Hernia (2014) 18:439–442
DOI 10.1007/s10029-014-1216-x

CASE REPORT

Rare case of obturator hernia in a patient with Marfan's syndrome

R. Peter · V. Indiran · K. Kannan · P. Maduraimuthu · C. Varadarajan

Received: 26 June 2013 / Accepted: 5 January 2014 / Published online: 12 January 2014

Abstract Obturator hernia is a very rare type of abdominal hernia which constitutes <1 % of all the hernias. It is an important cause of small bowel obstruction which is associated with a high mortality rate if left untreated. Obturator hernia typically occurs in an elderly women or patients with chronically raised intraabdominal pressure or previous multiple pregnancies. We report a case of obstructed obturator hernia in a young female patient with Marfan's syndrome complicated with dissecting aortic aneurysm and chronic kidney disease. Though recurrent and incisional hernia constitutes one of the minor diagnostic criteria of Marfan's syndrome, obturator hernia being a very rare entity, has been reported very rarely in a patient with Marfan's syndrome.

Keywords Marfan's syndrome · Obturator hernia · Dissecting aortic aneurysm · Small bowel obstruction

Introduction

An obturator hernia is a rare type of abdominal hernia with protrusion of abdominal contents through the obturator foramen in the pelvis. It occurs in an elderly multiparous women or patients with increased intraabdominal pressure causing increased mortality and morbidity. It commonly manifests as small bowel obstruction, obturator neuralgia or rarely a palpable mass. Marfan's syndrome is an inherited connective tissue disorder with recurrent or incisional hernia accounting for one of the minor diagnostic criteria. Obturator hernia is reported very rarely in a patient with Marfan's syndrome. Obturator hernia usually presents with nonspecific signs and symptoms making preoperative diagnosis difficult clinically. An abdominopelvic computed tomography has 100 % sensitivity for the diagnosis and helps in avoiding complications arising out of delayed diagnosis.

R. Peter · V. Indiran (✉) · K. Kannan · P. Maduraimuthu · C. Varadarajan
Department of Radio Diagnosis, Sree Balaji Medical College and Hospital, No 7 Works Road, Chrompet, Chennai 600 044, Tamil Nadu, India
e-mail: ivraman31@gmail.com

Case report

A 27-year-old female was referred to Department of Radiodiagnosis for abdominopelvic computed tomography (CT) with complaints of severe pain in the left groin for 1 day. The pain was continuous and radiating to the medial aspect of the left thigh. There were no other associated symptoms. The patient had a history of congenital heart disease with previous echocardiography revealing severe mitral regurgitation, tricuspid regurgitation and pulmonary hypertension. The patient also had hypertension and chronic kidney disease stage V. The patient had previously been operated for bilateral inguinal hernia.

On general examination, the patient had Marfanoid features like tall stature, arm span to height ratio more than 1.05, positive Steinberg's sign or thumb sign in which the thumb when opposed across the palm extends beyond the ulnar border of the palm, kyphoscoliosis, arachnodactyly (long slender fingers and toes) and positive wrist sign in which the patient can enclose the wrist with the thumb and little finger of the other hand and digits over each other. On local examination, a 3 × 2 cm tender, non-compressible swelling was noted in the medial aspect of the left thigh. Patient's laboratory reports apart from abnormal renal

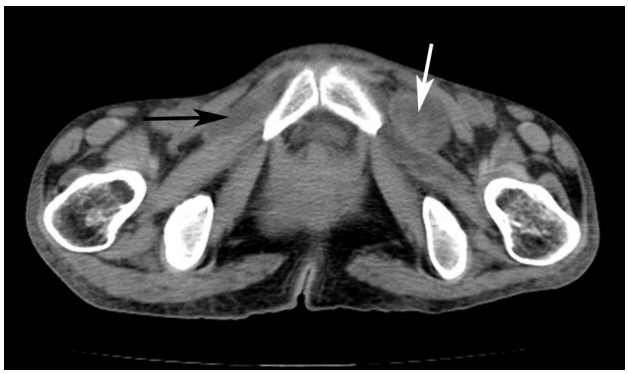

Fig. 1 Non-contrast CT axial image shows small bowel lying between the left obturator externus and pectineus muscles (*white arrow*). Fluid noted between the right obturator externus and pectineus muscle (*black arrow*)

