Clinical Case Report

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Combined laparoscopic and open technique for repair of congenital abdominal hernia

A case report of prune belly syndrome

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Abstract

Background: Prune belly syndrome (PBS) is a rare congenital disorder among adults, and the way for repairing abdominal wall musculature has no unified standard.

Materials and methods: We described combining laparoscopic and open technique in an adult male who presented with PBS. Physical examination and radiological imaging verified the case of PBS. The deficiency of abdominal wall musculature was repaired by combining laparoscopic and open technique using a double-deck complex patch.

Results: The patient successfully underwent abdominal wall repair by combining laparoscopic and open technique. Postoperative recovery was uneventful, and improvement in symptom was significant in follow-up after 3, 6, 12, and 24 months.

Conclusions: Combining laparoscopic and open technique for repair of deficiency of abdominal wall musculature in PBS was an exploratory way to improve life quality.

Abbreviations: CT = Computed tomography, MOS = Medical Outcomes Study, PBS = Prune Belly syndrome, SF-36 = 36-item short-form health survey.

Keywords: hernia, laparoscopy, prune belly syndrome

1. Introduction

Prune belly syndrome (PBS) is a rare congenital disorder, which is characterized by 3 abnormalities: an absence or deficiency of abdominal wall musculature, bilateral cryptorchidism, and a dilated dysmorphic urinary tract. This syndrome affects about 1 in 30,000[1] births of which about 96% of those affected are male.[2] Though some of the theories try to explain the possibility of genetic inheritance and possible association with trisomy 18 and 21[3]; however, the exact etiology of PBS remains unknown. We report a case of an adult male with PBS for its rarity and discussion of its intervention.

2. Case report (methods and results)

2.1. Clinical presentation and radiological findings

The patient's demographics, brief medical history, clinical presentation, laboratory results, and radiological findings were shown in Table 1. The patient came under category 3 PBS as described by Woodard in 1985[4] for unilateral cryptorchidism and deficiency of abdominal wall musculature (Fig. 1).

2.2. Surgical technique

After the patient signed the informed consent, we had a plan for the operation. The patient was placed supine with both arms adducted under general anesthesia, and 3 operative ports were used. A 10-mm trocar was placed to the superior to the umbilicus for camera, the rest 2 trocars were respectively in the right upper quadrant and left right upper. After the trocars were placed well, laparoscopic exploration was firstly operated, and the displacement of organs and cryptorchidism on the right side of the pelvis were found. Separation of the adhesion between liver and the abdominal wall was done laparoscopically with harmonic scalpel (Ethicon, Cincinnati, OH). A branch of vessel was found and we ensured it was a branch of vena cava by combining with preoperative abdominal CT scan (Fig. 2). It was difficult to be separated the tight adhesion in laparoscopic surgery. A 6-cm incision was made in the median upper abdominal. After slicing through the skin, subcutaneous tissue was separated carefully by electrotome until the branch of vena cava was exposed and separated. Separation was continued under direct vision until the deficiency area of abdominal wall musculature was sufficiently exposed. A 22×18 cm double-deck complex patch (Composix Kugel Mesh Patch; C. R. BARD, Murray Hill, NJ) was put in through the incision and then we sutured the median upper abdominal incision. The patch was flattened to overstep the edge of the deficiency of abdominal wall about 5 cm, and fixed with screwed nails after confirming avoiding the ribs or underlying nerves or vessel under laparoscopic vision (Fig. 3).

The operation time was about 120 minutes, and blood loss was about 100 mL. The patient's recovery was uneventful 10 days

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Table 1

Basic information.

Age, y	31
Sex	Male
Symptoms	Paroxysmal abdominal pain (especially forcibly)
History	Abdominal skin was paper-like when newborn (No intervention)
Duration of symptoms	>20 y
Physical examination	Wrinkled abdominal skin, about 16 × 12 cm Unilateral cryptorchidism
Family history of genetic or congenital anomaly	No
Abnormal in his physical growth and mental development	No
Routine hematological examination and renal function	No anomaly
Abdominal CT scan	Abdominal herniation, tortuosity of portal vein, and inferior vena cava Unknown tissue in the right side of the pelvis (cryptorchidism)

CT = computed tomography.

after operation, and the suggestion of urology department consultation was given. The Medical Outcomes Study (MOS) 36-item short-form health survey (SF-36) was referenced to use to evaluate quality of life in follow-up. The postoperative follow-up after 3, 6, 12, and 24 months showed the patient felt less pain and more confident in daily life.

