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Case report

Series of rare diagnosis and presentations of abdominal mass in children: a great learning experience

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ABSTRACT

Abdominal mass is a common finding in children, either palpable or radiologically evident surprisingly. Some of them are rare tumors. Functional active tumors are rarely palpable but present with varied arrays of symptoms. In this series authors are discussing few rare cases with atypical presentations such as- teratoma arising from adrenal gland, teratoma presenting with hypertension, fetus in fetu (Girl and Boy child), adrenocortical tumor presenting as precocious puberty and adrenal pheochromocytoma with features of cushing's syndrome. These atypical presentations may have pose a challenge in diagnosis and management for the treating team with first time occurrence specifically if they are handling them for first time.

Keywords: Abdominal mass in children, Adrenal tumor, Fetus-in-fetu, Teratoma

INTRODUCTION

Abdominal mass in children is a very common entity yet do get surprises when dealing with them. Sometimes the condition itself is rare and sometimes a not so rare condition can present in an atypical manner that catches us with a surprise. In a span of few years, authors had successfully managed a few such cases which was great learning experience and authors felt worth sharing. Authors had teratoma arising from adrenal gland, teratoma presenting with hypertension, fetus in fetu (girl and boy child), adrenocortical tumor presenting as precocious puberty and adrenal pheochromocytoma with features of Cushing's syndrome in children which are either rare or atypical to present with.[1-5]

CASE REPORT

Adrenal tumor

Case 1

A 7½ years old girl presented at OPD with ill-defined dull aching pain abdomen for 10 days without any associated symptoms. On further enquiry mother confirmed a swelling in right side of her abdomen since 2 years of age for which she was advised surgery but as the child was asymptomatic they did not follow up further. On examination her blood pressure was normal. There were no cervical lymphadenopathies, hepato-splenomegaly. A non tender firm flank mass was palpable in right side of abdomen. Radiology (CECT) (Figure 1).

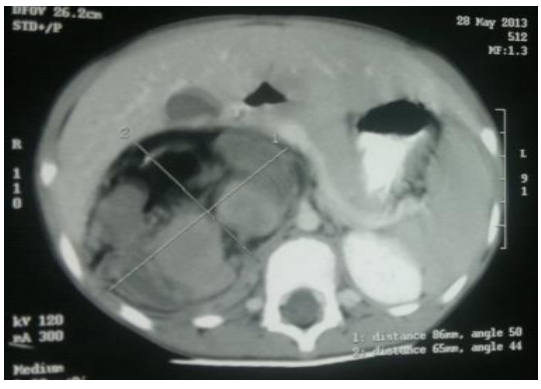

Figure 1: CECT Abdomen shows Rt Adrenal mass.

confirmed a right sided Adrenal mass of size 5cm × 7cm ×7cm. Laboratory values were all normal including tumor markers (AFP and Beta HCG) not elevated. Surgery was planned and after excision HPE report confirmed the diagnosis of adrenal teratoma. Post operative follow up was uneventful.

Case 2

A 4 years old girl presented with uncontrolled hypertension and facial puffiness with history of unconsciousness and vomiting (single episode). Investigation revealed a right side adrenal SOL of size 2.2cm ×3cm ×4 cm without any vascular involvement. Blood pressure was 124/74 mmHg (>95th percentile). Laboratory values did not match with classical pheochromocytoma with Urinary VMA 2.14mg/24hr (<2.20), Free metanorphine 12 pg/ml (<65) and elevated DHEA 62.5 µg/dl (<38) and Androstenedione 0.80ng/ml. Other values were normal including Blood Sugar, ALP, TSH, ACTH, Serum cortisol in (High and Low dose dexamethasone suppression test) and Testosterone. After preoperative stabilization of hypertension and intra-operative monitoring by expert anaesthesia team, right adrenalectomy was done and specimen was sent for (histopathology). Postoperative recovery was uneventful. HPE suggested features of Pheochromocytoma. IHC was atypical for Pheochromocytoma Melan A very focal faint positive, CD 56- patchy positive, Ki 67- less than 2% (NEGATIVE for chromogranin A, Synaptophysin, S100, Inhibin Alfa). Post operative follow up showed decreased facial puffiness and blood pressure was optimized with reduced number and dose of antihypertensive (Figure 2).

