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Case Report

Adrenocortical Tumor In Children – Report of a Case

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Abstract
Adrenocortical carcinoma in childhood is a rare, potentially fatal disease.Adrenocortical carcinoma has a bimodal age distribution with cases clustering in children under 6, and in adults 30–40years old. We are presenting a case of 2 years old girl who presented with symptoms of virilization. She was operated for the adrenal mass. A diagnosis of adrenocortical carcinoma was rendered.

Key words: Adrenocortical Carcinoma in Children, Virilization, Adrenal Tumors

Introduction
Adrenocortical neoplasms are rare in the pediatric population, accounting for less than 0.2% of all pediatric tumors and 6% of all adrenal tumors in children.[1] adrenocortical tumors occur more frequently in girls, with a male to female ratio of approximately 1:2 to 1:3.[2]Most adrenocortical tumors in children are hormonally active, it may lead to either a virilizing or a feminizing features. Virilization with or without hypercortisolisim is the most common presentation.[3]These virilizing tumors may be more difficult to recognize in boys than in girls. Boys may present with precocious puberty, including penile enlargement, acne, and premature development of pubic, axillary, and facial hair. Girls may develop clitoral hypertrophy, hirsutism, and acne. The treatment of choice is adrenalectomy. Adrenocortical tumors are associated with several congenital anomalies, including hemihypertrophy; other tumors associated with hemihypertrophy include nephroblastoma and hepatoblastoma. Patients with Beckwith-Wiedemann syndrome (exomphalos, macroglossia, and gigantism) also have a higher than expected incidence of adrenocortical carcinoma.[4] Most adrenocortical tumors, however, occur sporadically.[5]

Case Report
A 2 years old female patient presented with history of excessive hair growth on body, increased weight gain, and polyphagia for 6 months duration. Her past medical or surgical history & family history were not significant. Finding on examination were hirsutism, moon face, central obesity with BMI 21.6(>95th).

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Her baseline investigations were normal and hepatitis profile was negative for both hepatitis B &C.Her Testosterone level was elevated (11.8 nmol/L). Her Serum ACTH, 17-OH progesterone & cortisol were within normal limits with negative Dexamethaon Suppressive Test. Her 24 Hrs urinary Cortisol & Vinyl mandalic acid(VMA) were performed to rule out neuroblastoma, that turn to be in normal limits. The ultrasound abdomen showed right supra renal mass with evidence of focal calcification. The CT scan showed a well-defined lobulated soft tissue mass lesion approximately 11X6 cm in right supra renal region, arising from right adrenal gland with multiple calcific foci.

So the patient had undergone for right side laprotomy and right adrenalectomy was done. A solid mass of 17X 10X8 cm was resected in Toto. There was no extension to Para adrenal area. Post operative recovery was uneventful. Child was well in follow up.

Gross Examination:

The tumor measures 17x10x8 cm, well circumscribed, of homogeneous appearance and exhibits areas of hemorrhage and necrosis, with fatty infiltration. (Fig 1, 2)

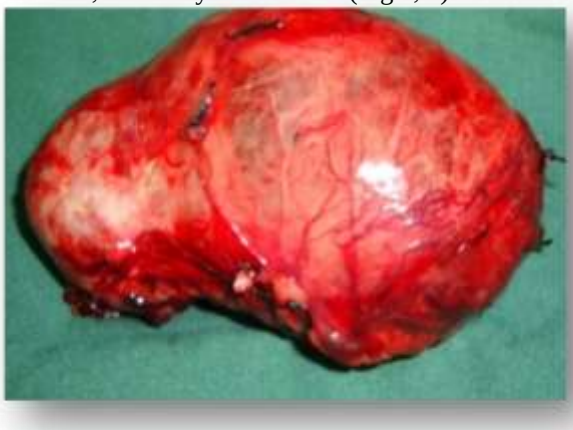

Fig: 1 External surface of the tumor

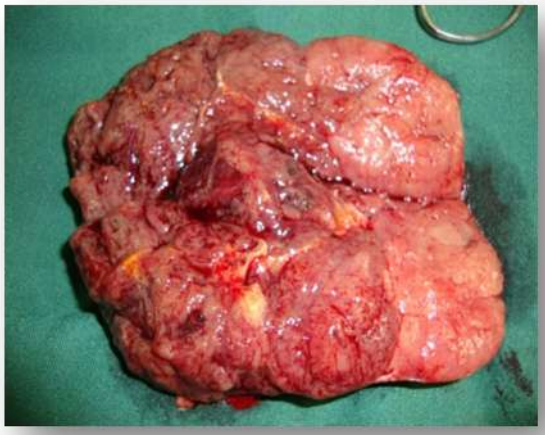

Fig:2 Cut surface of the tumor

Microscopic Examination:
There was prominent nuclear hyperchromasia,abnormal nuclear membrane contours and irregular distribution of chromatin and diffuse pattern of growth, and mitotic activity.(Fig 3-4)