3. Discussion

PBS is also known as Eagle-Barrett syndrome, the "prune-like" abdominal skin owing to an abdominal wall muscle defect, which is a characteristic manifestation and the first clue of PBS at physical examination.[5] PBS is rarely seen in adult population,[6–8] but the wrinkled abdominal skin provided an important clue for us to

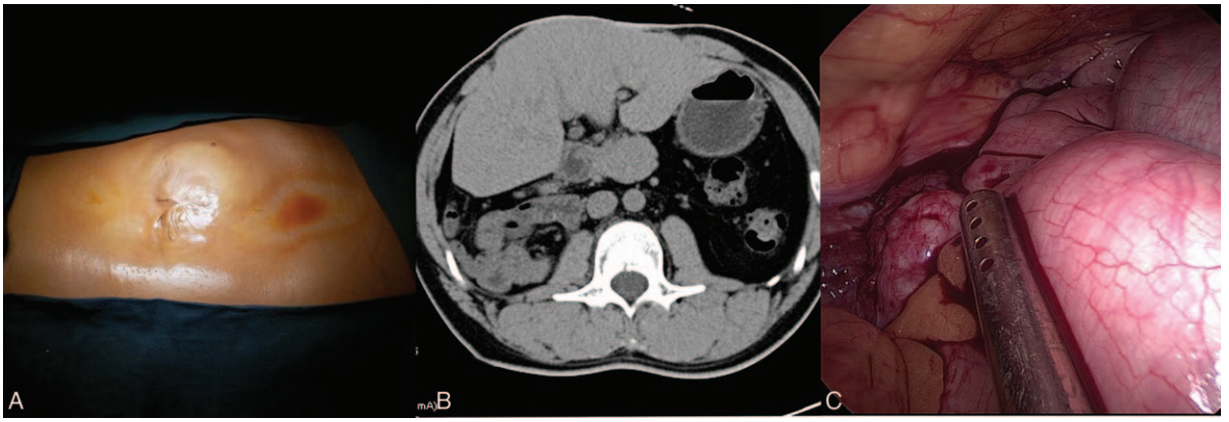

Figure 1. (A) Wrinkled abdominal skin, about 16 × 12 cm; (B) abdominal hernia; (C) cryptorchidism in the right side pelvic.

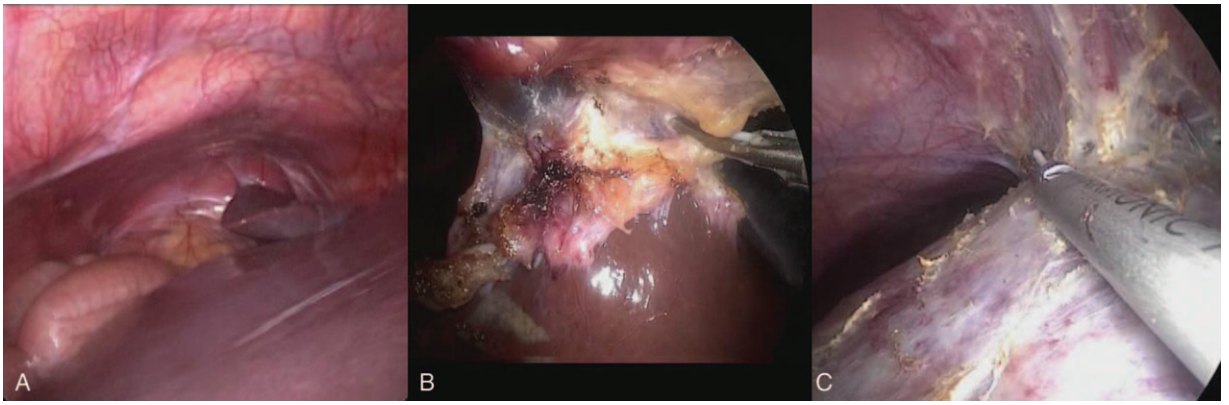

Figure 2. (A) Displacement of organs; (B) separation of liver and the abdominal wall; (C) a branch of vena cava was found (red arrow).

