

Education-Family Physician Corner

Mixed Corticomedullary Adrenal Adenoma in a Child

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We present a three-and-a-half-year-old female with an extremely rare tumor of the adrenal gland. There are less than 20 reported cases worldwide; it is mostly reported in adult females. The child presented with precocious puberty and the investigations revealed an adrenal mass. The tumor was resected. The morphology of the tumor suggested a mixed corticomedullary adenoma, which was confirmed with immunohistochemistry.

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Mixed corticomedullary adrenal tumors are very rare^{1,2}. Benign mixed tumor of the adrenal gland is an encapsulated mass, composed of an intimately intermixed population of adrenal cortical cells and pheochromocytosis³. The potential for malignancy should be considered. There is only one reported case of mixed corticomedullary carcinoma in the literature³.

The aim of this report is to present a rare case of an adrenal mass of mixed corticomedullary adenoma.

THE CASE

A three-and-a-half-year-old Bahraini female presented with precocious puberty. The patient's breasts and pubic hair were Tanner stage III. The child had normal blood pressure and no clinical evidence of glucocorticoid excess features. The bone age was four years advanced. The dehydroepiandrosterone (DHEA) level was elevated at >27 umol/L (normal range 0.27-1.63 umol/L). Both the follicle stimulating hormone (FSH) and the luteinizing hormone (LH) levels were normal. The estradiol was elevated at 157.2 pmol/L (normal level <130 pmol/L). The 24-hour urine vanillylmandelic acid was normal at 9.9 umol/24hours (normal range 10-50 umol/24hours). The cortisol level was normal based on a normal Synacthen stimulation test. The electrolytes including potassium and the glucose level were normal.

CT scan revealed a fairly defined right adrenal mass, measuring 5.7x4x3.7 cm. It contained multiple fine foci of calcification and a heterogeneous enhancement. No cavitation was seen within it. The superolateral border was not defined from the

liver. The mass displaced the kidney inferiorly, see figure 1. Consequently, the child underwent a resection of the right adrenal gland tumor. The tumor proved histopathologically to be a mixed corticomedullary adrenal adenoma.

The DHEA and the estradiol both normalized following the resection of the tumor. The breast regressed in size to Tanner stage II. The child is completely healthy six years after the resection.



Figure 1 (A): Well-Defined Right Adrenal Tumor with Tiny Calcification

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Figure 1 (B): Axial Section Showing Enhanced Adrenal Tumor

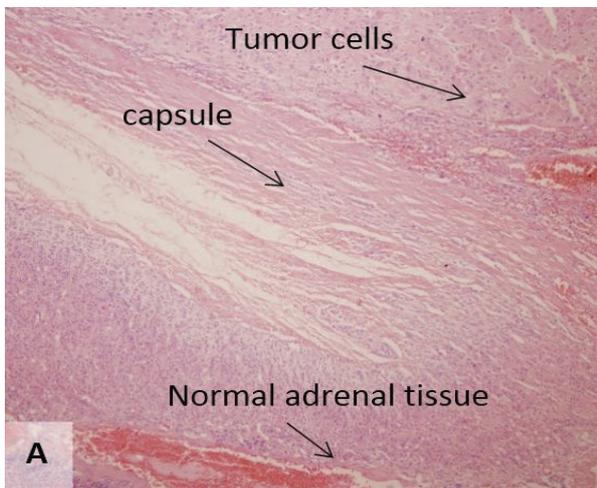


Figure 2 (A): Encapsulated Tumor with a Rim of Normal Adrenal Gland Tissue at the Periphery x40

