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Case Report

A Rare Case of Adrenocortical Carcinoma in an Infant

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Abstract

Background: Adrenocortical carcinoma might be regarded as an ultrarare disease, and it is even more rare to have a presentation as described, in an infant. The incidence is believed to be 1 to 2 per million per year, but valid data are lacking. Adrenocortical carcinoma typically presents late with a large mass. Recognition of the typical clinical, biochemical, and imaging findings is necessary for rapid diagnosis, prompt intervention, and early use of the appropriate therapy.

Case Presentation: Herein, we describe a rare case of adrenocortical carcinoma in an infant who presented with acne and pubic hair. Preoperative considerations, intra-operative technique, postoperative management, adjuvant therapies, and a brief review of the literature are discussed.

Conclusion: A multidisciplinary approach is integral in effectively diagnosing and managing adrenocortical carcinoma.

Keywords: Adrenocortical carcinoma; MRI; CT

Introduction

Adrenocortical Carcinoma is a rare malignancy with an incidence of 1-2 cases per 1 million population per year and a variable but generally poor prognosis [1]. A slight female predominance is observed (1.5:1) [1]. The age distribution is bimodal, with a first peak in childhood and a second between the fourth and fifth decade [1].

Case Summary

A one-year-old female child who came to the paediatric surgery department of Yenepoya Medical College Hospital with complaints of pubic hair since five months and acne since three months was admitted and evaluated. Routine investigations were within normal limits. A unilateral adrenal mass was diagnosed by ultrasound. Serum Thyroid Stimulating Hormone 9.29 (0.7-6.4 mIU/L), Free T4 0.75 (0.7-2.1 ng/dl), Dehydroepiandrosterone Sulfate 24.7(32.7-276 ug/dl), Luteinizing hormone <0.10, Estradiol 21.00 pg/ml, 8 am cortisol 1.79(4.4-22.7 ug/dl). Computed Tomography abdomen showed a large, well-defined, heterogeneously enhancing soft tissue attenuating lesion with few non-enhancing areas of calcification seen arising from the left suprarenal gland measuring 6.8 x 5.7 cms- suggestive of neuroblastoma. Ultrasound guided Fine Needle Aspiration Cytology from the swelling showed features suggestive of neuroganglioma. Surgical excision of the mass was performed via an open anterior transperitoneal approach, and the entire adrenal gland was removed. The intraoperative and postoperative period was uneventful, and the patient was discharged. Histopathology of the mass was reported as adrenal cortical carcinoma. The patient is doing well in the recent follow-ups, and there is no recurrence.

Discussion

The adrenocortical carcinoma is a rare type of paediatric tumour and represents <0.5% of all childhood neoplasms and 6% of all paediatric adrenal tumours [2,3].

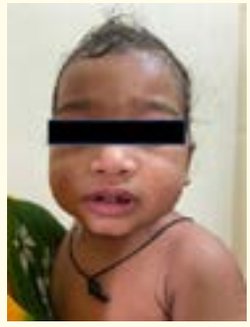

Figure 1: Clinical picture of patient showing acne.

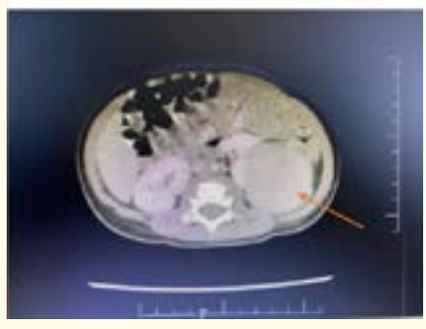

Figure 2: CT image showing left suprarenal mass.

Predisposing inherent genetic factors have also been identified in ~50% of infants exhibiting Adrenocortical carcinomas. Furthermore, two genetic syndromes, Li-Fraumeni and Beckwith Wiedemann, are associated with these [2,3].

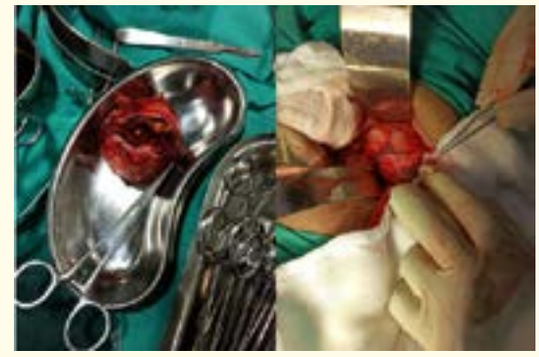

Figure 3: Specimen and intraoperative picture.

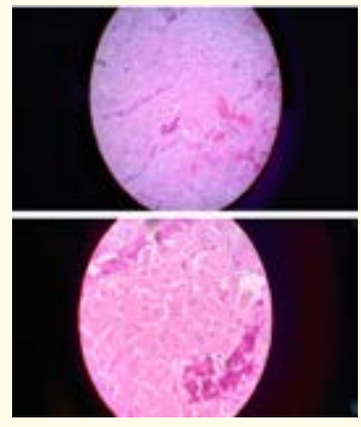

Figure 4: Histopathology slides.

