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Case Report

Suprarenal neuroendocrine tumour with intrathoracic extension in a child

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Introduction

Neuroendocrine tumours (NETs) are a diverse group of neoplasms with an incidence of 6.98/100,000 population. NETs are rare in children [1,2]. Common anatomical sites are the gastrointestinal tract and lungs. Occurrence in adrenal glands is rare. Majority of NETs originate sporadically. Some secrete hormones [1,2].

We report a right suprarenal NET extending through a diaphragmatic hiatus up to the hilum of lung in a three-year-old healthy girl with nonspecific central abdominal pain.

Case report

A healthy 3-year-old girl with vague central abdominal pain of four months' duration had an USS abdomen. This detected a right suprarenal solid tumour. CT scan identified a 3.8cmx4.5cmx7.3cm size, non-metastatic, right suprarenal neuroblastoma confined to the adrenal gland (MRI facility was not available). Tumour was seen pushing the inferior vena cava (IVC) forwards, without encroaching on it and extending up to the aorta. USS guided tumour biopsy diagnosed a benign neurogenic tumour. Tc99 bone scan was normal.

A multi-disciplinery team decided on primary tumour excision. This was the first such major paediatric surgery conducted at Teaching Hospital, Ratnapura which was the only paediatric surgical centre in the Sabaragamuva Province of Sri Lanka.

The team of oncological, vascular and paediatric surgeons approached the tumour via a standard right upper-transverse laparotomy [3]. A suprarenal tumour was found, occupying the entire suprarenal and retro-hepatic segments of the IVC, completely covering its right and posterior surfaces. Posterior part of the tumour extended up to the aorta.

Contrary to the CT findings, the tumour was found attached to the medial end of the right hemi-diaphragm, extending beneath the right crus into the thoracic cavity. Thoracic dissection through the diaphragmatic hiatus, although challenging, commenced to avoid the additional surgical trauma of a thoracotomy. The IVC together with the liver was mobilised towards the left side, facilitating dissection superiorly. The tumour extended beside the right pericardium up to the right pulmonary hilum. No intra-thoracic structure invasion or lymphadenopathy was identified. Rest of the abdominal viscera were normal.

The tumour was totally excised, together with a segment of the right hemi-diaphragm and the right-crus according to oncosurgical principles [3]. Liga-clips were applied to tumour excision margins to facilitate interpretation of follow up imaging. The child was electively ventilated and weaned off. She made an uneventful recovery 12 days after surgery.

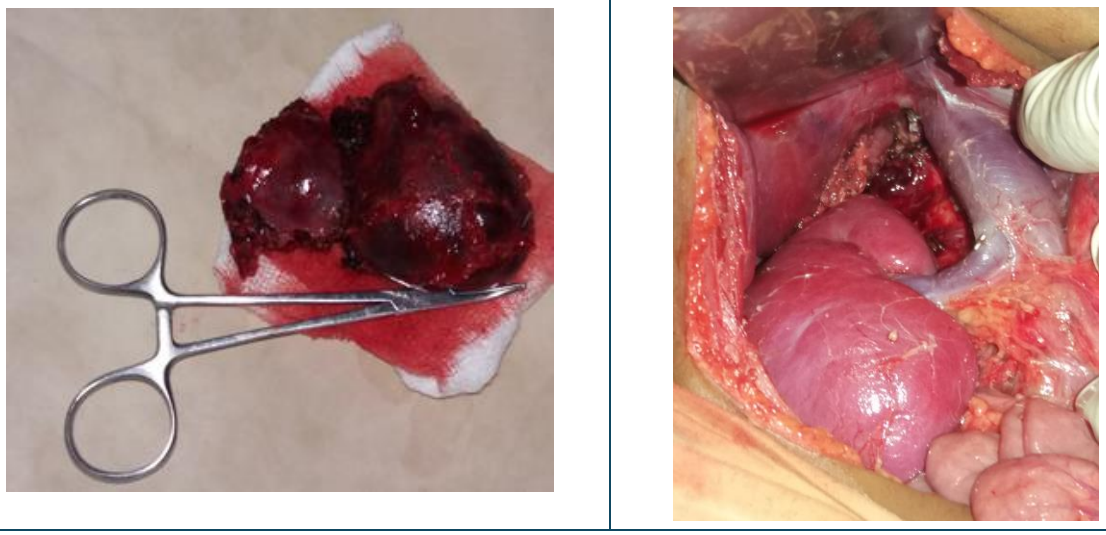

Figure 1: Excised tumour with post- excision operative field

Histology confirmed a totally excised, benign, encapsulated, neuro-endocrine tumour with nuclei with 'salt-and-pepper' appearance. Mitosis was inconspicuous. Immunohistochemistry, chromogranin, CD56 and CD99 were negative. Ki67 index=25%. The tumour was classified as Grade 3 WHO NET [1,2]. Serial follow-up imaging and

tumour-marker analyses under oncological supervision remain normal. She is healthy nearly four years after surgery.

Discussion

CT scan has 82% sensitivity and 86% specificity in diagnosing NETs [2]. Management of this radiologically detected suprarenal tumour commenced with USS guided biopsy for histology. Ideal management of WHO grade 3 NET is total excision followed by surveillance [1,2,3]. Incomplete tumour excision warrants adjuvant therapy. Despite unexpected operative finding of intra-thoracic extension, total tumour clearance avoiding thoracotomy was achievable because of the expertise of a multi-disciplinary team comprising of clinicians with an interest in NETs in children.

NETs with Ki67 index >20% or mitotic count 20/10HPF (whichever is higher) are classified as NET-Grade 3 [1,2]. This tumour with Ki67 25% (despite inconspicuous mitosis) was classified as a WHO Grade3 NET.

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