



Gynaecomastia With Growth Retardation In Child With Primary Empty Sella Syndrome

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ABSTRACT

We present a 12 year-old boy, who had chronic headache and gynecomastia associated with growth retardation (proportionate short stature). Bone age and blood tests suggested pituitary deficiency. CT and MRI scan revealed primary Empty sella syndrome (ESS) where the sella turcica was filled with cerebrospinal fluid flattening the normal pituitary. The absence of space occupying lesions on neuroimaging ruled out secondary causes like brain tumours. Resolution of symptoms with normalization of pituitary function within 6 months of placement of ventriculoperitoneal shunt reaffirmed the diagnosis. Our case suggests that gynecomastia may be the initial manifestation of primary Empty sella syndrome. Hence empty-sella syndrome should be included among the causes of gynecomastia with proportionate short stature in children

KEY WORDS : Gynecomastia, Empty Sella syndrome, Proportionate short stature, Ventriculoperitoneal shunt

Case Report

Primary empty sella syndrome is rare in childhood and to find empty sella syndrome associated with gynecomastia (probably due to the interruption of the normal hypothalamo-pituitary neurovascular connection) is all the more unusual. This is the first report on gynecomastia with growth retardation, due to 'primary empty sella syndrome' in a child.

A twelve years old boy, born to non-consanguineous parents presented to us with second episode of afebrile generalised tonic clonic seizures. The child had a similar episode four years ago, when he was started on phenytoin by a local practitioner. Phenytoin was self discontinued 2 years later. There was no history of trauma, drug intake, radiation, recent immunization, dog bite, exanthema, febrile illness, bleeding or any history suggestive of cranial nerve deficits e.g regurgitation of feeds, frequent falls, visual or hearing difficulty or contact with tuberculosis. The birth, family and the immunization history were not contributory. Examination revealed a short statured child with a height of 120 cm (Expected height 149cm.) The upper segment: lower segment ratio was 1:1. The testicular volume and penile length were normal (3 ml and 6 cm respectively). Gynecomastia was

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present. However there was no evidence of precocious puberty[1]. Fundus examination revealed mild papilloedema.

Investigations

Blood Sugar, urea, creatinine, electrolytes, urine (microscopy, biochemical analysis and specific gravity) were normal. The growth hormone and thyroid functions were depressed. The Growth hormone levels were 0.1ng/ml at basal level and 0.04ng/ml, 0.07ng/ml, 0.08, ng/ml (very low). Post stimulation at 30 minutes, 60 minutes, 90 minutes respectively (against the normal value of 7ng/ml-22ng/ml). Testosterone less than 0.01 nano gram per ml at basal level (against a normal of 1.7 to 7.8). Cortisol levels (8 AM) were 2.9 μ g/dl against a normal of 8.7–22.4 μ g/dl. T3-180ng/dl(NR 80–180 ng/dl),T4-2.9 ug/dl(NR 4.6–12 ug/dl),TSH- 5.2uU/ml (NR 0.5–6 uU/ml); In view of low serum cortisol, low GH(basal and post stimulation) and an inappropriate TSH for the low T3/ T4 levels, a diagnosis of hypopituitarism was established. Bone age of 4–5 years(delayed by 7 years) further confirmed the picture. The plain skull radiographs showed an enlarged sella turcica with no abnormal intracranial calcifications. Coronal computerized tomography scans (CT) through the sella turcica demonstrated an enlarged sella turcica filled with cerebro spinal fluid. Magnetic resonance MRI showed primary type empty sella syndrome. T2-weighted sagittal magnetic resonance image showed a compressed pituitary and cerebrospinal fluid-filling the sella.

Discussion

Empty sella syndrome often accompanies idiopathic intracranial hypertension. Normally the walls of the pituitary fossa protect the gland from the surrounding pressure, but due to the CSF fistula in the lamina cribrosa created by raised intracranial

pressure or a congenital deficiency in the diaphragm of sella, the CSF accumulates in an enclosed and in elastic space, causing increase in the intrasellar pressure which can obstruct the neuro-hypophyseal vessels between hypothalamus and pituitary gland. This may lead to hypopituitarism and also hyperprolactinaemia causing gynecomastia (due to disruption of the prolactin inhibiting factor released by hypothalamus) as seen in our case. The coexistence of any brain tumour (secondary empty sella) was ruled out by MRI[2]. Clinical studies of patients with ESS have revealed a variety of associated features and it seems likely that several aetiologic factors are involved. Most common hormone deficiency in empty sella syndrome is GH (30%) followed by TSH (15%). The presence of proportionate short stature with no familial /genetic element or chronic illness led us to suspect a pituitary disorder. A ventriculo-peritoneal shunt reverts hypopituitarism and hyperprolactinaemia due to release of this obstruction, so that the neuro-hypophyseal stalk can deliver the prolactin inhibition factor (PIF)[3]. Stalk compression may also sometimes, cause hypoadrenalinism due to lack of normal hypothalamic control over ACTH secretion and low serum sodium may indicate secondary adrenal insufficiency. However, the electrolytes were normal in our case.

This child was managed by placement of a ventriculo-peritoneal shunt and improvral in thyroid function and growth rate following the shunt showed the importance of early diagnosis of this treatable condition.

Conclusion

Empty-sella syndrome should be included among the causes of proportionate short stature in pediatric patients, especially if associated with gynecomastia. A high index of suspicion is needed in such cases so that early diagnosis can be made and necessary treatment given.

References

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