age of presentation is 70 years at a median of 72 months after RT.[3]

In the pre-BCT era, secondary cutaneous angiosarcoma of the breast and upper arm was described in association with lymphedema following mastectomy and axillary dissection (Stewart–Treves syndrome). Now, most secondary angiosarcoma is cutaneous radiation-associated angiosarcoma of the breast (RACA).[5]

Another rare and poorly understood cutaneous vascular lesion has been described in the context of RT.

Atypical post-radiation vascular proliferation (APRVP) was thought historically to be a benign, distinct entity but shares enough clinical and histologic features with angiosarcoma to raise concern that APRVP and angiosarcoma may represent ends of a spectrum of radiation-associated vascular lesions.[6] Several reports confirmed that some patients initially diagnosed with APRVP following BCT were subsequently found to have RACA at the same site.[7-9] The natural history of APRVP is as yet undefined.

Some authors proposed that the clinical, morphologic, and immunophenotypic overlap of APRVP and RACA suggests that they exist as a continuum along a morphologic spectrum.[10] In this case, MYC amplification may be a requisite genetic abnormality in transformation of APRVP to RACA.

Although most patients with APRVP do not develop RACA,[7, 9] small biopsies showing APRVP require careful interpretation. A diagnosis of APRVP in violaceous or erythematous skin in an irradiated breast should prompt further investigation to rule out RACA.

Several single institution studies and one meta-analysis have reported relatively poor prognosis for women with RACA.[2, 11-13]

Radiological aid, as seen in our case report, in diagnosing RACA is very limited and unspecific.

A thickening of the skin at mammography should always be compared with the baseline examination six month after radiation therapy, when radiation-induced skin thickening is mostly pronounced.

Any further thickening of the skin should therefore raise the suspicion of a secondary lesion.

Some surgical dedicated institutions suggest that a standard mastectomy may be sufficient for primary mammary angiosarcoma but is inadequate for RACA: the authors specifically remove all irradiated breast skin considered at-risk and that may contain occult sites of disease.[14]

Although they admit this do not universally prevent recurrence, they found that patients who did not undergo resection of all irradiated breast skin trended toward a worse median Local Recurrence Free Survival (10.0 *vs*. 80.8 months) and Overall Survival (29 months *vs*. not achieved). They also resect the pectoralis major when concerned about tumor extension.

Locoregional recurrences are very common, occurring at a median 5 months after initial diagnosis. RACA is histologically multifocal in a substantial number of patients.

The importance of locoregional control and careful clinical evaluation to detect presence of recurrences in short order should be emphasized. Distant metastases are associated with a dismal prognosis, with a mortality rate of 89 % in Morgan's cohort. In contrast, 30% of women with locoregional recurrences died of angiosarcoma.[14]

Although RACA remains an aggressive disease with a poor prognosis, we do not believe that it should dissuade breast cancer patients and oncologists from using RT as part of BCT. The absolute incidence of RACA is low: some authors previously reported an incidence of 6.6 per 100,000 person-years and others have reported incidence rates of <1% among patients receiving RT as part of BCT.[4]

The Early Breast Cancer Trialists' Collaborative Group reported that RT significantly reduced the risk of locoregional recurrence and 15-year mortality among breast cancer patients receiving BCT.[15]

However, women treated with RT for breast cancer should be evaluated regularly and educated by their oncologists regarding the risks of this secondary malignancy.

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Ileal duplication cyst in new born complicated with volvolus: 2 cases report and literature review

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Duplication of alimentary tract are uncommon congenital lesions that can occur anywhere from the mouth to the anus and have reported incidence of 1:25000 deliveries. The ileum appears to be the most common location and the majority duplications are cystic lesion. Symptoms vary with the location, the type and the complication like volvulus formation. Midgut volvulus is one of the most life-threatening abdominal emergencies in the neonatal period. It is necessary to be aware of this condition despite his rarity. We present two cases of acute abdomen in new born caused by volvolus secondary to ileal duplication cyst with a review of the literature. Patient 1: Male neonate, presenting at the birth cyanosis and progressive abdominal distension. The abdominal ultrasound showed free fluid within the peritoneal cavity and a only one dilated loop of bowel. On surgical exploration, large volume hemoperitoneum and a volvulus secondary to a mid ileum cystic duplication was found along the mesentery with gangrene of the involved segment. Local resection along with excision of 15 cm of involved bowel and end to end ileoileal anastomosis was done. The histopathology confirmed the diagnosis. Presently, the child is 8 years old and doing well. Patient 2: 48-hour-old infant female neonate present to us for abdominal distension and bilium vomiting. The abdominal radiographs revealed features of small intestinal obstruction. On surgical exploration, volvulus secondary to a terminal ileum cystic duplication was found along the mesentery with gangrene of the involved segment. Local resection along with excision of 16 cm of involved bowel and end to end ileoileal anastomosis was done. The histopathology confirmed the diagnosis. Presently, the child is 7 years old and doing well. The natural history of intraabdominal enteric duplication is quite variable ranging from causing complication that require surgery in the first day of life to being indefinitely asymptomatic. Volvolus is rare complication of enteric duplication and occurred near 23% of patient affected. We perfomed a review of the literature from 1957 to 2012 and found no more than 26 cases of volvulus secondary to jejunal-ileal duplication. All the cases (except 4 that occurred in patient older than 1 month) resulted in new born with the symptoms of acute abdomen.

