Endoscopic management of Mirizzi’s syndrome

R E England, D F Martin

Abstract

Background—The accepted management of Mirizzi’s syndrome is surgical, but endoscopic and percutaneous management have been described.

Aim—To review our experience of endoscopic intervention for Mirizzi’s syndrome.

Patients and Methods—ERCP reports of patients presenting for endoscopic management of choledocholithiasis between March 1989 and June 1995 were reviewed. Those with cholangiographic evidence of Mirizzi’s syndrome were selected for study. Patient records and cholangiograms were reviewed and follow up was recorded from the notes or by telephone contact with patients, their relatives, or doctors.

Results—Twenty five patients had Mirizzi’s syndrome. Sixteen were female and their median age was 67 years (range 28–91). Ten presented with painless jaundice, nine with painful jaundice, four with cholangitis, and two had pain as their only symptom. Twelve were referred for surgery and 11 of these had preliminary endoscopic therapy. Thirteen have been treated solely with endoscopic therapy. Treatment in this group was aimed at relieving jaundice and removing stones. Stones were completely removed in three patients. Nine patients have been treated with long term stents, and one awaits extracorporeal shockwave lithotripsy of the gall bladder. Complications of treatment occurred in four of 25 after ERCP.

Conclusions—Endoscopic treatment of Mirizzi’s syndrome is effective as a temporising measure before surgery and can be definitive treatment for unsuitable surgical candidates.

Methods

We searched our database for all patients with a cholangiographic diagnosis of Mirizzi’s syndrome between March 1989 and June 1995 and reviewed their notes, and retrograde cholangiograms. Demographic details, presentation, clinical and cholangiographic findings, endoscopic and surgical treatment, and complications of treatment were recorded. Patients were followed up by contacting their primary physician or referring doctor. Follow up was judged complete if the patient had cholecystectomy, cholangiography showed clear ducts, and they were discharged from review, or if the patient died.

Mirizzi’s syndrome was described in 1948 and consists of a stone impacted in the neck of the gall bladder or in the cystic duct, causing inflammation and extrinsic compression of the common duct, leading to obstructive jaundice. Mirizzi’s syndrome may be subdivided into types I and II: in the first there is extrinsic compression of the common duct by an inflamed or enlarged gall bladder (Fig 1) while in type II the stone erodes from the gall bladder or the cystic duct into the common duct resulting in a cholecystcholedochal fistula (Fig 2). The syndrome is rare occurring in only 0.7–1.1% of patients undergoing cholecystectomy. The established treatment is surgical but with the development of endoscopic and percutaneous biliary techniques non-surgical therapy has been described. We describe our experience of Mirizzi’s syndrome, emphasising the role of ERCP in diagnosis and treatment.

Keywords: Mirizzi’s syndrome, ERCP, endoscopic therapy.
TABLE 1  Clinical details of the surgically treated group

