dL), and 7.59 ng/mL (Female: 5-25 ng/mL, Male: 3-18 ng/mL), respectively. The bladder diary during the hospital stay was not suggestive of polyuria. Visual acuity was 6/6 in both eyes, with a visual field corresponding to that of the examiner with normal fundus.

The patient was started on gonadotropin-releasing hormone (GnRH) analog therapy (Inj. Leuprolide acetate depot at 150 µg/kg intramuscular every 4 weeks) and is on regular follow-up every 3 months. After discussion with the department of neurosurgery, considering the location of the lipoma, surgery is not planned and conservative medical therapy is to be continued with imaging follow-up for size increment.

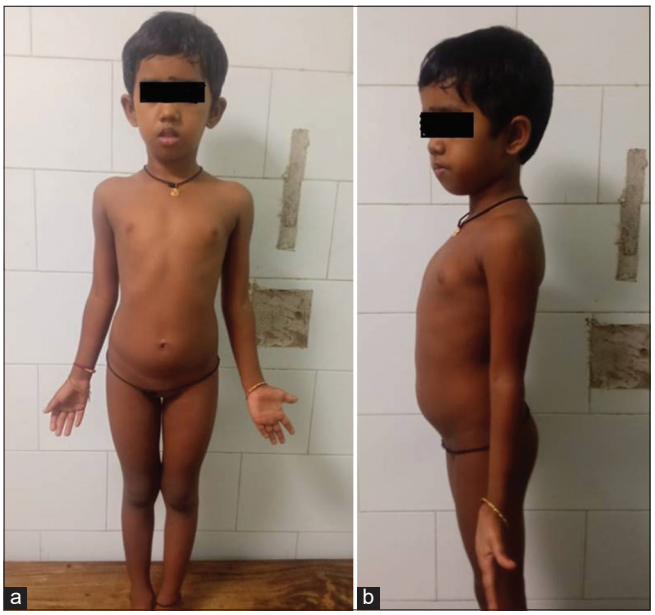

Figure 1: (a and b) Clinical image Tanner stage A1P1B1 (R), B2 (L).

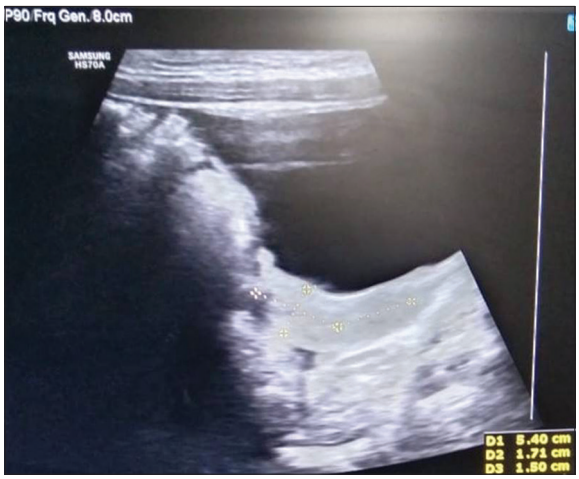

Figure 2: Ultrasound abdomen/pelvis: Uterus.

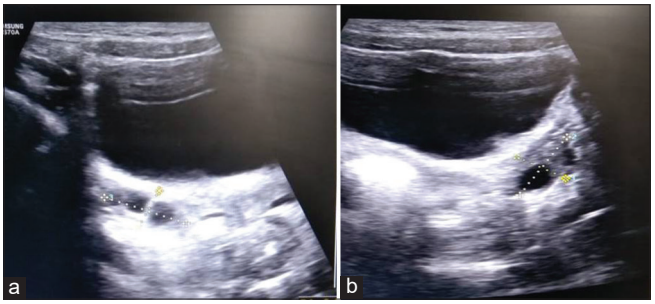

Figure 3: Ultrasound abdomen/pelvis: (a) left ovary and (b) right ovary.