TNM Staging for Adrenocortical Cancer			
Tumor	**Node**	**Metastasis**	**Stage**
T1	N0	M0	I
T2	N0	M0	II
T1	N1	M0	III
T2	N1	M0	III
T3	Any N	M0	III
T4	Any N	M0	III
Any T	Any N	M1	IV
Primary Tumor (T): T1, size<=5 cm without local invasion; T2, size >5cm without local invasion; T3, any size with local invasion but no involvement of adjacent organs; T4, any size with involvement of adjacent organs. Nodes(N): N0, no involvement of regional nodes; N1, positive regional lymph nodes. Metastasis(M): M0, no known distal metastases; M1, distant metastases present.			

Table 1: TNM staging of Adrenocortical Carcinoma.

Cushing's syndrome is the most frequent symptom of raised hormone levels; however, the feminizing and masculinizing syndromes are less common and present as the initial manifestation of Adrenocortical carcinoma in only 2.2% of cases. Adrenocortical carcinoma often secrete a variety of hormones and, thus, presents the signs and symptoms of multiple syndromes (mixed forms).

Non-functional tumours comprise around 10% of paediatric cases worldwide [4]. However, a significant clinical manifestation observed in patients with functioning Adrenocortical carcinomas and androgen and estrogen overproduction is growth disturbance in terms of overgrowth [4]. The implications of recognizing this as a symptom of Adrenocortical carcinoma are significant and may facilitate the early diagnosis of the tumour before the onset of other tumour-associated symptoms.

CT and MRI scans are helpful in these tumours. The size of the adrenal mass on imaging studies is the most important criterion to help diagnose malignancy. Other CT imaging characteristics suggesting malignancy include tumour heterogeneity, irregular margins, and the presence of haemorrhage and adjacent lymphadenopathy or liver metastases. FDGPET or PET-CT scans may have some utility in distinguishing benign from malignant lesions. Once adrenal cancer is diagnosed, CT scans of the chest and pelvis or FDG-PET or PET-CT scans are performed for staging. The tumour-node-metastasis (TNM) staging system for adrenocortical carcinoma is depicted in Table 1 [5].

The role of the pediatric surgeon is particularly significant in the pathology of Adrenocortical carcinomas, and surgical management must be considered for all cases of childhood Adrenocortical carcinoma. Numerous studies have proposed open transperitoneal resection as it is associated with a decreased risk of bleeding, improved visualization of the tumour boundaries, and it allows for the inspection and biopsy of the contralateral adrenal and adjacent lymph nodes. Thus, an open transperitoneal resection was conducted in the present case.

The histological features of this type of tumour remain controversial. The pathologist usually determines the definitive diagnosis and prognosis of the tumour; however, in paediatric adenocarcinoma, the pathologist's role is different. Due to the rarity of paediatric Adrenocortical carcinoma, the experience of pathologists is limited; thus, it is difficult for pathologists to establish a diagnosis. The histological features are not considered markers of malignancy in infants, and due to the heterogeneity and rarity of Adrenocortical carcinomas, the prognostic factors have been challenging to establish.

Paediatric surgeon along with expert opinion by an endocrinologist, radiologist as well as an experienced pathologist is required in adequate management of similar cases of Adrenocortical Carcinomas. Starting from the diagnosis, hormonal assessment till the final specimen histopathology.

Prognosis is dependent on numerous factors. A young patient (<3 years), presenting with symptoms within six months of onset, exhibiting a small mass (<150 g) who receives an open complete resection, and demonstrates negative results from histological analysis of the lymph nodes and does not exhibit metastasis is a typical case with an optimal prognosis [6].

Chemotherapy with Mitotane: Mitotane is the only adrenal-specific agent available for the treatment of Adrenocortical carcinoma. Mitotane exerts a specific cytotoxic effect on adrenocortical cells, producing focal degeneration of the fascicular and particularly the reticular zone, whereas changes of the zona glomerulosa are relatively slight. Cytotoxic chemotherapy by combining mitotane with etoposide, doxorubicin, and cisplatin is useful for advanced Adrenocortical cancer [7].

As described in the TREP (Italian Pediatric Rare Tumor) guidelines, the postoperative evaluation involves USG or MRI of the site of the excised tumour and hormone profiling. Follow-up, which will comprise the same analysis, will be performed at two-month intervals for the initial two years, at three-month intervals in the third year, at four-month intervals in the fourth year, and annually from the fifth year onwards. However, the infant in the present case did not follow up yet [8].

Conclusion

The current case report shows a unique presentation of Adrenocortical carcinoma and may be beneficial in increasing the knowledge of Adrenocortical Carcinomas. Furthermore, the present report aims to highlight the importance of a multidisciplinary approach, which is essential in early diagnosis and treatment.

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