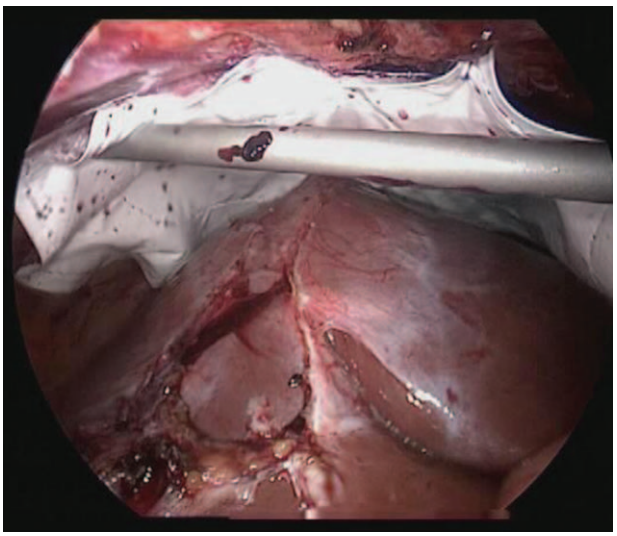

Figure 3. A double-deck complex patch was flattened and fixed.

consider congenital disease and diagnosis PBS for this patient who without history of surgical operation and trauma eventually. The etiology of PBS is still misted, though possible familial genetic inheritance was reported in some studies.[3] The prognosis of PBS is usually poor in newborns because of pulmonary hypoplasia or renal failure or a combination of congenital anomalies,[9] but the outcome of PBS in adults lacks effective evidence.

Multiple surgical interventions are necessary in surviving populations with PBS; however, the disorder remains challenging. A long-term follow-up of total abdominal wall reconstruction showed an effective intervention for absence or deficiency of abdominal wall musculature.[10] To reduce the surgical trauma and meet the demand of this patient, laparoscopic procedure was chosen to combine with open technique after discussing with the patient and our team members. Combined laparoscopic and open technique for abdominal wall repair was an exploratory treatment with less damage, faster recovery, and good cosmetic results. The abdominal CT scan should be analyzed carefully before operation to fully understand branches of vessels, for the reason that organ displacement may lead to anatomic abnormalities. The option of double-deck complex patch was according the deficiency of abdominal wall and over the edge about 5 to 6 cm. To evaluate the patient's recovery after operation, The SF-36 was referenced to use, and we called the patient many times to know the change in physiological and psychological. It is a pity that the patient's family members' life quality was lack of enough attention. Though our exploratory procedure acquired improvement in patient's life quality, this technical should be evaluated with long-term follow in large numbers of patients.

4. Conclusion

PBS is a rare congenital anomaly among adults, and the defect of abdominal wall muscle has a strong impact on daily life. Combined laparoscopic and open technique for repair of deficiency of abdominal wall musculature in PBS was an exploratory way to improve quality of life.

References

[1] Baird PA, MacDonald EC. An epidemiologic study of congenital malformations of the anterior abdominal wall in more than half a million consecutive live births. Am J Hum Genet 1981;33:470–8.
[2] Tagore KR, Ramineni AK, Vijaya Lakshmi ARNB. Prune belly syndrome. Case Rep Pediatr 2011;12:17–36.
[3] Ramasamy R, Haviland M, Woodard JR, et al. Patterns of inheritance in familial prune belly syndrome. Urology 2005;65:1227.
[4] Kelalis PP, King LR, Belman AB. Clinical Pediatric Urology. 2nd ed. Saunders, Philadelphia, PA:1985.
[5] Xu W, Wu H, Wang DX, et al. A case of prune belly syndrome. Pediatr Neonatol 2015;56:193–6.
[6] Tattoli F, De Prisco O, Gherzi M, et al. Prune-Belly Syndrome: a case report. G Ital Nefrol 2014;31:pii: gin/31.3.5.
[7] Salako AA, Takure AO, Olajide AO, et al. Prune belly syndrome in an adult Nigerian: case report. Afr J Med Med Sci 2009;38:357–60.
[8] Musone D, Nicosia V, D'Alessandro R, et al. Peritoneal dialysis in adult patients with prune belly syndrome: an impossible challenge? G Ital Nefrol 2013;30.
[9] Samal SK, Rathod S. Prune Belly syndrome: a rare case report. J Nat Sci Biol Med 2015;6:255–7.
[10] Lesavoy MA, Chang EI, Suliman A, et al. Long-term follow-up of total abdominal wall reconstruction for prune belly syndrome. Plast Reconstr Surg 2012;129:e104–9.