Full Title : Laparoscopic Assisted Anterior Resection in Kartegener's Syndrome

Running Title: Laparoscopy in Kartegener's Syndrome

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Kartagener's syndrome is a subgroup of primary ciliary dyskinesia. When situs inversus totalis (SIT), chronic sinusitis and bronchiectasis occur together a patient is said to have Kartagener's syndrome¹. In isolation SIT is a rare autosomal recessive inherited disorder with the incidence ranging between 1:20,000 and 1:10,000². While laparoscopic colonic surgery has been found to have equivalent oncological outcomes to open operations³, there are only eleven reports of laparoscopic surgery for colonic pathology in patients with situs inversus^{2,3-12}. This is the first case reported in the literature from Australia.

A 76 year old female with known Kartagener's syndrome presented with non-specific abdominal pain and rectal bleeding, and was later found to be anaemic. At colonoscopy a tumour was found in the sigmoid colon. Subsequent staging CT of the chest, abdomen and pelvis confirmed SIT with the tumour seen in the sigmoid colon in the right lower quadrant (Figure 1). There was no evidence of metastatic disease. There were no arterial anomalies, however the tributaries within the portal system converged at one point to form the portal vein (Figure 1). The patient had a number of comorbidities including obesity (BMI 34), valvular heart disease, atrial fibrillation (on warfarin), bronchiectasis and obstructive sleep apnoea.

The patient was placed in lithotomy position, both arms by her side. A hand port was inserted through a limited infra-umbilical vertical incision. A supra-umbilical 12mm balloon port was subsequently inserted using a cut-down technique. Two

5mm working ports were inserted under vision in the left iliac fossa and left upper quadrant respectively. A 30 Degree 10mm scope was used. The right-handed surgeon and assistant were positioned on the left side of the patient (Figure 2). Laparoscopic exploration showed complete transposition of abdominal viscera. The liver was located in the left upper quadrant and the spleen and stomach were in the right upper quadrant.

The omentum was dissected from the transverse colon. The splenic flexure in the right upper quadrant was mobilised. The flexure was lower and technically easier to mobilise than the orthotopic splenic flexure. The sigmoid colon, descending colon and upper rectum were then mobilised. The inferior mesenteric artery was identified and high ligated with a linear EndoGIA^R stapler. The inferior mesenteric vein (IMV) was located lateral to the DJ flexure to the right of midline and was ligated with Hem-o-lok^R clips (Figure 3). The bowel anastomosis was performed using a linear TL 60^R stapler and circular EC529^R stapler. A leak test was negative. Compared with orthotopic patients operating time was equivalent and blood loss was approximately 200ml. Post operatively the patient recovered well, bowels opened day 5, with discharge day 7. Discharge was delayed by two days due to mild cellulitis around IV cannula.

Histopathology showed a moderately differentiated adenocarcinoma, 35mm, with tumour extension through the muscularis propria into the pericolic fat. Sixteen

lymph nodes were harvested with 4/16 involved. Margins were clear. The patient was staged as IIIC, T4aN2aM0 (AJCC 7th edition).

Primary ciliary dyskinesia is inherited as an autosomal recessive disease¹. Left-right asymmetry is established during gastrulation through cell-cell interactions centred at the primitive node¹³. In situs inversus viscerum totalis, the handedness of all the viscera is reversed. Situs inversus in absence of Kartagener's syndrome is rarely complete and more often exhibits heterotaxy, where there is a discordance of sidedness¹³. It is important to note that when there is heterotaxia there is abnormal arrangement of organs e.g. midline liver and major blood vessels which differs from the orderly arrangement of either situs solitus or situs inversus¹⁴. Careful preoperative planning is required to ensure no vascular anomalies are present⁷, particularly if there is heterodextra. Vascular anomalies have been found in patients with Kartagener's syndrome, such as an absent inferior vena cava¹⁵.

In this case for a right-handed surgeon the overall technical difficulty was comparable to surgery in orthotopic patients, with the splenic flexure comparatively easier. In this case the surgeon began the mobilisation with a flexure down approach, rather than medial to lateral or lateral to medial. Combination techniques with flexibility between lateral dissection, medial dissection or flexure down approach need to be used, as patients will not always be mirror image. The position of operator was mirror imaged, with the operator, camera assistant and working ports

on the left, and the hand port in usual position in the midline. Operative time and blood loss were comparable to orthotopic laparoscopic case.

