

β -hCG secreting mediastinal tumour, a rare cause for gonadotropin independent precocious puberty (GIPP)

Jananie Suntharesan¹, Nayana Samarasinghe², Sanjewa Gunasekara³, Navoda Atapattu¹

Ceylon Medical Journal 2019; **64**: 158-160

DOI: <http://doi.org/10.4038/cmj.v64i4.8992>

Introduction

Precocious puberty (PP) is defined as the onset of secondary sexual characteristics before the age of 8 years in girls and 9 years in boys. PP can be due to gonadotropin dependent or independent causes. GIPP some of the secondary sexual characteristics appear without activation of normal hypothalamic-pituitary-gonadal axis. GIPP can present as phenotypically similar (iso sexual) or heterosexual puberty. Untreated congenital adrenal hyperplasia (CAH) and McCune-Albright syndrome are the commonest causes of iso sexual GIPP. Androgen secreting tumors such as adrenocortical tumor and gonadal tumors, leads to peripheral precocious puberty in boys and virilization in girls. β -hCG producing germ cell tumor and hepatoblastomas lead GIPP mostly in boys. We present a rare case of β -hCG producing anterior mediastinal seminoma in an eight years old boy, who presented with GIPP.

Case report

8 years old boy presented with deepening of voice, increase height velocity and associated penile enlargement, axillary and pubic hair growth for one-month duration. On examination child was not pigmented, not dysmorphic and there was no café au lait spots or bony deformities suggestive of Mc Cune Albright syndrome. His height was 134.5cm in 75th centile. He had pubic hair stage II. His phallus length was 9.5cm with good width, but his testicular volume was 3-4 ml bilaterally. He was normotensive with normal respiratory and abdominal examination. Neurological examination was unremarkable. During evaluation, noted to have rapid increase in height

velocity (97th centile) and pubertal progression to tanner stage V with bilateral testicular volume 6-7 ml.

Discrepancy with the testicular volume and pubertal stage suggested, GIPP in this child. Bone age at presentation was appropriate to the chronological age (Figure 1), which indicated the rapid onset of puberty. Biochemical investigations revealed suppressed gonadotropins with elevated testosterone and β -hCG (Table). Even though there was no mediastinal widening on his initial chest x-ray (Figure 2), CT chest showed a well-defined soft tissue mass with heterogenous contrast enhancement in the anterior mediastinum (7 × 4 × 5.6 cm) suggestive of germ cell tumor (Figure 2).



Figure 1. Bone age 7 years at presentation.

¹Endocrine and Diabetes Unit, Lady Ridgeway Hospital for Children, ²Department of Radiology, Lady Ridgeway Hospital for Children, ³National Cancer Institute, Apeksha Hospital Maharagama.

Correspondence: JS, e-mail: <jananiesuntharesan@gmail.com>. Received 30 April 2019 and revised version 14 October 2019 accepted 14 December 2019.



This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

Table.

Time period	At presentation (26/10/2018)	At one month (30/11/2018)	Post operatively (5/03/2019)	Post OP 4 months (8/06/2019)
S Testosterone (0.1-1nmol/l)	25		0.15	
17 OHP (<6nmol/l)	8.17			
LH (0.08-3.9)IU/l	<0.07		0.2	0.66
FSH (0.1-1.3)IU/l	<0.05		0.35	2.27
β hcg (<0.7 IU/l)	90.33	261.9	0.07	< 0.9
α Feto protein (<10ng/ml)	3.64	0.91		