<table>
<thead>
<tr>
<th>Patient</th>
<th>Sex</th>
<th>Age (y)</th>
<th>Presentation</th>
<th>Mirizzi type</th>
<th>Endoscopic treatment</th>
<th>Surgery</th>
<th>Comment</th>
</tr>
</thead>
<tbody>
<tr>
<td>4</td>
<td>F</td>
<td>37</td>
<td>Jaundice+pain</td>
<td>Type I</td>
<td>ES, CBD stent</td>
<td>LC at 15 days</td>
<td>Stone had been fragmented. Alive and well</td>
</tr>
<tr>
<td>8</td>
<td>F</td>
<td>65</td>
<td>Jaundice+pain</td>
<td>Type I</td>
<td>ES</td>
<td>LC converted to OC at 365 days</td>
<td>Surgery recommended. Alive and well</td>
</tr>
<tr>
<td>9</td>
<td>M</td>
<td>56</td>
<td>Jaundice+pain</td>
<td>Type I</td>
<td>ES</td>
<td>OC+CBD exploration at 3 days OC at 116 days</td>
<td>Surgery recommended. Alive and well</td>
</tr>
<tr>
<td>11</td>
<td>F</td>
<td>60</td>
<td>Jaundice</td>
<td>Type I</td>
<td>ES, CBD stent</td>
<td>OC+CBD exploration at 1 day OC+CBD exploration at 7 days OC at 120 days OC at 63 days OC at 165 days OC at 9 days</td>
<td>Invasive carcinoma of gall bladder on histology and died from this 388 days after cholecystectomy Alive and well No response to endoscopic treatment Alive and well after surgery Polya gastrectomy. Alive and well ES not performed as young and wanted to preserve sphincter. Alive and well Surgery postponed till renal failure resolved Alive and well Coincident pseudoaneurysm of cystic artery with haemobilia. Hepatic embolisation performed, bleeding stopped, but died with multi-organ failure at 18 days No improvement with stent, developed acute cholecystitis. Alive and well after surgery Alive and well</td>
</tr>
<tr>
<td>12</td>
<td>F</td>
<td>28</td>
<td>Jaundice+pain</td>
<td>Type I</td>
<td>ES, CBD stent</td>
<td>OC+CBD exploration at 1 day OC+CBD exploration at 7 days OC at 120 days OC at 63 days OC at 165 days OC at 9 days</td>
<td>Laparotomy at 11 days. Unable to remove gall bladder No response to endoscopic treatment Alive and well after surgery Polya gastrectomy. Alive and well ES not performed as young and wanted to preserve sphincter. Alive and well Surgery postponed till renal failure resolved Alive and well Coincident pseudoaneurysm of cystic artery with haemobilia. Hepatic embolisation performed, bleeding stopped, but died with multi-organ failure at 18 days No improvement with stent, developed acute cholecystitis. Alive and well after surgery Alive and well</td>
</tr>
<tr>
<td>14</td>
<td>M</td>
<td>65</td>
<td>Cholangitis</td>
<td>Type II</td>
<td>ES, CBD stent, mechanical lithotripsy</td>
<td>OC+CBD exploration at 1 day OC+CBD exploration at 7 days OC at 120 days OC at 63 days OC at 165 days OC at 9 days</td>
<td>No response to endoscopic treatment Alive and well after surgery Polya gastrectomy. Alive and well ES not performed as young and wanted to preserve sphincter. Alive and well Surgery postponed till renal failure resolved Alive and well Coincident pseudoaneurysm of cystic artery with haemobilia. Hepatic embolisation performed, bleeding stopped, but died with multi-organ failure at 18 days No improvement with stent, developed acute cholecystitis. Alive and well after surgery Alive and well</td>
</tr>
<tr>
<td>15</td>
<td>M</td>
<td>70</td>
<td>Jaundice+pain</td>
<td>Type I</td>
<td>CBD stent, cystic duct dissected, cystic duct stent</td>
<td>OC at 120 days OC at 63 days</td>
<td>No response to endoscopic treatment Alive and well after surgery Polya gastrectomy. Alive and well ES not performed as young and wanted to preserve sphincter. Alive and well Surgery postponed till renal failure resolved Alive and well Coincident pseudoaneurysm of cystic artery with haemobilia. Hepatic embolisation performed, bleeding stopped, but died with multi-organ failure at 18 days No improvement with stent, developed acute cholecystitis. Alive and well after surgery Alive and well</td>
</tr>
<tr>
<td>17</td>
<td>F</td>
<td>28</td>
<td>Jaundice+pain</td>
<td>Type I</td>
<td>CBD stent, cystic duct dissected, cystic duct stent</td>
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</tr>
<tr>
<td>20</td>
<td>M</td>
<td>59</td>
<td>Cholangitis+renal failure</td>
<td>Type I</td>
<td>ES, CBD stent</td>
<td>OC at 165 days</td>
<td>No response to endoscopic treatment Alive and well after surgery Polya gastrectomy. Alive and well ES not performed as young and wanted to preserve sphincter. Alive and well Surgery postponed till renal failure resolved Alive and well Coincident pseudoaneurysm of cystic artery with haemobilia. Hepatic embolisation performed, bleeding stopped, but died with multi-organ failure at 18 days No improvement with stent, developed acute cholecystitis. Alive and well after surgery Alive and well</td>
</tr>
<tr>
<td>22</td>
<td>F</td>
<td>71</td>
<td>Cholangitis</td>
<td>Type II</td>
<td>ES, CBD stents, mechanical lithotripsy</td>
<td>OC at 9 days</td>
<td>No response to endoscopic treatment Alive and well after surgery Polya gastrectomy. Alive and well ES not performed as young and wanted to preserve sphincter. Alive and well Surgery postponed till renal failure resolved Alive and well Coincident pseudoaneurysm of cystic artery with haemobilia. Hepatic embolisation performed, bleeding stopped, but died with multi-organ failure at 18 days No improvement with stent, developed acute cholecystitis. Alive and well after surgery Alive and well</td>
</tr>
<tr>
<td>23</td>
<td>F</td>
<td>58</td>
<td>Jaundice+pain</td>
<td>Type I</td>
<td>CBD stent</td>
<td>OC at 9 days</td>
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</tr>
<tr>
<td>25</td>
<td>M</td>
<td>62</td>
<td>Pain</td>
<td>Type I</td>
<td>ES done previously for CBD stone.</td>
<td>Awaits surgery</td>
<td>No response to endoscopic treatment Alive and well after surgery Polya gastrectomy. Alive and well ES not performed as young and wanted to preserve sphincter. Alive and well Surgery postponed till renal failure resolved Alive and well Coincident pseudoaneurysm of cystic artery with haemobilia. Hepatic embolisation performed, bleeding stopped, but died with multi-organ failure at 18 days No improvement with stent, developed acute cholecystitis. Alive and well after surgery Alive and well</td>
</tr>
</tbody>
</table>

