Mirizzi syndrome: Report of a case and the challenge of management in our environment.

OO Akute[1,2], BJ Alegbeleye[1] and AO Afolabi[1]

Department of Surgery[1,2], College of Medicine, University of Ibadan, Ibadan, and New Address: Bowen University Teaching Hospital[2], Ogbomosho, Nigeria.

Abstract

Background: Mirizzi Syndrome is a rare complication of cholelithiasis reported to occur in 1% of all patients with gall stones and an incidence of 0.7-1.4% in all cholecystectomies. It is characterized by an impaction of a large calculus in the Hartman's pouch of the gall bladder (GB) or in the cystic duct, causing an extrinsic obstruction of the common hepatic duct. This can, with time, result in varying degrees of fistula formation between the duct and the GB. Types I, IIa, b and c have been described depending on the circumference of the duct involved in the cholecysto-choledochal fistula. This syndrome presents clinically as surgical jaundice. The pre-operative diagnosis is difficult as well as the surgical management of the type II subtypes. The aim of this paper is to draw attention to this clinico-pathological entity as it occurs with the same frequency in our environment as in the environment with high incidence of cholelithiasis.

Method: We report a case in our practice (Mirizzi Type IIa) and discuss the difficulties encountered in pre-operative diagnosis and subsequent management. The literature is also reviewed

Results: The diagnosis of Type IIa Syndrome was made on the operating table. The on-table cholangiogram was inconclusive. A choledochoplasty was performed over a T-tube and this was removed after 12 weeks. Patient has done well thereafter.

Conclusion: The local surgeon is advised to have a high index of suspicion about this condition so as not to be caught unawares.

Keywords: *Mirizzi syndrome, pre-operative diagnosis, operative management.*

Résumé

Introduction : Le syndrome de Mirizzi est une rare complication de la cholelithiasie chez 1% des patients ayant des calculs biliaires et une incidence de 0.7-1.4% chez toutes les cholécystectomies. Elle est caractérisée par une concentration de larges calculs dans la poche d'Hartman de la vésicule biliaire ou de la voie cystique, causant une obstruction extrinsèque de la voie hépatique commune.

Méthodologie : En fonction du temps, ceci peux résulter a différent dégrée de formation de fistules entre les voies et la vésicule biliaire. Les types I, II a, b et c ont été décrit dépendamment de la circonférence des voies impliquées dans la fistule cholécysto-choledochale. Ce syndrome se présente comme la jaunisse chirurgicale. Le diagnostic pré-opérative est aussi difficile que le soin chirurgical des sous types de types II. Le but de cet article est de tirer l'attention sur cette entité clinico-pathologique vu sa fréquence d'apparition dans notre environnement avec une incidence élevée de cholelithiasie

Résultats : Nous rapportons un cas dans notre pratique de Mirizzi type IIa et discutons les difficultés rencontrées dans le diagnostic pré-opérative et les soins. La littérature est aussi revue. Les résultats du diagnostic de ce syndrome de type IIa ont été faite dans la salle d'opération et le cholangiogramme était non-concluant. La choledochoplastie était faite par le tube en T et a été enlevé après 12 semaines et l'individu regagnant sa santé parfaite.

Conclusion : Le chirurgien local a conseillé 'avoir un indexe de suspicion et attention élevées par rapport à cette condition.

Introduction

Gallstones are still relatively an uncommon disease in our environment when compared to the technologically advanced countries and even Asia, though the trend is rising [1].

Mirizzi syndrome (M/S) which is an uncommon disease deserves to be chronicled when it occurs in our practice. This syndrome, which was first described in 1948 by Mirizzi is characterized by the impaction of a big stone in the cystic duct or the Hartmann's pouch of the gall bladder (GB) whilst resting on the common hepatic duct (CHD). This results in mechanical obstruction of the CHD with attendant intermittent or persistent jaundice. There may be varying degree of fistulation to compound the picture. It is

Correspondence: Dr. O.O. Akute, Department of Surgery, Bowen University Teaching Hospital, Ogbomoso, Nigeria. E-mail: papakute@yahoo.co.uk

reported to occur in 1% of all patients with gallstones with a frequency of 0.7-1.4% of all cholecystectomies [2].

A patient is presented and a review of the literature done to draw attention to the condition in our environment and highlight the challenges of diagnosis and treatment.

Case presentation

A 54 year old clergyman was referred to our service with a 5-month history of intermittent jaundice, dull right hypochondrial pain and weight loss. He had a history of pale, bulky foul smelling stool (3-4 episodes daily), deeply coloured urine, anorexia and occasional post – prandial vomiting (1-2 episodes weekly). He had a Truncal Vagotomy and Pyloroplasty for a duodenal ulcer in 1978.