With respect to anaesthetic preparation, congenital cardiac abnormalities are occasionally associated with primary ciliary dyskinesia, including transposition of great vessels. Therefore preoperative cardiac evaluation is recommended. In addition ECG sensors are placed on the right side given the dextrocardia. Laparoscopic assisted surgery should be considered in patients with Kartagener's syndrome and situs inversus totalis. Our case demonstrates that laparoscopy is safe with a good oncologic resection and nodal harvest.

References

1. Uptodate^R. [updated 11 March 2016]. Available from http://www.uptodate.com/contents/primary-ciliary-dyskinesia-immotile-cilia-syndrome?source=search_result&search=kartageners&selectedTitle=1~27
2. Kim Y, Ryu H, Kim D, Kim I. Double primary malignancies associated with colon cancer in patients with situs inversus totalis: two case reports. *World J Surg Oncol*. 2011 Sep 23;9:109.
3. Green B, Marshall H, Collinson F, Quirke P, Guillou P, Jayne D, Brown J. Long-term follow-up of the Medical Research Council CLASSICC trial of conventional versus laparoscopically assisted resection in colorectal cancer. *Br J Surg*. 2013 Jan;100(1):75-82.

4. Kobus C, Targarona E, Bendahan G, Alonso V, Balague C, Vela S, Garriga J, Trias M. Laparoscopic surgery in situs inversus: a literature review and report of laparoscopic sigmoidectomy for diverticulitis in situs inversus. *Langenbecks Arch Surg*. 2004 Oct;389(5):396-9.
5. Hirano Y, Hattori M, Douden K, Hashizume Y. Single-incision laparoscopic surgery for colon cancer in patients with situs inversus totalis: report of a case. *Indian J Surg*. 2015 Apr;77(Suppl 1):26-8.
6. Yaegashi M, Kimura T, Sakamoto T, Sato T, Kawasaki Y, Otsuka K, Wakabayashi G. Laparoscopic sigmoidectomy for a patient with situs inversus totalis: effect of changing operator position. *Int Surg*. 2015 Apr;100(4):638-42.
7. Sumi Y, Tomono A, Suzuki S, Kuroda D, Kakeji Y. Laparoscopic hemicolectomy in a patient with situs inversus totalis after open distal gastrectomy. *World J Gastrointest Surg*. 2013 Feb 27;5(2):22-6.
8. Berencsi A, Bezsilla J, Sikorszki L, Temesi R, Bende S. Laparoscopic sigmoid resection in total situs inversus. *Magy Seb*. 2013 Feb;66(1):30-3.
9. Kim H, Choi G, Park J, Lim K, Jang Y, Park S, Jun S. Laparoscopic right hemicolectomy with D3 lymph node dissection for a patient with situs inversus totalis: report of a case. *Surg Today*. 2011 Nov;41(11):1538-42.
10. Jobanputra S, Safar B, Wexner SD. Laparoscopic diverticular resection with situs inversus totalis (SIT): report of a case. *Surg Innov*. 2007 Dec;14(4):284-6.

11. Fujiwara Y, Fukunaga Y, Higashino M, Tanimura S, Takemura M, Tanaka Y, Osugi H. Laparoscopic hemicolectomy in a patient with situs inversus totalis. *World J Gastroenterol*. 2007 Oct 7;13(37):5035-7.
12. Fang J, Zheng Z, Wei B, Chen T, Lei P, Huang J, Huang L, Wei H. Laparoscopic resection for rectal cancer and cholecystectomy for patient with situs inversus totalis. *J Minim Access Surg*. 2015 Jul-Sep;11(3):210-2
13. Davies H, Slater GH, Bailey M. Laparoscopic sigmoid colectomy for diverticular disease in a patient with situs inversus. *Surg Endosc*. 2003;17:160–161.
14. Winer-Muram HT. Adult presentation of heterotaxic syndromes and related complexes. *J Thorac Imaging*. 1995 Winter;10(1):43-57.
15. Tabeta H, Hiroshima K. A case of Kartagener's syndrome with bilateral superior vena cava and absent inferior vena cava. *Nihon Kyobu Shikkan Gakkai Zasshi*. 1991 Apr;29(4):507-11.
16. Larsen's human embryology 4th ed. Schoenwolf, Gary C; Larsen, William J, (William James). Philadelphia, PA: Elsevier/Churchill Livingstone, c2009.

Figure 1 Abdominal CT .tif

Figure 2 port position.tif.png

Figure 3 Intraop.tif

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