ESWL=extracorporeal shockwave lithotripsy, CBD=common bile duct, OC=open cholecystectomy, LC=laparoscopic cholecystectomy.
with plastic stents (8) and sphincterotomy (6), extracorporeal shockwave lithotripsy of the gall bladder (2), and adjuvant mechanical lithotripsy (2). Sphincterotomy was avoided in case 17 as she was young, and was not performed for technical reasons in case 15 as the patient had a Polya gastrectomy. Case 23 had stenting without sphincterotomy as the initial diagnosis was thought to be a malignant stricture; however, on review of her imaging Mirizzi’s syndrome was diagnosed. Treatment in this group was aimed at relieving jaundice and allowing elective planned cholecystectomy. Surgery was performed at a median time of 15 days after ERCP (range 1–365 days).

Three patients failed to improve after endoscopic therapy: case 23 developed acute cholecystitis, case 14 had persistent cholangitis, and case 22 was operated on for continued haemobilia but surgery failed to identify the source and cholecystectomy was not possible because of severe inflammation of Calot’s triangle. This patient subsequently had transcatheter embolisation of a pseudoaneurysm of the cystic artery but although this controlled her haemorrhage she succumbed a week later to multorgan failure. All but one of the patients who had surgery required an open procedure.

Thirteen patients had been treated solely with endoscopic therapy (Table II). Treatment was aimed at relieving jaundice and removing stones. Those in whom complete clearance was not achieved were treated with long term stenting. The stones were completely removed in three patients (cases 2, 6, and 21) using a combination of sphincterotomy, mechanical lithotripsy, methyl tert butyl ether, extracorporeal shockwave lithotripsy, and common duct bile stents. Nine patients were treated with long term stenting and of these five are alive and well at follow up ranging from four months to five years and two months. Four patients in this group died, two of non-biliary causes, another from bronchopneumonia 10 days after ERCP, and one of secondary biliary cirrhosis at two years and seven months. The remaining patient in this group is awaiting extracorporeal shockwave lithotripsy of the gall bladder.

Complications of endoscopic treatment occurred in four of 25 patients: (case 7) bronchopneumonia after ERCP, (case 23) acute cholecystitis after stenting, (case 11) liver abscess treated conservatively with antibiotics, and (case 5) secondary biliary cirrhosis.

The overall mortality of the group was seven of 25, with a 30 day mortality of two of 25. Four of seven deaths were biliary related: (case 5) secondary biliary cirrhosis; (case 7) bronchopneumonia after ERCP; (case 11) carcinoma of gall bladder; (case 22) multiorgan failure secondary to prolonged haemobilia from a pseudoaneurysm of the cystic artery.

**Discussion**

Mirizzi syndrome, a rare complication of gallstone disease, is increasingly recognised with the widespread availability of cholangiography. In the past the diagnosis was often first made at laparotomy, and because of the severe inflammation in Calot’s triangle associated with a shrunken gall bladder, the risk of bile duct injury was high. The syndrome, as originally described, comprised a stone
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impacted in the cystic duct or neck of the gall bladder and a functional disorder of a putative sphincter within the common hepatic duct, without any lesion of the common bile duct. Inflammation, aberrant vessels or an impacted cystic duct stone were thought to stimulate contraction of this 'sphincter' resulting in obstructive jaundice and biliary fistula. It is now known that such 'sphincter' exists and that biliary obstruction results from extrinsic compression of the common duct by the inflamed gall bladder and that the inflammatory process may result in common duct disruption.

