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Original Article

Adrenal tumors in children – Clinicopathological experience

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ABSTRACT

Objective: To document general baseline data on the patterns of childhood adrenal tumors.
Design, setting, and participants: This is a retrospective analysis of 10 cases of pediatric adrenal tumors (age group 0–12 years) in surgical pathology encountered over a period of 5 years.
Results: In the adrenal gland, adrenal medullary tumors were more common than adrenal cortex with neuroblastoma as common individual tumor. Two were tumors of adrenal cortex and 8 were of adrenal medulla with ratio of medullary to cortical adrenal neoplasms as 4:1.
Conclusion: Different types of tumors were seen in the adrenal gland. A high index of suspicion should be maintained with an aim of surgical treatment. Histological type is important for understanding etiology and progression of disease.

1. Introduction

Adrenal tumors occur rarely in childhood. Most antenatally detected suprarenal tumors are attributed to neuroblastoma. Neuroblastoma accounts for more than 90% of these tumors. Neuroblastoma along with ganglioneuroblastoma and ganglioneuroma constitutes a group of ganglion cell origin tumors that originate from primordial neural crest cells, which are the precursors of the sympathetic nervous system.[1] It is the third most common childhood cancer, after leukemia and brain tumors, and is the most common solid extracranial tumor in children.[2] The most commonly encountered pediatric adrenocortical tumors are adrenocortical carcinoma and adrenocortical adenoma. The adrenal cortex accounts for 6% of adrenal cancers in children, and adrenocortical carcinoma constitutes 0.2% of malignant neoplasms children less than 15 years of age.[3,4] Early diagnosis increases the possibility of curative treatment. As most adrenocortical tumors in children are hormonally active, they should be considered in the evaluation of a child presenting with abnormal hair growth, penile or clitoral enlargement, acne, hypertension, and rarely, unexplained constitutional symptoms. Furthermore, adrenal chromaffin cells contribute to pediatric neoplastic processes in the form of pheochromocytoma. The term pheochromocytoma is used generally to classify both intra-adrenal and extra-adrenal tumors that arise from chromaffin cells that may be located anywhere along the sympatho-adrenal system, including the adrenal medulla, Zuckerkandl body, paravertebral chain, hilum of the kidney and liver, aortic bifurcation, bladder,

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and mediastinum. Herein, pheochromocytoma shall designate tumor arising from the chromaffin cells within the adrenal medulla.

2. Materials and methods

10 cases of pediatric adrenal tumors (age group 0–12 years) were analyzed retrospectively over 5 years. Surgical specimens and biopsy tissues received were fixed overnight in 10% buffered formalin and submitted for processing. Paraffin sections were cut at 4–6 μm thickness and routine H&E staining was performed. All cases were reevaluated histologically on sections from routinely processed formalin fixed, paraffin embedded blocks. Special stains and immunohistochemistry were studied wherever necessary. The clinical, radiological, and therapeutic data were obtained from patients' case paper records.

3. Results

A total of 10 cases of adrenal surgical specimens presented over a period of 5 years. Of this, 6 were diagnosed as malignant tumors. Adrenal medullary tumors were more common than adrenal cortex with neuroblastoma (4 of 6 cases) as common individual tumor. Male to female ratio was 1:1. A single case of both adrenal adenoma and carcinoma presented at an age of 7 years within a male and female child respectively. There were two cases of pheochromocytoma in an 8-year-old female and 9-year-old male child with swelling and hypertension. Both of them showed elevated levels of VMA and catecholamines. Four of neuroblastoma cases were presented at the age of 11 months, 18 months, 3 years, and 5 years with male and female ratio of 1:1. One case of each ganglioneuroblastoma and ganglioneuroma at the age of 4 and 5 years, both in females, was documented presenting as suprarenal mass (Table 1).

4. Discussion

Neoplasms of the adrenal cortex both benign and malignant in the pediatric age group are rare. Adrenocortical tumors are very rare in children with a worldwide annual incidence of 0.3 per million children below the age of 15 years.[5] However, when present, they can cause many problems by excess secretion of certain hormones. In a review of the earlier literature, Hayles et al.[6] identified 222 hormonally active adrenal cortical neoplasms with a female-to-male ratio of 3:1, slightly over 50% of cases manifested in children at or before 3 years of age. Almost 80% of cases occurred under 8 years of age. Adrenocortical carcinoma accounts for less than 0.5% of pediatric malignancies. There is a female predilection (male-to-female ratio, 1:3).

The present study had 10 cases of the adrenal gland tumor. Two were tumors of adrenal cortex and 8 were of adrenal medulla with ratio of medullary to cortical neoplasms as 4:1. A single case of both adrenal adenoma and carcinoma at an age of 7 years within a male and female child respectively was documented. In contrast to adults, most children with adrenal cortical neoplasms have evidence of a functioning tumor at presentation with virilization as the most common manifestation, followed by Cushing syndrome and feminization.[6] An adrenal cortical neoplasm accounts for 50–70% of cases of pediatric Cushing syndrome. In the present study, adrenocortical adenoma presented with swelling, hypertension and facial puffiness. However, adrenocortical carcinoma presented only with the hypertension. The gross and microscopic features of adrenocortical neoplasms were studied in details. Adenoma on gross examination was of 4 cm × 4 cm, weight 40 g, well circumscribed and encapsulated, homogenous, soft and yellow. Microscopically, the cells of the adenoma were polygonal, large with clear cytoplasm in nests and cords. Adrenocortical carcinoma grossly has shown size of 15 cm × 12 cm, weight 650 g, nodular mass with solid, cystic, and necrotic areas. Microscopically, the growth pattern was diffuse sheets of tumor cells with bright pink cytoplasm and broad mitotically active cells separated by broad fibrous bands. Marked nuclear pleomorphism with areas of necrosis, hemorrhage and capcular invasion was also seen.

Cagle et al.[6] specifically studied the adrenal cortical neoplasms in children; they found that only size (expressed as weight) was a reliable predictor of malignancy, with a weight greater than 500 g indicative of a carcinoma. The survival of children with cortical carcinoma varies with stage at presentation and the completeness of surgical resection. In general, the overall survival rate is reported as 50% (with most metastases occurring within 2 years of diagnosis), with a higher survival rate in infants (53%) than in children older than 9 years of age (17%). Lynette et al.[7] studied six cases of

Table 1 – Clinical features of non-neuroblastic adrenal tumors.

Tumor	Case no.	Age in years	Sex	Duration of disease in months	Clinical presentation				VMA and catecholamines	CT/USG/MRI
					Swelling or lump	Hypertension	Facial puffiness	Pain		
Adrenocortical adenoma	1	7	M	11	P	P	P	A	NA	MRI – adrenal adenoma
Adrenocortical carcinoma	1	7	F	5	A	P	A	A	NA	CT – right adrenal tumor
Pheochromocytoma	1	8	F	2	P	P	P	A		CT – left suprarenal tumor
	2	9	M	2	P	P	A	A		CT – left adrenal mass