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Congenital Infantile Fibrosarcoma of The Duodenum, A Case Report

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Abstract

Background: Congenital infantile fibrosarcoma is a rare soft-tissue tumor in the pediatric age group and seldom involves the gastrointestinal tract.

Methods: This study is a case report study done in the Raparin Pediatric Hospital in Erbil on 9 days old boy with signs of intestinal obstruction. After explorative laparotomy a solid mass involving duodenojejunal junction was found, resection of the mass was done with end-to-end anastomosis and approval was taken from patient's parents.

Results: Histopathologic examination showed extensive replacement and effacement of the small bowel architecture by a cellular neoplasm. The tumor was extended through muscularis propria to the serosa with histological features consistent with congenital infantile fibrosarcoma, the diagnosis was confirmed by immunohistochemistry and genetic analysis which demonstrated the t(12;15) translocation. The baby was discharged on the 5th hospital day and followed up for 2 years without recurrence.

Conclusion: Here we reported a rare case of congenital infantile fibrosarcoma arising from the small bowel in a new born baby, operated on, totally resected and histopathology proved the diagnosis and the patient was disease free after 2 years follow up.

Keywords: Children, Congenital**,** Fibrosarcoma

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Case report

A nine-day old male was reported with a duodenojejunal junction tumor. He belonged to a twin delivered by caesarean section. He presented with bilious vomiting & upper abdominal distension since birth, passed meconium normally, no other significant signs and symptoms were found. No associated congenital abnormalities were found. Plain abdominal X-Ray showed multiple air fluid levels with dilated upper abdomen Figure (1). Abdominal Ultrasound exam showed a mass in the duodenojejunal junction of 3×3cm, related to the duodenojejunal junction.Explorative laparotomy was done, A mass in the DJJ was found of about 3x3cm involving the whole bowel diameter Figure (2). The mass was firm, fleshy, yellowish pale in color. Resection done with end-end anastomosis of the jejunum. The mass sent for histopathological examination.The postoperative period was uneventful the patient started oral feeding after 3 days and discharged home after 5 days uneventfully. Histopathologic evaluation demonstrated a 3x3x2cm mass with a fleshy grey cut section. Microscopically, sections showed extensive replacement and effacement of the small bowel architecture by a cellular neoplasm. The tumor was extended through muscularis propria to the serosa and it was composed of a predominantly spindle cell population running in fascicles producing a well-developed herringbone appearance Figure (3). There was moderate cellular pleomorphism, mitotic count was > 5/ 10 HPF and Ki-67 labelling index was around 10%.Immunohistochemical stains were negative for MyoD1, C- Kit(CD 117), S100 protein, CD34, actin and desmin but was positive for Vimentin Figure (4).The diagnosis of congenital infantile fibrosarcoma (CIF) was confirmed by detection of t(12;15) ETV6-NTRK3 fusion transcript on reverse transcriptase polymerase chain reaction (RT-PCR) done on formalin fixed paraffin embedded sections of the specimen(the molecular diagnosis was thankfully done in Our Lady's Children Hospital in Dublin, Ireland).No further treatment used. The patient followed for two years post operatively with no untoward sequelae.

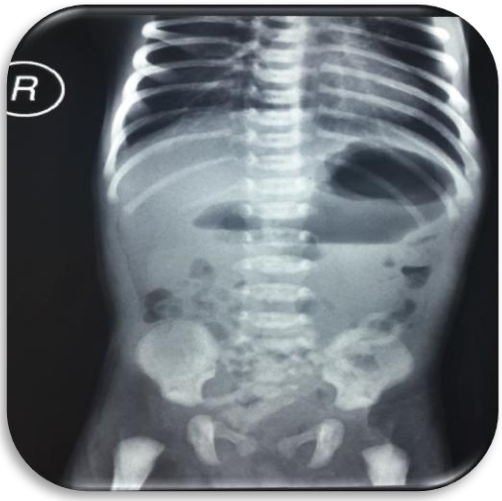

Figure (1): Plain abdominal X ray showed Multiple air fluid levels.

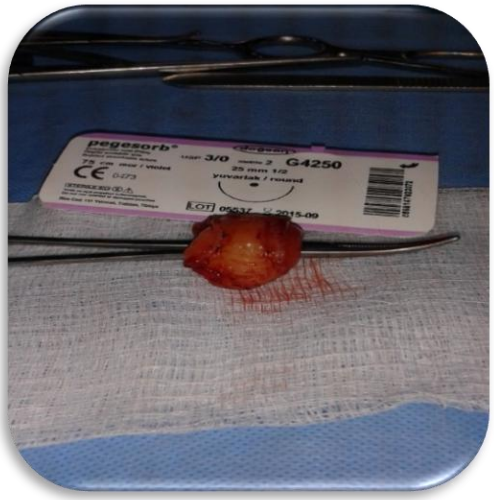

Figure (2): The resected mass in DJJ.