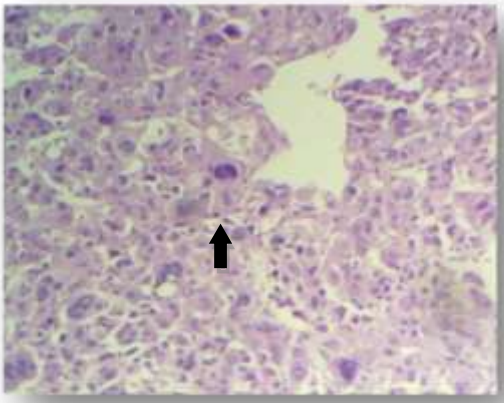

Figure: 3 Highly dysplastic cells with frequent mitosis; arrow indicates mitosis (H&E X 100)

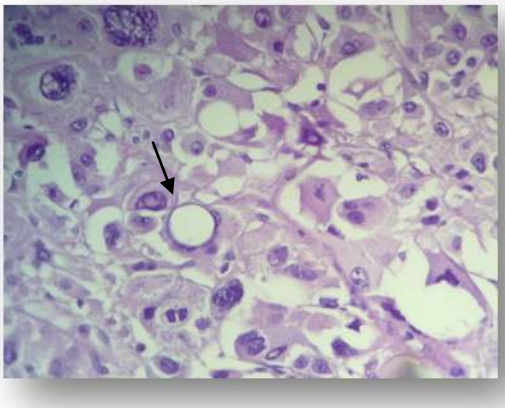

Fig: 4 Intracytoplasmic lipid vacuole (arrow) (H&E x 400)

Discussion

Adrenocortical carcinoma is rare. There is a female-to-male predominance of about 2:1. The tumors occur equally on the right and left sides and are hormonally functional in 80% of patients. [6]

Adrenal tumors can be classified as functional (FT) when their hormonal secretions result in clinical consequences: Cushing syndrome (CS), virilization syndrome (VS), feminization syndrome (FS), or a mixed Cushing-virilizing syndrome (CVS). Tumors are considered nonfunctional (NF) when the tumors do not secrete excessive hormones or produce hormonal precursors and/or active hormones in quantities insufficient to have clinical consequences. Such tumors still may secrete excessive amounts of steroids detected during laboratory evaluations.[7]

The staging system for patients with adrenocortical carcinoma originally proposed by MacFarlane,[8]and modified by Sullivan et al.[9]is used frequently. Patients with Stage I and II disease presented with tumor confined to the adrenal gland without local invasion or distant metastases and with a greatest tumor dimension of <5 cm (Stage I) or >5 cm (stage II), respectively. Patients with these tumors have the best chance of cure with surgical excision. When there is local tumor invasion that does not involve adjacent organs or regional lymph nodes, the tumor is considered to be Stage III. In Stage IV disease, there are distant metastases or invasion into adjacent organs plus regional lymph nodes. In the majority of reports of adrenal carcinoma, the tumors generally are in an advanced stage (Stage IV) of disease.

All children with virilizing features had elevated serum testosterone levels ranging from 30 ng/dL to 2300 ng/dL. Ultrasonography (US) of the adrenal gland has proven to be an effective technique for identification of adrenal masses but depends substantially on operator skills. Computed tomography (CT) generally is considered the method of choice in the radiodiagnostic evaluation of adrenal diseases. The location, size, and shape of adrenal masses and nodules as small as 1 cm generally are demonstrated using conventional CT techniques; however, using thin-section CT scanning, nodules as small as 3–5 mm can be identified.[10] Magnetic resonance imaging (MRI) improves adrenal imaging and provides prognostic help by appearance of the lesion on T1 and T2 imaging modes.[11]The adrenal cortical scintigraphy functional localization technique takes advantage of the accumulation of radiocholesterol into adrenal cortical tissues.

Inferior Vena Cava (IVC) invasion by tumors occurs with reasonable frequency in adrenocortical carcinoma, and the diagnosis can be established using US, CT, or MRI. Precise information regarding the presence and extent of IVC involvement is essential before deciding on surgical removal of thrombus.

The most common sites of metastases were the liver (85%), followed by lung (60%), bone (10%), and lymph nodes (10%), the latter more frequently in the retroperitoneum.[12]

The only curative treatment is complete surgical excision of the tumor, which can be performed even in the case of invasion into large blood vessels, such as the renal vein or inferior vena cava. Radiation therapy and radiofrequency ablation may be used for palliation in patients who are not surgical candidates.[13]

Chemotherapy regimens typically include the drug mitotane, an inhibitor of steroid synthesis which is toxic to cells of the adrenal cortex, [14] as well as standard cytotoxic drugs. A retrospective analysis showed a survival benefit for mitotane in addition to surgery when compared to surgery alone.[15]The two most common regimens are cisplatin,doxorubicin,etoposide + mitotane and streptozotocin + mitotane. It is unknown which regimen is better.

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