KEY WORDS: Intestinal volvulus - Abdomen, acute - Infant.

Gastrointestinal duplications (GID) are uncommon congenital lesions that can occur anywhere from the mouth to the anus and have a reported incidence of 1:25000 deliveres. Ileum is the most common site of occurence.[1]

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The natural history of intra-abdominal enteric duplications is quite variable, ranging from causing complications that require surgery in the first day of life to being indefinitely asymptomatic.[2] The clinical presentation can range from an asymptomatic abdominal mass to a clinical bowel obstruction secondary to a complication like volvolus formation.[3]

Volvolus as result of duplication occurs rarely and it almost exclusively develops in the neonatal period.

The ideas for this article was born from the clinical history of two neonates referred to our clinic for a rare cause of acute abdominal. They showed an acute abdomen caused by volvolus secondary to ileal duplication cyst. We perform a review about the cases of volvulus secondary to a GID.

Clinical series

Case 1.—Male, full term neonate, his weight at the birth was 2480 grams. The antenatal ultrasound showed a dilatation involving the descending colon. He presented at the birth a progressive abdominal distension and cyanosis that required intubation. On examination his abdomen was distended and tender. Nasogastric suction yielded 15 mL of gastric secretions. On the fourth hours of life he passed meconium with the aspect of melena. The abdominal X-ray revealed no intraluminal gas and the abdominal ultrasound showed free fluid within the peritoneal cavity and a only one dilated loop of bowel. In view of this finding the 10-hours-old child was underwent to a laparotomy. A right supraumbilical transverse incision was performed. A large volume hemoperitoneum and a volvulus secondary to a mid ileum cystic duplication was found along the mesentery. The loop of the ileum had undergone torsion of four turns with resultant gangrene of the involved segment (Figure 1). Local resection along with excision of 15 cm of involved bowel and end to end ileoileal anastomosis was done. The histopathology confirmed the diagnosis. Postoperative course was regular and presently the child is 8 years old and doing well.

Case 2.—Female, full term neonate, her weight at the birth was 3,519 grams and the one and five minutes APGAR score was 10. There was a suspect of right ovarian cysts in the prenatal ultrasound.

Figure 1.—The loop of the ileum secondary to the cyst.

She passed meconium in the first 36 hours. On the second days of life developed abdominal distension and bilious emesis. On examination his abdomen was distended and tender. Nasogastric suction yielded 20 ml of bilious secretions. The abdominal radiographs revealed features of small intestinal obstruction. A contrast enema was performed and it showed an obstruction of terminal ileum. She was underwent to a surgical exploration and a right supraumbilical transverse incision was performed. On further delivery of the bowel outside the abdominal cavity, volvulus secondary to a terminal ileum cystic duplication of 4x5 was found along the mesentery with gangrene of the involved segment (Figure 2). Local resection along with excision of 16 cm of involved bowel and end to end ileoileal anastomosis was done. The histopathology confirmed the diagnosis and did not reveal any evidence of gastric mucosa. Postoperative course was regular and presently, the child is 7 years old and doing well.

Discussion

GID are uncommon malformation of the intestinal tract that can occur anywhere from the mounth to the anus. Ileum is the most common site of occurance.[1]

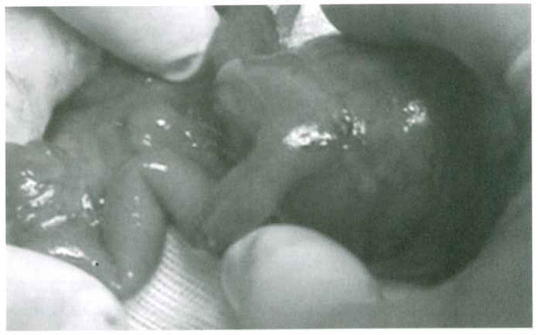

Figure 2.—Ileum cystic duplication.

The characterization of enteric duplication is not simple given that there is a large range of anatomical variability.[4]

The majority are cystic but they can be cystic or tubular, varabily for size and may communicate with intestinal tract.

GID tend to be paramesenteric and share a common muscular wall and blood supply with native intestine.[5]

Duplication arise from disturbances in the embryonic development of gastrointestinal tract.