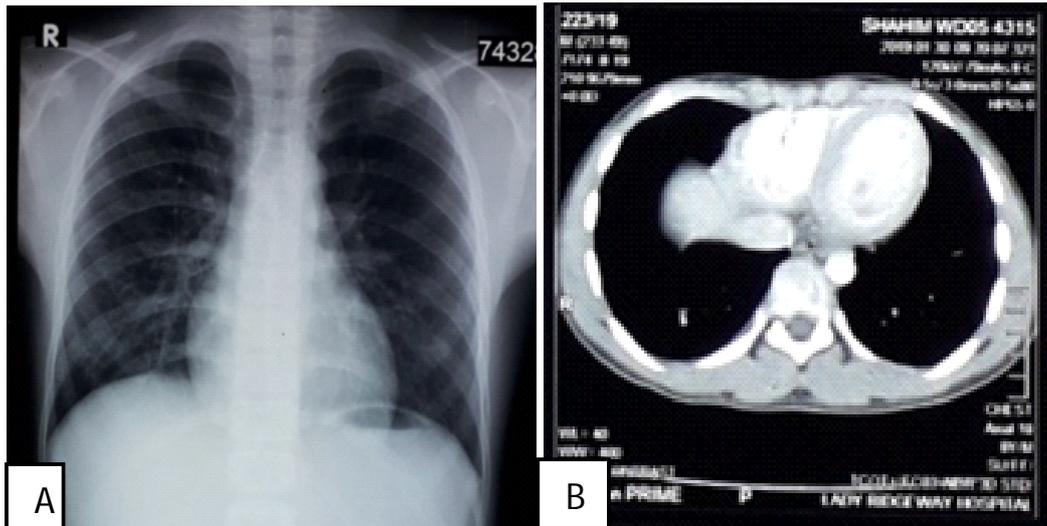


Figure 2. Chest X-ray no mediastinal widening (A) CT thorax showed well-defined soft tissue mass the anterior mediastinum (B).



Figure 3. Bone age advanced to 12.5 years over eight months.

Surgical resection was carried out as the frozen sections were inconclusive. Histology revealed seminoma without other evidence of germ cell components (yolk sac tumor, embryonal carcinoma, choriocarcinoma or teratoma). He received cisplatin-based chemotherapy for his seminoma. During follow up child entered into gonadotropin dependent puberty with elevated LH and advanced bone age (Figure 3) for which he was started on gonadotropin analogs.

Discussion

The prevalence of non CAH GIPP is 14 per 1,000,000 children. HCG secreting tumors account for 5.5% of GIPP (1). β -hCG causes peripheral precocious puberty in boys via cross-reaction with LH receptors, which produce testosterone. In girls FSH is needed for initiation of puberty. Boys present with isosexual precocious puberty with low LH. Boys have less pronounced testicular enlargement due to lack of FSH, which is needed for Sertoli cell proliferation (2). Extra gonadal germ cell tumors (EGCT) represent up to 5% of all germ cell tumors (GCTs) with an incidence around 1/1,000,000 (3). EGCTs derive from germ

cell, which migrate during the embryogenesis. EGCTs are mainly localized along the median axis. Extragonadal germ cell tumors can be benign-teratoma or malignant tumour. 90% of five-year survival rate is observed in mediastinal seminomas, whereas only 45% of five-year survival rate is observed in mediastinal nonseminomas [4].

The most commonly used chemotherapeutic regimen for germ cell tumors include cisplatin, etoposide, and bleomycin. Patients who have normal imaging or tumour size less than 3 cm after chemotherapy can be observed without further intervention.

Conclusion

β -HCG producing germ cell tumors are very rare, which can manifest as GIPP. High clinical suspicious along with early appropriate imaging is needed for definitive diagnosis. Cisplatin based chemotherapy is the treatment of choice followed by surgical resection if needed.

Conflict of interests

None.

References

1. Turan S, Guran T, Atay Z, Bereket A, Ozen S, Kara C, et al. The Etiology and Clinical Features of Non-CAH Gonadotropin-Independent Precocious Puberty: A Multicenter Study. *J Clin Endocrinol Metab* 2016; **101**: 1980-8.
2. Stephen MD, Zage PE, Waguespack SG. Gonadotropin-dependent precocious puberty: Neoplastic causes and endocrine considerations. *Int J Pediatr Endocrinol* 2011 (1): 184502.
3. Busch J, Seidel C, Zengerling F. Male extragonadal Germ Cell Tumors of the Adult. *Oncol Res Treat.* 2016; **39**: 140-4.
4. Bokemeyer C, Nichols CR, Droz JP, et al. Extragonadal germ cell tumors of the mediastinum and retroperitoneum: Results from an international analysis. *J Clin Oncol.* 2002; **20**: 1864-73.