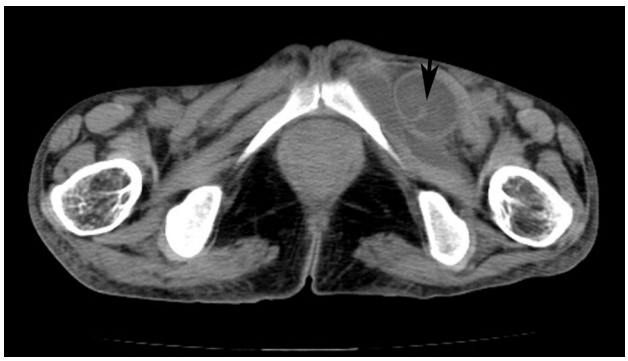

Fig. 2 Non-contrast CT axial image shows dilated ileal loop and surrounding fluid between the left obturator externus and pectineus muscles (*black arrow*)

parameters (Blood urea—156 mg/dl and serum creatinine—5.5 mg/dl) were unremarkable.

Non-contrast abdominopelvic CT with few sections of the upper thigh was done. Intravenous contrast was not given as the renal parameters were deranged. Scanogram showed scoliosis of dorsolumbar spine with convexity to the right side and dilated small bowel loops in the left lower quadrant. CT sections revealed findings consistent with obstructed obturator hernia—a dilated ileal loop and surrounding fluid in the left upper thigh, lying between obturator externus and pectineus muscles. The dilated ileal loop within the sac showed a maximum dimension of 2.9 cm (Figs. 1, 2). The ileal loop proximal to the hernial orifice appeared dilated for a length of about 20 cm with a maximum dimension of about 3 cm (small bowel obstruction) (Fig. 3). Rest of the small and large bowel loops appear collapsed. Small fluid collection was also noted between the right obturator externus and pectineus muscles (Fig. 1). CT revealed a large lower thoraco-abdominal aorta dissecting aneurysm with eccentric

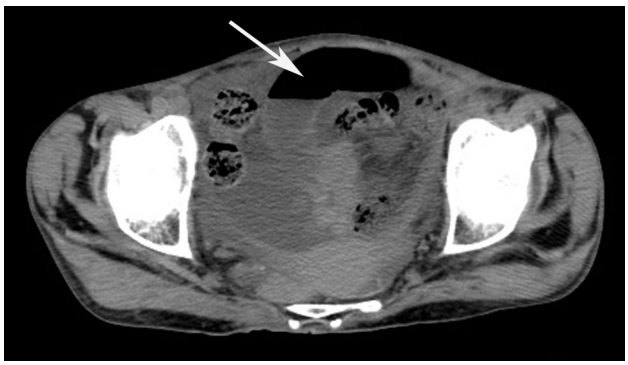

Fig. 3 Non-contrast CT axial image shows dilated small bowel loop with fluid level (*white arrow*) in the left lower quadrant and pelvis

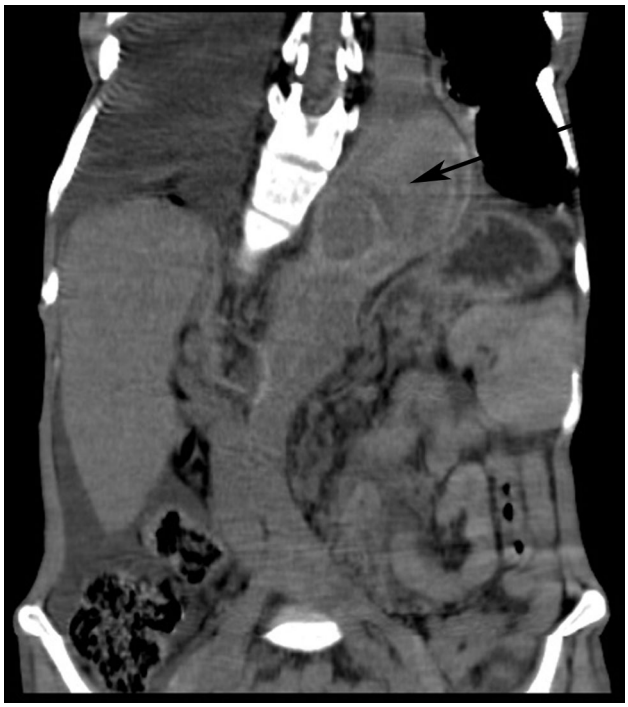

Fig. 4 Non-contrast CT curved coronal reformat image shows lower thoraco-abdominal aorta dissecting aneurysm with eccentric luminal thrombus (*black arrow*). Scoliosis of the dorsolumbar spine is also seen

luminal thrombus extending from D9 to L3 level with maximum cross-sectional dimension of 6.2 cm at D10–D11 level (Fig. 4). Rest of the aorta was normal in dimension. Scoliosis of the dorsolumbar spine was seen with convexity to the right side and apex at L1 level. Moderate right-sided pleural effusion and ascites were noted.