Several theories have been proposed for the formation of the duplication including the aberrant luminal recanalizion, the diverticular theory and the split nothocord theory but no single theory adequately explains all the know duplication.[6]

The presentation of GID can vary according to the size, type, location and the presence of gastric mucosa in the duplication.[5] Some GID may be totally asymptomatic, identified on routine physical examination or during investigations for other problems.[3]

Over than 60% of the GIDs are symptomatic in the first year of life.[7, 8]

Abdominal pain, vomiting, and an abdominal mass are the most common symptoms and signs attributable to GIDs located in the abdomen.[9, 10]

TABLE I.—*Twenty-six patients with volvolus secondary to GID.*

Author	Year	Cases	Sex	Age of diagnosis	Localization	Exitus
Kirkmann	1957	1	M	30 days	Torsion isolated jejunal duplication cyst	No
Howanietz	1967	1	F	3 days	Ileal duplication cyst	Yes
Bhargava JS	1976	1	M	5 years	jejunal duplication cyst	Yes
Richard J Bower	1977	1	M	13 years	Ileal duplication cyst	Yes
George W. Holcomb III	1988	4	?	Newborns	Ileal duplication cyst	Yes
A. Ciardini	1992	3	?	Newborns	Ileal duplication cyst	?
M.D. Stringer	1995	1	?	Newborn	Ileal duplication cyst	Yes
P.T. Foley	2003	1	M	2 days	Torsion isolated ileal duplication cyst	Yes
P. S. Puligandla	2003	5	?	Newborns	Ileal duplication cyst	1 yes; 4 ?
Abdur-Rahaman Oljaide	2010	2	F	10 days	Ileal duplication cyst	No
			F	2 days		
Okoro PE	2010	1	M	10 month	Ileal duplication cyst	?
Pablo Laje	2010	1	M	1 day (prenatal)	Ileal duplication cyst	No
Minakshi Sham	2011	1	M	4 days	Ileal duplication cyst	No
N. Pant	2012	1	M	18 months	Torsion isolated ileal duplication cyst	No
Messina	2012	2	M	Prenatal	Ileal duplication cyst	No
			F	4 days		

The potential complications of an intra-abdominal GIDs are many and can be fatal.[4]

The complications can occur because GID can serve as the focal point for volvolus or intususception or because gastric mucosa can lead to peptic ulcerations, bleeding, perforation with peritonitis.[5]

From 1957 to 2012 resulted no more than 26 cases in the literature including our patients (Table I). We performed a review of the literature searching all the case of volvulus secondary to GID.

Volvolus, caused by a duplication, was observed rarely and based on a study of 76 patients, Puligandla *et al.* found that the volvolus occurred in 23,8% of patients affected by GID.

Ileum was the most frequent site of ED, jejunal duplications were found in two patients and in six patients the localization remain unknown.

The type of ED, when it was descripted, was cystic. In the 26 cases found, we also considered two cases of acute abdomen due to torsion of the pedicle of a completely isolated duplication cyst.[11, 12]

Twenty-two patients were symptomatic during the newborn and in 92,3% of patients the volvulus occurred earlier until the first two years. In the second case that we presented the volvulus appeared when she was 2-days-old. We suspect that in the first case the volvulus become symptomatic in the first day of life but it appeared in the prenatal period because the patient presented occlusion signs at the birth. In the literature we found only one other patients in addition to our case with volvulus probably occurred during fetal life.[2] Pablo Laje *et al.* suspected that the volvulus, in the patient of his series, occurred most likely during fetal life because the proximal and distal ends of the volvulized segment were near-atretic and the segment was not viable.[2]

The remaining 2 patients become symptomatic respectively at 5 (Bhargava *et al.*) and 13 (Richard J Bower *et al.*) years old.[10, 13]

Abdominal swelling and bilious vomiting were the most common presentation.

Melena was observed only in one patient of our series.

The diagnosis is rarely made until at surgery because of nonspecificity of symptoms and presentations.[14] The prenatal diagnosis it is possible (GID are believed to occur between the fourth and eighth weeks of embryonal development), but the differential diagnosis to be considered includes neuroenteric cyst, anterior myelomeningocele, mesenteric cyst, choledochal cyst, and ovarian cyst.[2]

Both of our patient had a delayed prenatal diagnosis. In the first case the antenatal ultrasound showed a dilatation involving the descending colon while in the second case there was an ultrasound image compatible right ovarian cyst.

Treatment of EDs is by surgical means.[3]

Most of the reported cases showed a good survival, 4 patients died for volvolus or postoperative complications and in the 4 cases reported by Ciardini A *et al.* and Okoro PE *et al.* the prognosis was unknown.[15, 16]

Conclusions

This is the first review about the case of GID complicated by volvolus. GID are uncommon malformations and volvulus secondary to GID is a very rare cause of acute abdomen in neonate. Despite his rarity it is necessary keep it in mind in the differential diagnosis of acute abodomen in new born primarily if there is a prenatal ultrasound suspect of abdominal mass.

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