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 syndrome\(^1\). In isolation SIT is a rare autosomal recessive inherited disorder with the incidence ranging between 1:20,000 and 1:10,000\(^2\). While laparoscopic colonic surgery has been found to have equivalent oncological outcomes to open operations\(^3\), 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\textsuperscript{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\textsuperscript{R} clips (Figure 3). The bowel anastomosis was performed using a linear TL 60\textsuperscript{R} stapler and circular EC529\textsuperscript{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 disease. Left-right asymmetry is established during gastrulation through cell-cell interactions centred at the primitive node. In situ 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 sidedness. 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 inversus. Careful preoperative planning is required to ensure no vascular anomalies are present, particularly if there is heterodextra. Vascular anomalies have been found in patients with Kartagener's syndrome, such as an absent inferior vena cava.

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