In 1982 McSherry classified Mirizzi's syndrome into type I and type II based on cholangiographic findings at ERCP. Type I is present when there is extrinsic compression of the common duct by a stone in the gall bladder or in the cystic duct (Fig 1). Type II is present when a cholecystocholedochal fistula exists (Fig 2). Type II has been further subclassified by Csendes et al. into the at types IVA to VIB denoting the size of the defect in the common duct—type IVA: erosion of less than one third the circumference of the bile duct; type IIB: erosion of up to two thirds of the common duct; type IV: complete destruction of the entire wall of the common duct. The size of the fistula in Csendes' series was determined at per operative cholangiography. In our experience exact quantification of fistula size at pre-operative cholangiography is impossible, and we therefore use McSherry's classification. The importance of recognising type II Mirizzi is stressed as the operative approach differs in the presence of a cholecystocholedochal fistula. An open surgical approach is considered more appropriate than a laparoscopic approach if a fistula is present as dissection may be difficult and a direct common duct repair or bilioenteric anastomosis is necessary. Indeed an open procedure is often necessary for Mirizzi's syndrome irrespective of type due to severe inflammation and this is reflected in our series where all but one of our patients who had surgery required an open procedure.

There are a number of published reports of non-surgical management of Mirizzi's syndrome. Percutaneous management is usually reserved for patients who have failed endoscopic access and who are unsuitable surgical candidates. Cairns and Oxtoby describe percutaneous cholecystodocholithotomy and electrohydraulic lithotripsy, performed under light general anaesthesia, in patients with Mirizzi type I. Adam used a percutaneous metal stent to decompress the biliary tree in an elderly patient with Mirizzi syndrome, and left the gall stones in situ. This patient was well at follow up at 2-5 years.

Endoscopic management of Mirizzi syndrome comprises biliary drainage of the common duct with or without gall bladder drainage, and stone removal with either basket or balloon, mechanical, electrohydraulic or extracorporeal lithotripsy or dissolution therapy. Endoscopic sphincterotomy is followed by either placement of a nasobiliary catheter or more commonly insertion of an indwelling prosthesis into the common bile duct, cystic duct or both. After resolution of jaundice or cholangitis attempts are made to clear the stones when surgery is not deemed appropriate. The largest published series of solely endoscopic management of Mirizzi syndrome is by Binmoeller et al. In this series electrohydraulic lithotripsy was attempted under cholangioscopic guidance to 14 patients with Mirizzi's syndrome. In all patients the stone was impacted distally in the cystic duct and stone size ranged from 1·5 cm to greater than 3 cm. Stones were cleared in all 14 patients with only one complication: there was leakage of contrast from the duct system into the peritoneal cavity after removal of a large 3·5 cm stone. This leak was caused by pressure necrosis from the large stone rather than injury to the duct by the electrohydraulic lithotripsy probe and resolved spontaneously with conservative management. The authors conclude that this is a safe and effective method of surgery. Limitations of this technique include the need for considerable expertise at ERCP, cost of equipment including mother and baby scope system, and electrohydraulic lithotripsy probe per patient treatment session and the procedure is time consuming. In addition stones must be accessible to the electrohydraulic lithotribe and hence Mirizzi's syndrome patients with a stone impacted in Hartmann's pouch are not suitable for this treatment (12 of 25 of our patients).

