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Case Report

Case Report of Large and Symptomatic Collision Low Grade Glial and Glioneural Tumor Originated in Adrenal Mass with Complex Extension in Early Infancy: Difficulties in Management, and Surgical Decision Making

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Abstract

Background: *Large and rapidly growing abdominal tumor may result in fatal outcomes in early infancy. In this case, a rapidly worsening clinical condition requires surgical decision-making despite the presence of a precise histological diagnosis.*

Case: *We highlight our experience with 3 month old 4 .5 kg female infant with large abdominal tumor and assess the available literature for the best possible management of a rare condition. Conclusion: In these cases, laparotomy should be considered as a life-saving procedure with challenging pre , intra, and postoperative complication*

Keywords: *abdominal compartment syndrome, case report, congenital cystic disorder.*

Introduction

Solid neonatal and infancy tumors are rare, representing less than 2% of childhood tumors.(1) Among them, large and rapidly growing abdominal tumors can seriously endanger the lives of newborns. There is a lack of consensus in managing these situations due to their rarity and diagnostic difficulty. Uncertainty in the nature and extent of the lesion despite well-conducted imaging studies further question whether a correct diagnosis was made. The optimal timing for surgery remains questionable due to the possibility of the evolution of homeostasis, ventilatory state, and concerns about oncological and long-term prognosis.

Case Series

A 50 days old female gender infant who is product of consngious marriage and full term caesarian section delivery presented by huge right sided abdominal mass causing marked abdominal destination. Abdominal ultrasound revealed a huge right sided retroperitoneal mass of right supra renal origin seen as complex multi-lobulated cystic mass with central calcification and size 15 by 11 by 10 cm in dimension. This mass displacing the liver anterior and to the left and displaces the right kidney inferiorly to the region of the pelvis. Anteriorly it displaces the intestinal loops. It crosses the midline and displacing the great vessels to

the left. Complementary PETCT done and revealed positive FDG avid RT solid and cystic huge mass suvmax 12. Ultrasound guided biopsy taken by senior interventional radiologist emphasizing that core of tissue taken from the wall of the solid component of the huge cystic solid mass. Pathology team described that this core of tissue showed an embryonal tumor consisted of neuroblasts amidistscanty amount of fibrillary eosinophilic stroma (neuropil). Mitosis index is low and this core of tissue is positive to stain by synaptophysin confirming diagnosis of schwannian stroma poor neuroblastoma, favorable histology tumor (INPC) and no amplification of NMYC gene. Furtherly the case boarded and in view of being inoperable chemotherapy for 2 courses of chemotherapy as per risk adapted neuroblastoma protocol given. Follow up of this mas after the 2 courses of this combination chemotherapy on clinical and ultrasound base showed clinical deterioration in term of increasing the abdominal girth and size of swelling. Patient urgently admitted for symptom control but further clinical deterioration in term of marked respiratory distress and mental state change. Urgent laparotomy done aiming at decompression or successful resection.