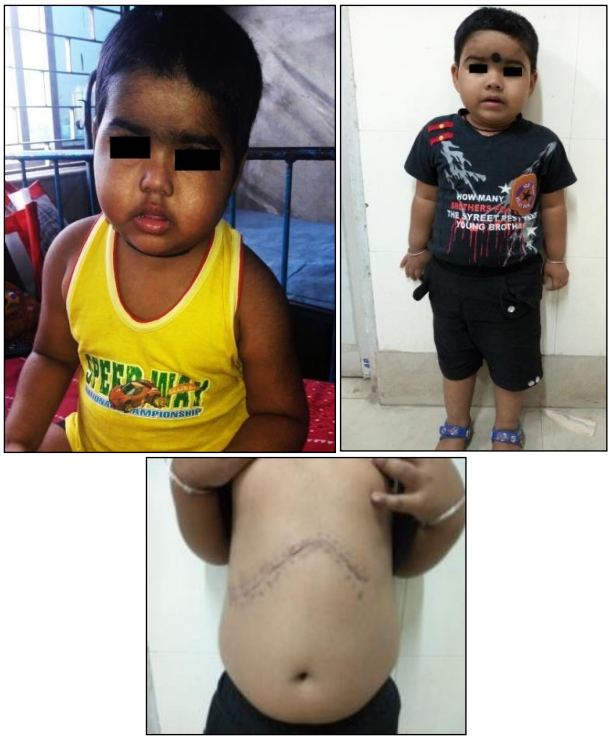

Figure 2: Preoperative and post operative facial changes, inset scar mark.

Case 3

A 6 years old Boy was brought to OPD with complaints of pre-mature appearance of pubic hair and increase in phallus size for last 2 months. On examination there was presence of curved and dark pubic hair, normal and well developed scrotum and testis and an unusual long Penis compared to age (Figure 3).

Figure 3: Enlarged phallus size with pubic hair.

Stretch penile length was 8.2 cm (expected 4.5cm) and mid-penile diameter 2.4cm (expected 1.2cm). Anthropometry measures body weight 30.2 kg (expected 20.7), height 1.2 m(expected 1.16m) a mid-arm circumference 23 cm with a BMI of 20.3 kg/m^2. Tanner Stage 3 was made based on Pubertal Maturation. Investigations revealed a Right Adrenal gland SOL measuring 5.5cm x 4cm× 4cm without any evidence of calcification, necrosis, lymphadenopathy (para-aortic) or renal parenchymal involvement. A provisional diagnosis of Virilizing Adrenocortical tumor (probably malignant) was suspected and Rt sided adrenalectomy was done. HPE report was Adrecortical Adenoma.

Teratomas

Case 4

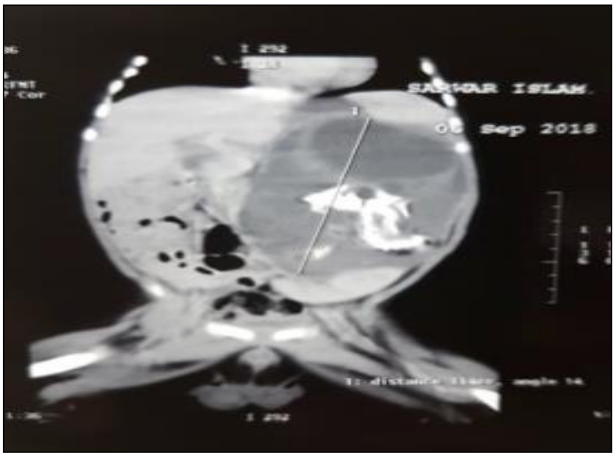

Figure 4: CECT whole abdomen showing features of teratoma.

Another boy of 6 months age presented with a lump in left flank for 2 months. The size of the lump was increasing gradually and the child became irritable.

This baby boy was born of caesarean section from a primi mother. His bladder and bowel habits were normal. A firm immobile retroperitoneal lump in left flank crossing the midline was palpable in his abdomen and radiology (CECT) suggested a retroperitoneal teratoma with both solid and cystic component (Figure 4).