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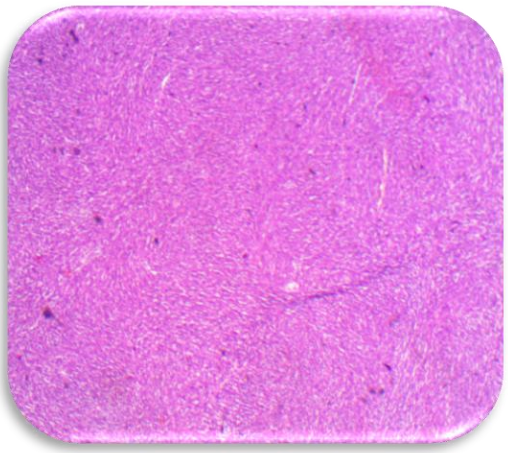

Figure (3). Compact spindle cells arranged in fascicular pattern (Hematoxyline & eosin x200).

Figure (4). Positive immunohistochemical staining for Vimentin (x200).

Discussion

Congenital infantile fibrosarcoma is a rare malignancy affecting infants and young children and is characterized by a rapidly growing invasive mass. Although most commonly presents in the extremities, it has been known to arise in virtually all other areas of the body.[1] Intra-abdominal CIF is particularly rare and most cases tend to arise from soft tissue structures rather than bowel or solid organs. There have been few case reports of CIF involving either the small intestine or colon, and these were all identified due to either intrinsic bowel obstruction or perforation Table (1).[2-12] Furthermore, these patients were nearly all identified at parturition or within the first several days of life

Table (1): Main previously reported cases of Intestinal congenital fibrosarcoma

Study	Age	Gender	Presentation	Location	Treatment/outcome
Wright,1971 (2)	16 months	M	Vomiting without obstruction	Duodenum	Pancreaticoduodenectomy, well at 24 months
Shearburn et al., 1975 (3)	1 day	F	Obstruction	Duodenum	Pancreaticoduodenectomy, well at 14 months
Shermanband Neustein,1985 (4)	1 day	M	Obstruction	Ileum	Resection, well at 3 years
Shima et al., 2003 (5)	1 day	F	Perforation, meconium peritonitis	Jejunum	Resection, full recovery
Buccoliero et al., 2008 (6)	3 days	M	Perforation	Ascending colon	Ileocolectomy, well at 12 months

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Islam et al., 2008 (7)	0 days	M	Meconium peritonitis	Descending colon	Resection, well at 5 years
van Niekerk et al., 2010 (8)	2 days	M	Obstruction	Ileum	Ileocecectomy, well at 9 months
Rizkalla et al., 2011 (9)	5 days	M	Perforation	Ileum	Resection, well at 12 months
Kim et al.,2013 (10)	2 days	M	Pneumo-peritoneum	Sigmoid colon	Sigmoid colectomy, well at 1 year
Sulkowski et al, 2014 patient 1 (11)	4 months	F	Asymptomatic abdominal mass	Jejunum	Resection, full recovery
Sulkowski et al, 2014 patient 2 (11)	15 days	F	Obstruction	Ileum	Ileocecectomy, full recovery
Kaiser et al, 2017 (12)	New born	F	Perforation	Jejunum	Resection with jujenostomy, full recovery
Current study 2021	9 days	M	Obstruction	Duodenum	Resection, full recovery

The differential diagnosis for an infant with a solid abdominal mass includes neuroblastoma, Wilms tumor, hepatoblastoma, rhabdomyosarcoma, and lymphoma. With respect to tumors arising from the small intestine, in infants these are most typically leiomyomas, leiomyosarcomas, or other tumors of mesenchymal origin, all of which are extremely uncommon.[13] Congenital infantile fibrosarcoma is distinct from fibrosarcoma in adults, in that metastases are rare, local recurrence is common and the prognosis is good. This low-grade malignant lesion is slightly more common in boys than in girls and has a 90% survival at 5 years.[14] CIF expresses tyrosine kinase receptor 3, NTRK3, and the fusion protein product, ETV6-NTRK3, associated with a chromosome 12 to 15 translocation. This chromosomal translocation and fusion protein are characteristic of CIF. [15] Which confirmed the diagnosis in our case.

Conclusion

Here we reported a rare case of congenital infantile fibrosarcoma arising from the small bowel in a new born baby, operated on, totally resected and histopathology proved the diagnosis and the patient was disease free after 2 years follow up.

Declaration of conflict of interest:

There are no conflicts of interest among the writers.

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