



PRECOCIOUS PUBERTY SECONDARY TO ADRENAL ADENOMA –AN UNUSUAL PRESENTATION : A CASE REPORT

Paediatric Surgery

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ABSTRACT

Precocious puberty in pediatric practice can be due to different causes, including adrenal tumors. Adrenocortical tumor is a rare cause of precocious puberty resulting in androgenic hormonal excess and is termed as pseudo-precocious puberty. The coexistence of myelolipoma within adrenal cortical adenoma is extremely rare, for both tumors present usually as separate entities. There are only 16 such cases reported. We present the case of a four-year-old boy with precocious puberty and hormonal assays confirmed of pseudo-precocious puberty. Abdominal magnetic resonance imaging (MRI) reported a right-sided well-defined adrenal adenoma. This case report emphasizes adrenocortical tumor as a rare cause of pseudo-precocious puberty in young children.

KEYWORDS

precocious puberty, adrenocortical tumor

INTRODUCTION:

Precocious puberty is defined as the onset of secondary sexual characteristics before 8 years of age in females and 9 years in males. Evaluating precocious puberty in children is a complex process that requires a thorough understanding of the underlying mechanisms and causes. While many cases are benign and represent early, certain evaluations must consider rare but serious conditions. Malignant conditions, such as hormone-secreting tumors of the gonads or adrenal glands, although not usually the initial thought, must be ruled out to ensure accurate diagnosis and appropriate treatment. Adrenal tumors in children are categorized as functional (hormone-secreting), which are most commonly found in children and adolescents, or non-functional (silent), which are usually found in adults presenting with symptoms of abdominal discomfort or back pain caused by the large mass of the tumor.

Case Presentation:

A 7-year-old male who presented with signs of precocious puberty, including the appearance of pubic hair and an increase in penile length over the course of two months. On further evaluation hormonal evaluation suggestive

- AFP:<2 (Decreased)
- HCG:<1.2 (Normal)
- Serum Cortisol:165 (Normal)
- Serum ACTH:40.5 (Normal)
- Serum testosterone: 6(Increased for age)
- DHEA:331 (Normal)
- SLH:0.12 (Normal)
- 17 Alpha Hydroxy PG: 0.51 (Normal)
- 25 H Vitamin D: 0.51 (Insufficient)



Fig.1- clinical picture

MRI abdomen plus pelvis was done which was suggestive 4.7x 3.1x2.6cm lesion arising from right adrenal gland superiorly abutting segment VI and segment V of liver, inferiorly upper pole of right kidney, and medially right crus of diaphragm with no evidence of calcification or haemorrhage within ; with features suggestive of adrenal adenoma.

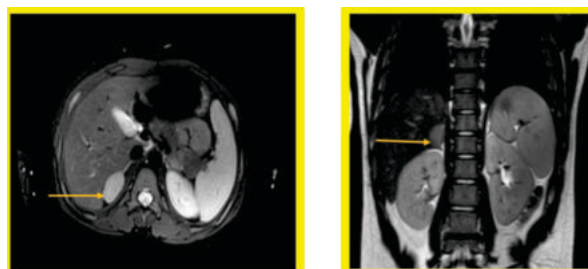


Fig.2 – MRI – right adrenal adenoma

The patient underwent laparoscopic right adrenalectomy for adrenal adenoma.

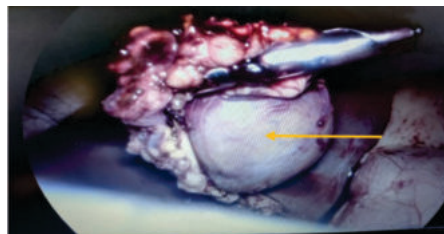


Fig.3- intraoperative image showing adrenal adenoma

Postoperatively child thriving well. Histopathological examination (HPE) revealed the presence of an adrenocortical adenoma with a focal myelolipoma component. Immunohistochemistry (IHC) showed SF1 was diffusely positive, while Inhibin was focally positive. The Mib-1 labeling index was notably increased in the myelolipoma component, indicating a higher proliferative activity.

DISCUSSION:

Adrenocortical tumors, comprising about 0.3-0.5% of neoplasms detected in patients under the age of 15, are functionally active in 95% of cases². These tumors often lead to the overproduction of androgens, corticosteroids, aldosterone, and estradiol, in decreasing order of frequency.³ However normal hormonal levels does not rule out adrenal cortical tumours.

At times the child is asymptomatic, however most commonly presents with virilisation. Children may also present with signs of cushings disease with symptoms such as central obesity, moon face, and hypertension. Hyperaldosteronism is uncommon.⁴ When evaluating children with hyperandrogenism, it is imperative to consider adrenocortical tumors in the differential diagnosis. Key hormonal assays include testosterone, DHEA-S, androstenedione, and 17(OH) progesterone. An elevated level of LDH marks a risk of malignancy.⁵ Higher levels of testosterone and DHEA-S, along with a lack of suppression after corticosteroid treatment and evidence of an adrenal lesion on imaging, should heighten the suspicion of an adrenocortical tumor. Features that may suggest a malignant character of adrenal mass include age <3.5 years, duration of less than 6 months⁶.

First line of imaging remains ultrasonography. CT scan gives more precise evaluation of the tumour in terms of size, capsule, demarcation, areas of calcifications, necrosis, or bleeding.⁷

Complete surgical resection is the mainstay treatment.⁸ Laparoscopic technique offers a large working space and clear anatomic landmarks, contributing to overall success. Compared with open procedures, laparoscopic surgery is associated with shorter postoperative hospital stays (2 to 3 days) and decreased blood loss⁹. While the retroperitoneal approach avoids entering the peritoneal cavity and reduces postoperative ileus, it provides a smaller working space and is generally limited to tumors smaller than 5 cm, making the transperitoneal approach necessary in this instance.

CONCLUSION:

This case report emphasizes the importance of considering adrenocortical tumors as a rare but critical cause of pseudo-precocious puberty in young children. It highlights the need for a thorough evaluation of patients presenting with signs of early puberty, including detailed hormonal assays and imaging studies, to ensure accurate diagnosis and appropriate management. Increasing awareness among paediatric practitioners about the potential for such tumors can lead to earlier detection and better outcomes for affected patients.

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