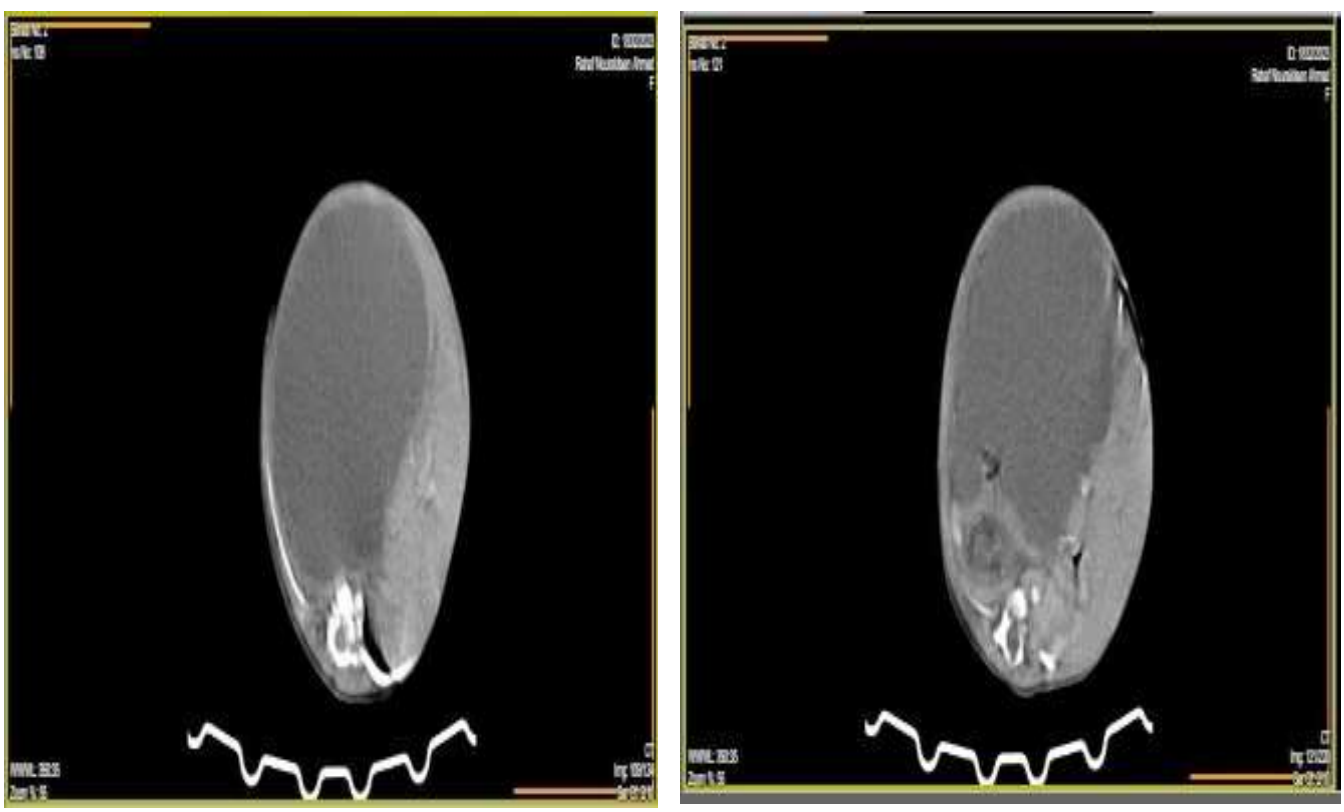

Figure 1: huge RT sided abdominal multi-loculted mass cystic with central focal dense calcification originated from RT adrenal gland.

Surgical strategy, anesthetic concern and post-operative care:

Once this patient had arrived to operative theatre there was huge risk for respiratory failure so caudal analgesia beside general anesthesia had adopted this plan aimed at avoiding postoperative atelectasis due to pain of the extended incision also postoperative resuscitation through central line and complete hemodynamics monitoring. As it was unclear that decompression could lead to improve prognosis rapid and careful resection of the mass with the right kidney after mobilization of the bowel and securing the great vessels done followed by hemostasis and insertion of two drain then closure of the incision done. Upon arrival the PICU she had satisfactory hemodynamics and spontaneously breathing and accepted laboratory panel expect for mild respiratory acidosis. The respiratory acidosis was spontaneously corrected after short period of time. Few hours later her blood pressure started gradually declined to less than for her age normal range although her central venous pressure was close to 7 also capillary refill was normal. We immediately started fluid resuscitation by crystalloids. although repeated boluses of crystalloid given blood pressure failed to attain the normal for her age so inotropic support as per dopamine at 10 u /kg then treated doses of adrenaline up to 0.3 u /kg /mi till blood pressure attained target off 88 /55 and mean blood pressure of 60 mhg. inotropic support then withdrawn and patient peacefully discharged with satisfactory lab panel and removal of surgical drains. Patient seen at OPD for wound care that had normal healing and post-operative CT documenting that was no residual of the swelling. Stitches were removed and patient discharged.