Laboratory values was unremarkable except elevated LDH. Surgery was done and after removal of mass hair , bone (vertebra like structure) and intestine like tubular structure was identified (Figure 5) and radiology of the specemen confirmed bony component (Figure 6).

Figure 5: Gross features of excised specemen.

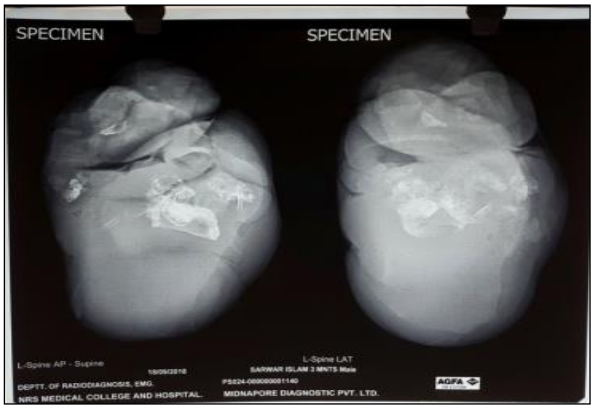

Figure 6: Radiology of excised specemen shows bone and vertebra.

Thus "Fetus -in- fetu" was diagnosed.

Case 5

Among 4 months old girl presented with abdominal distension with palpable lump in abdomen.

Though the baby was asymptomatic investigation revealed a well defined, lobulated, heterogenous lesion arising from adnexa with fat density and internal calcification measuring 9.7cm× 7.7cm× 11.7cm (Figure 7).

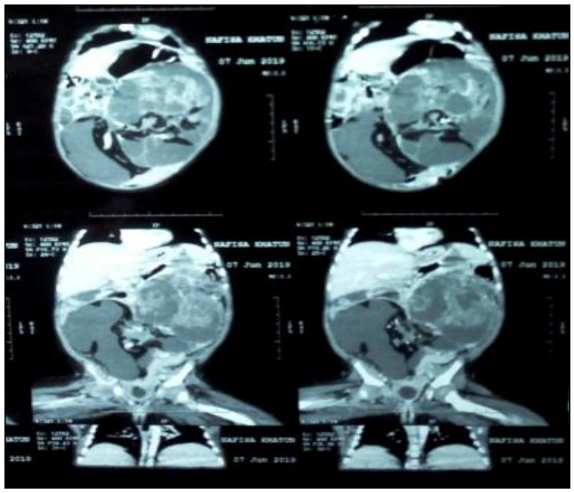

Figure 7: CECT whole abdomen.

CECT also suggests post contrast homogeneous contrast enhancement. The mass pushed the bowel up and anteriorly (Figure 8).

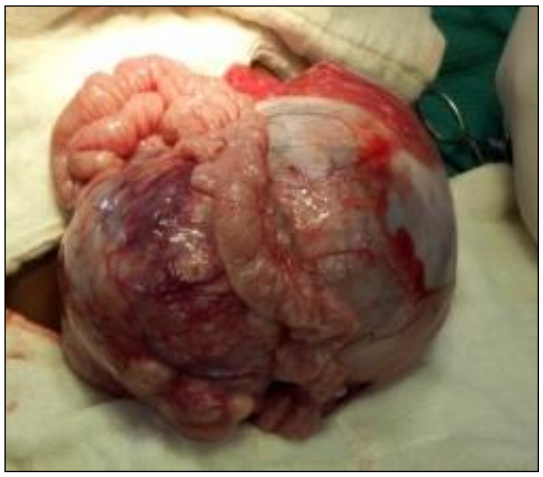

Figure 8: Excised specemen fetus in fetu.

After surgery a retroperitoneal mass with intact bone and bowel like structure was found and and on HPE it was diagnosed as **"**Fetus -in- fetu".

Case 6

Another 7 months old girl presented with a lump in left flank which was present for last 2 months. It increased in size over last 1 month. The girl was hypertensive (BP 180/120 mmHg) and had to be managed with 4 anti hypertensive medications- Amlodepine, Labetelol, Clonidine and Enalapril.