The patient underwent exploratory laparotomy with an infraumbilical midline incision. A circumference of ileum approximately 50 cm away from the ileocaecal junction was found entrapped in the left obturator foramen. The bowel was congested and viable. The contents were

reduced intraabdominally and preperitoneally and a prolene mesh (15 × 7.5 cm) was placed to cover the left obturator foramen and anchored to the pectineal ligament. The patient's symptoms improved following the surgical correction of the obturator hernia.

Discussion

Marfan's syndrome is an inherited connective tissue disorder that has an autosomal dominant pattern. The syndrome is caused by isolated fibrillin 1 (FBN1) gene mutation on chromosome 15, which codes for the tissue protein fibrillin. Both male and female sexes are found to be affected equally [1]. Clinical presentation is predominantly due to the abnormalities of the musculoskeletal, cardiovascular, ocular, central nervous and integumentary system involvement [2]. The diagnostic criteria for the diagnosis of Marfan's syndrome are revised according to GHENT classification and it is illustrated in the Table 1.

Though recurrent and incisional hernia account for one of the minor diagnostic criteria for Marfan's syndrome, the obturator hernia has been reported very rarely in a patient with Marfan's syndrome. Our literature search yielded only one similar case report so far [4].

Obturator hernia is a very uncommon type of hernia, with an incidence of about 0.073–1 % of all the hernias. This type of hernia was first reported by Ronsil in 1724 [5, 6]. It is the protrusion of the hernia sac through the obturator canal and foramen. Obturator hernia predominantly affects females. The females are affected six to nine times more than men [6, 7]. The female predominance is due to the larger and oblique inclination of the obturator canal in the female pelvis. The right side is commonly affected than the left side because the sigmoid colon overlying the obturator foramen on the left prevents it [7, 8].

The risk factors of obturator hernia include elderly females and patients with increased intraabdominal pressure. It is also seen in patients with rapid weight loss with

Table 1 Criteria of Marfan's syndrome [3]

System affected	Diagnostic criteria	Major criteria	Minor criteria
Musculoskeletal system	2 major (or) 1 major and 2 minor	Pectus carinatum	Pectus exacavatum of moderate severity
		Pectus exacavatum	Joint hyper mobility
		Upper to lower segment ratio <0.9	High arched palate
		Arm span to height ratio >1.05	Facial appearance—enopthalmos, retrognathia, down slanting palpebral fissures
		Positive wrist sign	
		Positive Steinberg's sign(thumb sign)	
		Thoracolumbar scoliosis of more than 20°	
		Pes planovalgus deformity	
		Acetabuli protrusion	
Ocular system	Positive major (or) 2 minor	Lens dislocation (ectopia lentis)	Flat cornea
			Increased axial length of the globe
			Hypoplastic iris or Hypoplastic ciliary muscle
Cardiovascular system	1 major (or) 1 minor	Dilatation of the ascending aorta with or without regurgitation	Mitral valve prolapse with or without regurgitation
		Dissecting aortic aneurysm	Dilatation of the main pulmonary artery in the absence of valvular or peripheral pulmonic stenosis
			Calcification of mitral valve annulus
			Dilatation or dissection of the thoracic or abdominal aorta
Pulmonary system	1 minor	–	Spontaneous pneumothorax
			Apical blebs
Skin and integumentary system	1 minor	–	Striae atrophicae
			Recurrent or incisional hernia
Central nervous system	1 major	Lumbosacral dural ectasia	–
Genetics	1 major	Family history	–
		Genetic mutations	
		Inheritance of DNA marker haplotype.	

decreased fatty tissue surrounding the obturator foramen [9].

Anatomy of the obturator foramen and canal

The obturator foramen is a large oval or a triangular aperture which is located in the anterolateral aspect of the pelvis. It is bounded by the pubis, ischium and their rami. The foramen is covered by a thin obturator membrane except for a small passage through which the obturator nerve and the vessels enter the medial aspect of the thigh [10]. The obturator canal is a 2–3 cm long canal which exits through the obturator foramen. The canal is bounded superiorly and laterally by the pubic bone and inferiorly by the obturator muscles [11].