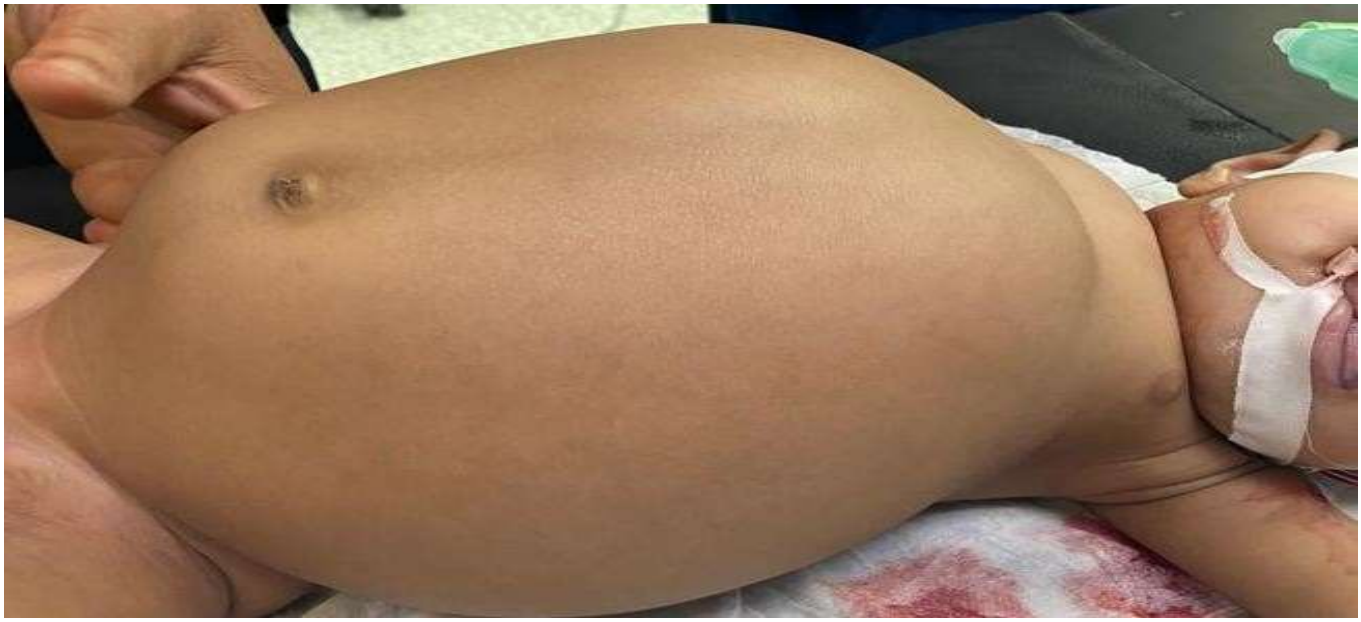

Figure 2 huge distended abdomen with marked respiratory compromise due to thoracic cage compression

Figure3 large specimen of post-operative excision of the complex mass en block with adherent not separable from the kidney

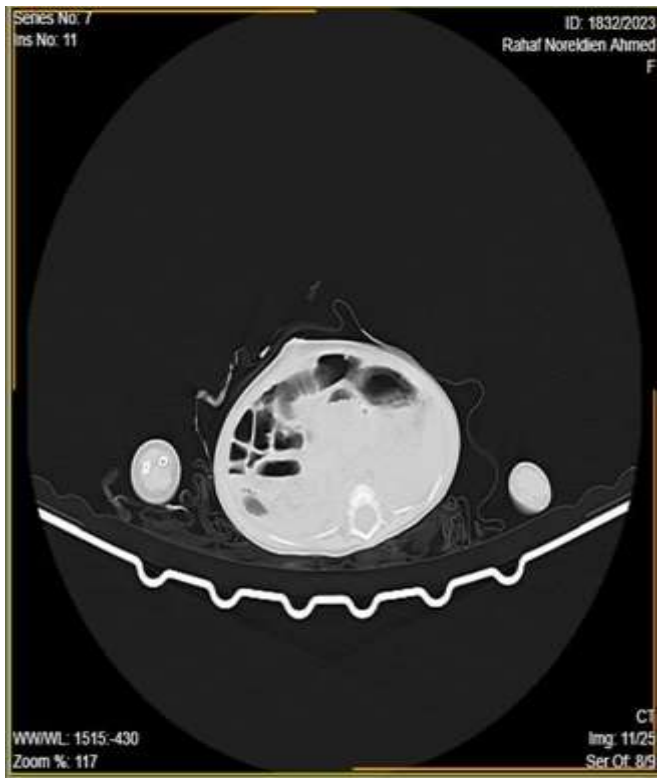

Figure 4 MSCT abdomen pelvis showed clear postoperative bed of the previous mentioned Rt adrenal mass.

Pathology of postoperative specimen and further oncologic plan

Gross pathology revealed Mass 18x15x12 cm attached to kidney 6x4x2.5 cm. Outer surface of mass shows a semilunar rim of golden yellow adrenal tissue 1x0.5 cm. Cut section of the mass shows a multinodular mass within adrenal gland. Mass shows greyish soft areas 0.5-1.5 cm as well as hard bony areas 9x8 cm. Capsule is intact. Cut section of kidney is negative for gross tumor. Soft parts were processed in seven blocks and different parts of cyst were processed in eight blocks and kidney was sampled in two blocks. Microscopic examination revealed Extensive sampling from cyst shows a teratoma with tissues from the three germ layers including skin, glandular epithelium, mature smooth muscles, mature adipose tissue, and mature brain with ependymal tissue, mature bone and cartilage. No evidence of immature elements. Extensive sampling from the soft parts (blocks 2A-2G) revealed a biphasic tumor composed of an admixture of neuronal and glial elements. In some sections the two components are intermixed and in other are