The antihypertensive medicine and doses were regulated and titrated gradually to achieve an optimum normalized blood pressure.Radiology confirmed a well defined left

supra renal mass with septation, calcification, cystic fatty element (Figure 9).

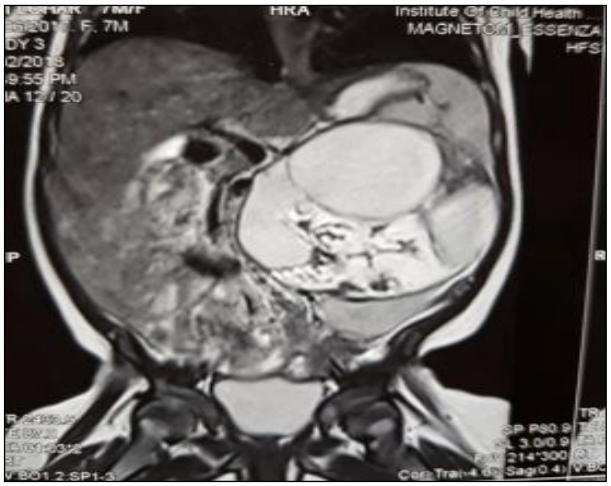

Figure 9: CECT whole abdomen.

MR Angioreconstruction showed a huge mixed intensity mass in left upper abdomen (11x10x10 cm) and left renal vessels showed adaptive elongation (Figure 10).

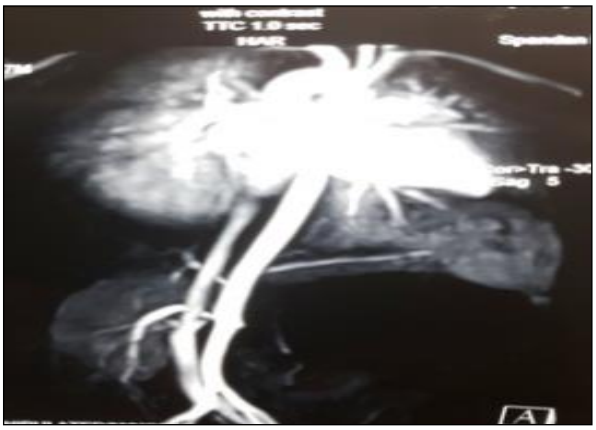

Figure 10: MR angiography showing adaptive changes in lefty renal vessels.

Nuclear scan (DTPA and DMSA) showed a non functioning left Kidney. Urinary VMA (1.52/200 ml of urine),B HCG (0.52), AFP (17.19 ng/ml) were normal and LDH (321 U/L) was elevated.

Surgery was done and Specemen in Gross was a Single globular soft tissue measuring 12x7x5.5 cm with attached tubular structure (? intestine) measuring 5cm in length (Figure 11).

Cut sections were partly solid and partly cystic. HPE confirmed Mature Teratoma.

Postoperative period was uneventful and Blood pressure was optomized by reduced number and doses of antihypertensives. 1 year after surgery BP (80/50 mmHg) was maintained by Clonidine (25mcg,QDS), Enalapril (1.25mg,BD) and Labetelol (50mg,BD).

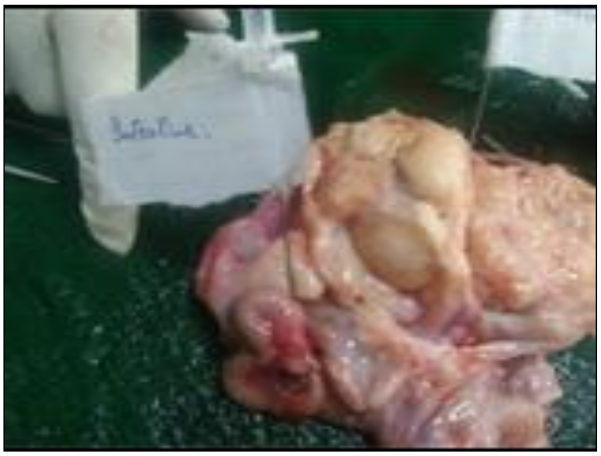

Figure 11: Gross specemen after excision.