Clinical features

The various clinical presentations of the patients include small bowel obstruction, obturator neuralgia, positive Howship Romberg's sign, positive Hannington Kiff sign and rarely a palpable mass. Small bowel obstruction is due to the involvement of either jejunum or ileum within the hernia sac. Obturator neuralgia presents as either hypoesthesia or hyperesthesia extending from the inguinal crease to the anteromedial aspect of the thigh [11]. Howship Romberg's sign is the pain radiating down the medial aspect of the thigh due to the compression of the obturator nerve. This sign is only reported in 15–50 % of the cases [6, 8]. Hannington Kiff sign is the absence of obturator reflex in the thigh due to the compression of the obturator nerve [11]. This sign is considered pathognomonic for diagnosing obturator hernia [10]. A very few percent of the patients may present with a palpable mass on the medial aspect of the thigh, in the adductor region, which is palpated on per vaginal examination [6, 10, 11]. Rarely the patient may present with ecchymosis in the medial aspect of the thigh due to the collection from the hernia sac.

In our case, patient had positive Howship Romberg's sign, features of small bowel obstruction and a palpable mass.

The diagnostic imaging modalities of the obturator hernia include plain radiograph, ultrasonography, and abdominopelvic CT [10]. Abdominopelvic CT gives 100 % accuracy for diagnosing obturator hernia [8, 9, 11]. The abdominal plain radiographs may show findings consistent with small bowel obstruction and also air in the obturator region [6]. Ultrasonography is also a reliable modality for the diagnosis, but it is operator dependent. CT may reveal a hypodense area with surrounding fluid collection in the obturator foramen. The loop of small intestine herniating through the sac may be demonstrated.

The management for obturator hernia is invariably surgical intervention. Various approaches have been described which include the abdominal approach through the low midline incision, obturator approach, retro pubic approach, inguinal approach and the recent laparoscopic approach [5]. The repair may include either a simple closure of the hernia defect or closure by the use of a synthetic mesh [11].

Obturator hernia, though a rare entity, should always be kept in mind whenever a patient with Marfan's syndrome presents with features of intestinal obstruction and pain in the medial aspect of the thigh. Early diagnosis is established with computed tomography scan. Immediate surgical intervention by reducing the obturator hernia through the various approaches reduces the high morbidity and mortality rate.

Conflict of interest RP, VI, KK, PM and CV declare no conflict of interest.

References

1. Ha HI, Seo JB, Lee SH, Kang JW, Goo HW, Lim TH, Shin MJ (2007) Imaging of Marfan syndrome: multisystemic manifestations. Radiographics 27:989–1004. doi:10.1148/rg.274065171
2. Magid D, Pyeritz RE, Fishman EK (1990) Musculoskeletal manifestations of the Marfan syndrome: radiological features. AJR 155:99–104
3. De Paepe A, Devereux RB, Dietz H, Hannekam RCM, Pyeritz R (1996) Revised diagnostic criteria for the Marfan syndrome. Am J Med Genet 62(4):417–426
4. Hirano Y, Watanabe T, Kato H, Uchida T, Yoshida S, Hosokawa O (2010) Single-incision laparoscopic hernioplasty for obturator hernia. Surg Laparosc Endosc Percutan Tech 20(3):e144–e145. doi:10.1097/SLE.0b013e3181e10f98
5. Cai X, Song X, Cai X (2012) Strangulated intestinal obstruction secondary to a typical obturator hernia: a case report with literature review. Int J Med Sci 9(3):213–215. doi:10.7150/ijms.3894
6. Lin HH, Juan CW, Chew BT, Chao JH (2010) Obturator hernia. Int J Gerontol 4(2):104–106
7. Nakayama T, Kobayashi S, Shiraishi K, Nishiumi T, Mori S, Isobe K, Furuta Y (2002) Diagnosis and treatment of obturator hernia. Keio J Med 51(3):129–132
8. Yokoyama Y, Yamaguchi A, Isogai M, Hori A, Kaneoka Y (1999) Thirty-six cases of obturator hernia: does computed tomography contribute to postoperative outcome? World J Surg 23:214–217
9. Chang SS, Shan YS, Lin YJ, Tai YS, Lin PW (2005) A review of obturator hernia and a proposed algorithm for its diagnosis and treatment. World J Surg 29:450–454. doi:10.1007/s00268-004-7664-1
10. Polat C, Yavas Y, Kosar MN, Yazicioglu B, Degirmenci B, Arikan Y (2011) An unusual cause of mechanical intestinal obstruction: strangulated obturator hernia. Eur J Surg Sci 2(1):19–22
11. Mantoo SK, Mak K, Tan TJ (2009) Obturator hernia: diagnosis and treatment in the modern era. Singapore Med J 50(9):866–870