During the first 3 month follow-up evaluation, the pubertal staging was static and prepubertal hormonal parameters

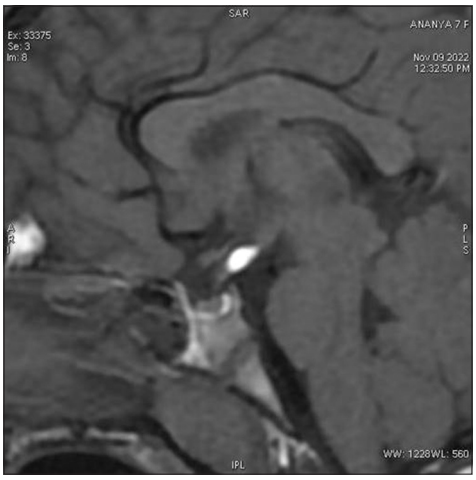

Figure 4: T1 magnetic resonance imaging pituitary showing hyperintense lesion in pituitary stalk.

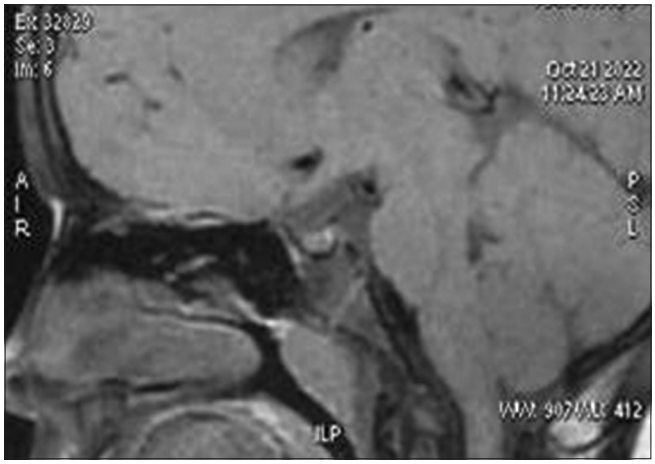

Figure 5: Hyperintense lesion on T1 suppressed in fat suppression sequence suggestive of lipoma.

(Basal LH – 0.2 mIU/mL and 3-h leuprolide stimulated LH 3.4 mIU/mL, respectively). USG pelvis revealed prepubertal uterine and ovarian parameters. On follow-up after 1 year of therapy, growth velocity was 4 cm/year, and bone age had not advanced further. The patient is continuing on monthly GnRH analog therapy.

DISCUSSION

Intracranial lipoma presenting as precocious puberty has been rarely described. Intracranial lipomas are described in the midline region, usually per callosal and dorsal mesencephalic.[6] Lipomas are mostly asymptomatic except in rare cases when their excessive growth or specific location can lead to neurological signs. In a recent case report, Giacomozzi *et al*. described a 7-year 3-month-old girl child with progressive breast enlargement.[7] The detailed evaluation revealed CPP and MRI pituitary and hypothalamus revealed a suprasellar lipoma stretching the pituitary stalk. Similarly, a case report by Vivanco-Allende *et al*. described a case of CPP with an osteolipoma of the tuber cinereum in a girl child of 7 years 8 months. The mechanism of precocious puberty has been postulated to be due to pituitary stalk compression.[8]

Surgical excision has been performed in a case of lipoma with precocious puberty which led to regression of pubertal characteristics and normalization of hormonal parameters to prepubertal level.[9] CPP is a rare condition, and cases produced by an organic cause are extremely rare. We present this case as an example of the medical treatment of a patient with CPP and a lipoma of the infundibulum. As most lipomas remain stable over time, the patient can be monitored serially for any change in size. Evidence for a causative correlation between lipoma and CPP has not been established.

CONCLUSION

CPP in girls is most commonly idiopathic. Organic causes of CPP in girls are rare with intra-cranial lipoma being one of the rarer causes. MRI reveals hyperintense lesion in T1 and T2. The use of fat-suppressed T2-weighted imaging technique aids in the diagnosis.

Intracranial lipoma is usually stable in size and associated precocious puberty can be medically managed with a follow-up MRI for size evaluation.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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