DISCUSSION

Authors are discussing all 6 cases of lump abdomen in children in two broad groups - adrenal tumors and teratoma (Table 1).

Incidence of germ cell tumors has been estimated at about 0.9/100,000 population.[6] Teratoma has been reported as the leading neoplasm.[7,8] Only 4% of Germ Cell tumors are retroperitoneal or intrabdominal. Mostly diagnosed during infancy or less than 5yrs age. (CCG/POG). Histologically most are mature > immature teratomas. 0-24% reported malignancy (63% Yolk Sac tumor). Benign tumors have a tendency to encase major vessels and vascular and visceral injuries are reported and thus surgery is a challenge.

The adrenal gland as the origin of a teratoma is extremely rare.[6-10] Adrenal teratomas have no specific clinical manifestation.[8,9] They are often found on ultrasonography examination. As the findings from laboratory examinations will often be normal, a postoperative pathologic examination required for a definitive diagnosis.[9] Surgical complete resection and close follow-up recommended for mature teratomas.[10,11] For Immature teratoma, adjuvant therapy, will also be necessary.[12] For a mature teratoma the prognosis is excellent.[11]

Liang Zhou et al in a Review of Literature of Primary adrenal teratoma found only 18 adults and 7 paediatric cases.[13] Among Paediatric age group Boys are more common than Girls. Only few cases are reported as Girl child in literature where as our series has two female cases.

That study also narrates that symptoms are not typical. Sometimes incidental finding and no left or right side predominance were observed. Lastly 5 children were followed up without recurrence or death. Observations of our study found to be close to that literature review.

Table 1: Summary of cases.

Age	Sex	Clinical features	Diagnosis	Surgery	Histopathology and immuno-histochemistry	Postoperative follow up
Adrenal tumors						
7y 6 m	F	Pain abdomen + Lump in Right flank	Right Adrenal Tumor	Right Adrenalectomy	Adrenal Teratoma	U/E
4y	F	Facial puffiness + Hypertension	Functional adrenal Tumor (Rt)	-do -	Pheo-chromocytoma with Atypical IHC (Melan A, CD 56, Ki 67 +ve & chromogranin A, Synaptophysin, S100, Inhibin Alfa -ve)	Uneventful, Facial puffiness reduced, Reduced dose and number of anti-hypertensive medicines
6y	M	Precocious puberty in boy	Virilizing Adrenal Tumor (Rt)	-do-	Adreno cortical Adenoma	U/E
Tetatomas						
6m	M	Lump abdomen	Retro-peritoneal mass	Excision of Retro-peritoneal mass	Mature Teratoma (Fetus-in-Fetu)	U/E
4m	F	Lump abdomen	Retro-peritoneal mass	-do-	-do-	U/E
7m	F	lump in Left flank + Hypertention	Left Suprarenal mass with non functioning Left Kidney	Excision of mass from left suprarenal area	Mature Teratoma	U/E, Reduced Number and dose of anti-hypertensive medicines

(*U/E – uneventful)

Functional Adrenal tumour is a rare incidence in paediatric population and its management requires specialised multimodality management. Its incidence is <0.2% of all Paediatric neoplasms.[14]

presence of pubic hair and increased penile size, advanced bone maturation, markedly elevated DHEA and 17 Hydroxyprogesterone levels and imaging showing the presence of a right suprarenal mass suggestive of adrenal tumour as the cause of precocious puberty.

PP is a rare entity and its incidence is 1:5000 to 1:10000. It is defined by expression of Secondary sexual characters in males before 9 years and in females before 8 years age. It occurs due to excessive production of sex steroids - Activation of Hypothalamic- pituitary- gonadal axis (Central) and Nonhypothalamic mediated increase in sex steroid(peripheral). Central precocious puberty or GnRH dependent PP is commoner than GnRH independent PP. More in girls than in boys incidence being 0.3 to 0.5 per million child per year. Manoj et al revealed bimodal age of distribution of adrenal tumors.[14] 1st peak before 5 years and 2nd peak 4th and 5th decade. Virilizing adrenocortical adenoma in girl child was reported by Maji et al and Shenge et al. 3 more such cases was reported earlier.[15]

Rarity of this tumor in boys- 10 month infant is revealed in Razavi et al.[16] In our case it was a 6 years male child with sudden progression of the disease.