geographically separate. Tumor shows dysmorphic ganglion cells that appear to demonstrate many foci of abnormal clustering with lack of cytoarchitectural organization. Nuclei are focally enlarged. The glial component is composed of proliferating cells some resemble fibrillary astrocytoma and others resemble oligodendroglia. There are wide areas of dystrophic calcification with related necrosis. Scattered micro-calcific (psammoma) bodies are also seen. Sections from adrenal gland show unremarkable cortex and atrophic medulla. Sections from kidney are also unremarkable. In view of pathological findings encountered in the surgical excision specimen, recuts from previous biopsy were made and additionally stained for GFAP and Ki67. GFAP was positive and Ki67 labels about 12% of the nuclei indicating an origin from the observed glioma within this huge teratoma. Thus in view of these new facts, the previous diagnosis of neuroblastoma is retracted. Final diagnosis is Collision low grade glial and glioneural tumor (subependymoma & ganglioglioma) arising in intra-adrenal mature cystic teratoma.

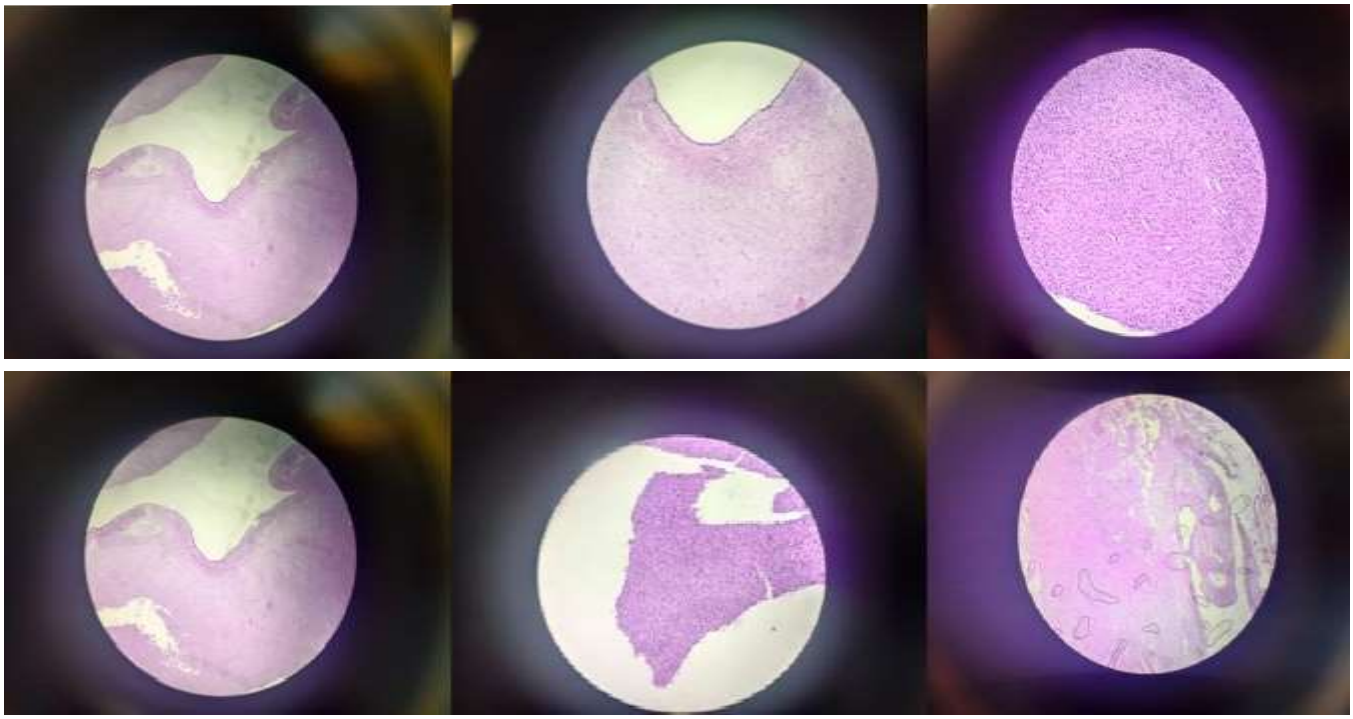

Figure 5: Serial H and E section of adrenal mass showed teratoma with tissues from 3 germ layers

Reference

1. Orbach D, Sarnacki S, Brisse HJ, et al. Neonatal cancer. Lancet Oncol. 2013;14:609-620. doi:10.1016/S1470- 2045(13)70236-5

2. European Low and Intermediate Risk Neuroblastoma Protocol: A SIOPEN Study Version 8.0. 8 June 2021. EudraCT Number 2010- 021396-8

3. European Low and Intermediate Risk Neuroblastoma Protocol: A SIOPEN Study Version 8.0. 8 June 2021. EudraCT Number 2010- 021396-8.