In our series almost half had definitive surgery after endoscopic therapy. As can be seen from Table I those who were referred for surgery were generally fit patients or patients who did not initially respond to endoscopic drainage (cases 14, 22, and 23). The importance of preliminary cholangiography should be emphasised: the diagnosis was only suspected in one of 25 patients after ultrasound. Cholangiography permits diagnosis, classification of type, and provides a roadmap for subsequent surgery. If ERCP is performed attempts can also be made to dislodge or remove the obstructing calculus at the initial diagnostic procedure. Interestingly in our series only two (case 14 and 22) of the surgically treated group had type II Mirizzi syndrome. Case 14 had an initial ERCP with sphincterotomy and mechanical lithotripsy and partial removal of the obstructing calculus. A stent was left for temporary drainage; however, the patient had persistent cholangitis and was referred for cholecystectomy and common duct exploration. Surgery was difficult as anatomical landmarks were very distorted and there was a large fistula between the shrunken gall bladder and the common duct with no identifiable cystic duct. During dissection a hole was made accidentally in the right main duct. The gall bladder was opened, stone debris was removed from the duct with the endoscopic stent. The gall bladder was resected and the T tube placed across the right hepatic duct with the gall bladder stone removed and the T tube was directly repaired over a T tube. A second T tube was placed across the right hepatic duct defect. The T tubes were removed after a subsequent cholangiogram, which confirmed clear ducts and no biliary leak. Case 22 was a patient who at ERCP for cholangitis was found...
to have haemobilia. Sphincterotomy was performed and attempts to remove the large 3 cm calculus with mechanical lithotripsy failed. Two large bony stents were placed for temporary drainage; however, a laparotomy was performed for continued haemorrhage. Surgery proved difficult because of anatomical distortion and bleeding and a cholecystectomy and stone removal were not possible. A T tube was placed in the common duct for drainage. Subsequent selective hepatic artery embolisation was performed of a pseudoaneurysm of the cystic artery and although this controlled her haemorrhage she succumbed to multiorgan failure. Surgery for Mirizzi type II is difficult and may be hazardous and endoscopic management may be preferable in this group as one can avoid the difficult surgical repair of the choledochocholangioenteric fistula.

Seven of nine patients in this series with Mirizzi type II were treated endoscopically.

The timing between preliminary endoscopic treatment and surgery was very variable in our series (Table I) and reflected patient response to initial biliary drainage as well as surgical waiting lists for elective cholecystectomy. Provided the patient’s symptoms are relieved with initial endoscopic treatment definitive surgical treatment can be postponed until the patient is fit for elective surgery. In patients unsuitable or unwilling to undergo surgery further endoscopic attempts are made to disimpact and remove the stones, using a combination of mechanical lithotripsy, dissolution agents or extracorporeal shock wave lithotripsy. We were successful in doing this in three patients, cases 2 and 21 who were medically unfit and case 6 who refused surgery.

There were nine patients who had long term stenting. The median age of this group was 78 years, range 69–91, and all but one (case 18) had coexistent medical problems making them unsuitable surgical candidates (Table II). Bergman et al.13 recently reported the outcome of long term stenting in 58 of 117 patients for irretrievable common duct stones and showed a 40% complication rate, most commonly cholangitis, and nine of 44 patients had a biliary related death. The authors conclude that permanent biliary stenting (that is, stent exchange only for recurrent symptoms) should be restricted to patients unfit for surgery or further endoscopic therapy and those with a short life expectancy and we agree with this policy. In our unit we change long term stents at yearly intervals but we review patients if they have recurrent symptoms between stent exchanges. Of the nine patients with long term stenting five are alive and well. Of the four who died there was one biliary death (case 5) from secondary biliary cirrhosis. This patient was initially referred for ERCP with suspected primary biliary cirrhosis and Mirizzi’s syndrome was diagnosed. She was a diabetic and not anxious for surgery. She had five ERCPs with mechanical lithotripsy and extracorporeal shock wave lithotripsy, however her stone remained. She was asymptomatic with stenting until finally presenting at two years and seven months with ascites, and oesophageal varices, but without jaundice. She succumbed after gastrointestinal haemorrhage. In retrospect this patient may have had a better outcome had she agreed to surgery. Complications of treatment occurred in a further three patients: bronchopneumonia resulting in the demise of one patient at 10 days; acute cholecystitis after stenting treated surgically; and a liver abscess that was treated conservatively.

The latter patient (case 11) had an open cholecystectomy at 116 days and histological examination of her gall bladder revealed an unsuspected gall bladder carcinoma. Coexistence of gall bladder carcinoma and Mirizzi’s syndrome has been recognised previously14 and is not surprising as both conditions are associated with long term gall stone disease.

In conclusion ERCP plays an important part in diagnosis and provides not only a roadmap for surgery but permits therapeutic decompression of the biliary tree as a temporising measure before surgery. In some patients endoscopic clearance of stones is possible, and long term stenting for those in whom clearance is not achieved is an alternative, effective option to surgery — with the caveat that these patients should be followed up at regular intervals to prevent complications. Finally those patients with Mirizzi type II may be more suited to endoscopic therapy as this can avoid what is often very difficult open surgery.

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