Cases presenting with virilizing signs must alert for a probable diagnosis of adrenal tumor (Maji et al). In our case tumor size was 5.3 cms. Monika et al found an usually large functional adrenal adenoma measuring 12 cms. Hence, rare tumor larger than 5 cms could be adenoma.[17]

Adrenalectomy is the treatment of choice in GnRH independent precocious puberty. Postoperative there is decrease in the hormone levels and improvement in secondary sexual characters. Medical treatment is the first choice suggested in GnRH dependent precocious puberty (hypothalamic cause).[18,19]

Manoj et al in his retropspective study on 42 cases of Adreno cortical adenoma and adreno cortical carcinoma found Weiss scoring system to be useful in distinguishing the tumors but this was possible only after histopathology.[14] After adrenalectomy based on Weiss criteria HPE examinations revealed a adrenocortical adenoma.

Our next Case of the series is a 4 years girl with clinical feature of Cushing's syndrome and Pheochromocytoma with Adrenal SOL (Right Side). No extra Adrenal source of ACTH is identified. Adrenal Pheochromocytoma in Children below 6 years presentation is rare (<1: 500000).[20] When an adrenal tumor presents with malignant hypertension and features of Cushing's Syndrome without any Pituitary or extra adrenal source of ACTH it is challenging from both diagnostic and management point. Adrenal Pheochromocytoma is a medullary tumor and Cushing's syndrome secreting hormone comes from Adrenal cortex but combination of both is not so common.

Hypercortisolism in patients with tumors of the autonomic nervous system has been reported previously. Luton et al. reviewed cases with pheochromocytoma and hypercortisolism due to adrenal tumors.[21]

Besides pheochromocytomas, extra-adrenal paragangliomas have been associated with Cushing's syndrome. Immunologic assays of tumor tissues have identified the usual cause as ectopic ACTH production.[20,22]

In Immunohistochemistry staining Chromogranin A, Synaptophysin, S100, Inhibin Alfa was negative which is typical for Pheochromocytoma. Melan A-(very focal faint positive),CD 56- (patchy positive), Ki 67- (less than 2%) positivity showed rarity of the disease pattern.

FIF is a rare anomaly of embryogenesis in which a malformed parasitic twin is found within the body of the normally developed host.[23] The majority of cases appear in infancy with an incidence reported at 1 in 500,000 births.[24] In most cases, the parasitic twin is anencephalic and usually contains a vertebral column and budding limbs. The upper limbs are less developed than lower limbs, and are usually located in the abdomen of the autosite.[25] The monozygotic diamniotic twin is quite similar to a mature teratoma; however, a key difference from fetus in fetu is a mature teratoma has an independent growth ability and malignant potential.[26] Diagnosis is often made preoperatively with ultrasonography, plain radiography, computed tomography (CT) or magnetic resonance imaging (MRI). Histopathologic findings confirm the diagnosis, and the recommended treatment is complete excision. Our two cases both presented in infantile age group. In a literature review of 95 FIF cases >60% was found in male, >70% was retroperitoneal and >75% had vertebral column.[27]

CONCLUSION

Abdominal mass is a common finding in children, either palpable or radiologically evident. Asymptomatic mass in abdomen sometimes present with a rare tumor that require surgery whereas functional/hormonally active tumors are rarely palpable but present with varied arrays of symptoms other than that caused by a lump and tumor removal is important from their treatment point of view. These atypical presentations may pose a challenge in diagnosis and management. Often these are first time occurence for the treating team and therefore, require careful judgement, gathering as much information possible and execute them efficiently with help of shared experience in literature and help of multi-disciplinary teams. Our experience although limited still has provided us with enriching learning experience, which authors hope will help us in managing such cases or still rarer variations successfully and there lies the idea of sharing them. These children need to be followed up to look out for long term outcome for such diseases.

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