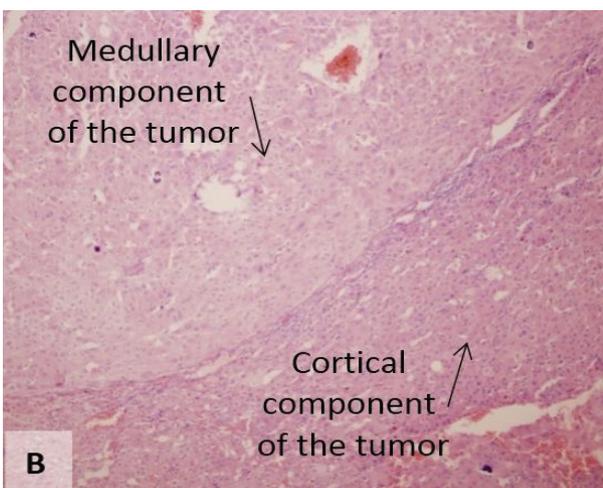


Figure 2 (B): Cortical and Medullary Components of the Tumor H and E Stain x100

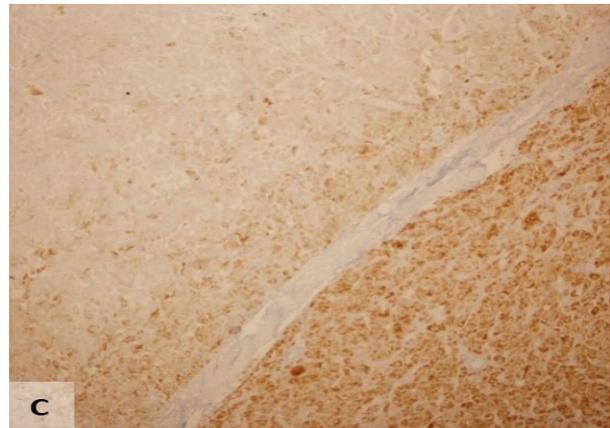


Figure 2 (C): Inhibin Positive in the Cortical Component of the Tumor x100

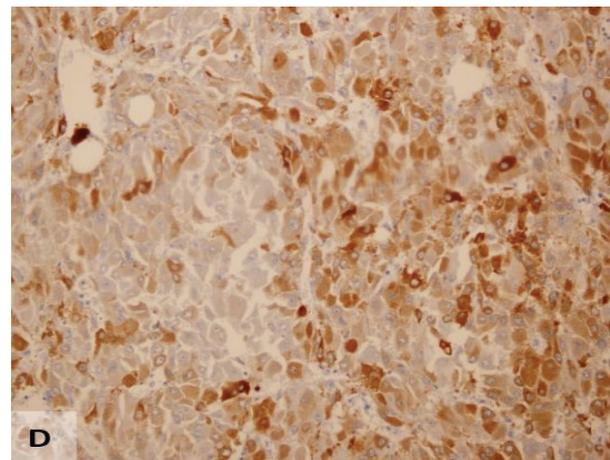


Figure 2 (D): Chromogranin Positive in the Medullary Component x400

Macroscopically, the tumor was an encapsulated greyish firm mass, weighing 30g and measuring 6x4x2.5 cm. The cut surface was fleshy in consistency and variegated in color. No macroscopic evidence of necrosis, hemorrhage or cystic degeneration was seen.

Microscopically, the tumor was well circumscribed and encapsulated, surrounded by a rim of the normal adrenal gland, see figure 2A. It showed an intimately admixed population of both adrenal cortical cells and pheochromocytes along with foci of calcification. No nuclear hyperchromasia, mitoses, hemorrhage, necrosis or vascular invasion was found.

Immunostaining revealed a focal positivity with calretinin and inhibin in cortical component and synaptophysin and chromogranin positivity in medullary component indicating an intimately admixed population of adrenal cortical and medullary cells. There was no predominant component.

DISCUSSION

Mixed corticomedullary adrenal tumors are described in less than 20 cases in the literature⁴⁻¹⁰. This rare adrenal tumor is characterized pathologically by a single well-circumscribed and encapsulated mass, with both cortical and medullary components.

The cases reported in the literature are in adults, who presented with hypertension or Cushing's syndrome. There are no reported cases of such tumor in children and in particular, the peculiar presentation in our case. The tumor in our case was a functional (hormone secreting) tumor, but most secreting tumors in children result in hypercortisolism or hyperaldosteronism, and rarely with virilization or feminization. In addition, the child had both excessive androgen and estrogen secretion which had resulted in both breast development and manifestations of adrenarche.

CONCLUSION

This is the first reported child with mixed corticomedullary tumor in the literature. The child's presentation was unusual compared with the few adults who were reported with the same tumor.

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