Physical examination revealed a chronically ill-looking emaciated middle aged man with jaundice. Besides a mid-line supra-umbilical scar, there was no other remarkable finding. The clinical diagnosis was surgical jaundice, secondary to peri-ampullary carcinoma or choledocholithiasis.

An abdominal USS showed a smooth enlarged liver, a gall bladder containing a huge oval gall stone and a normal caliber common bile duct (CBD). The liver function tests confirmed conjugated hyper-bilirubinaemia, elevated transaminases and alkaline phosphatase. Urinalysis revealed bilirubinuria. All other relevant tests including HIV screening, HbSag, alpha-fetoprotein, stool, E/U and serum creatinine, FBC, INR-ratio, chest x-ray and ECG were normal. The computed – axial tomography was omitted because of cost.

The patient was prepared for operation and had a laparotomy on 26/05/08. The findings at operation included – dense adhension in the whole sub-hepatic space, a complex mass consisting of a shrunken GB communicating with the right side of a grossly dilated duct and in the centre of everything was a big stone (4cm in diameter), causing cholecysto-choledochal fistula (Mirizzi Type IIa). On table cholangiogram under image Intensifier was inconclusive.

The cholecysto-choledochal mass was then opened and the stone removed. A partial cholecystectomy was carried out and a choledochoplasty performed over a T-tube using the remnant of the GB. His post-operative course was uneventful and serial clamping of the T-tube was commenced on 14th day post-operatively. A T-tube cholangiogram (fig. 1) demonstrated spillage of contrast around the T-tube but the duodenal loop was well outlined with contrast. The patient was discharged home with the T-tube in situ. An attempt to repeat T-tube cholangiogram under

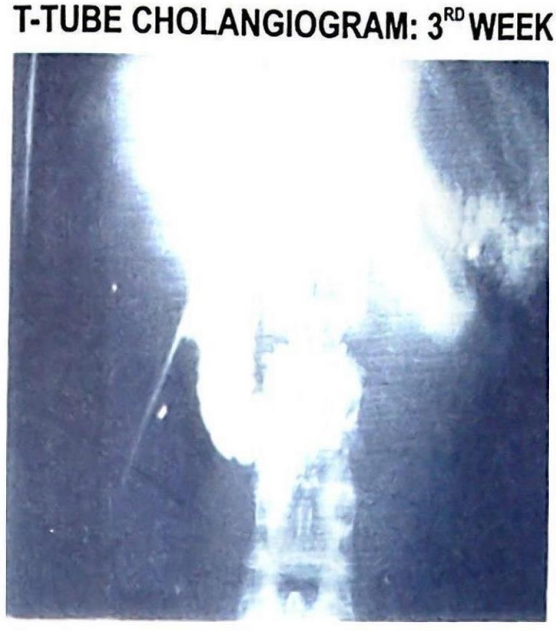

Fig. 1

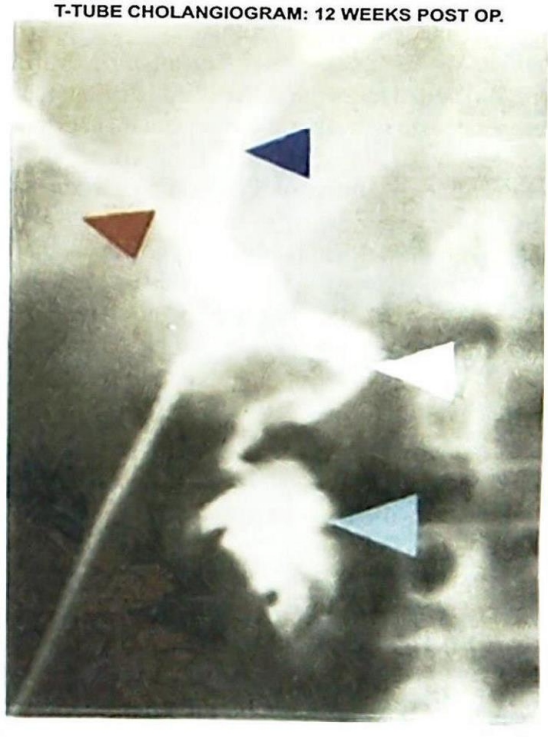

Fig. 2

fluoroscopy was unsuccessful because of faulty equipment. The repeat T-tube cholangiogram at 12th week post-operation showed contrast within the duodenal loop and no evidence of leakage (fig. 2). The right and left hepatic ducts were well outlined. The T-tube was removed thereafter. The patient has since returned to work with no abdominal pain or jaundice. Liver function tests have gradually returned to normal.

Discussion

Four different sub-types of this syndrome have been identified after the original description of this clinico-pathological entity by Mirizzi in 1948 [3,4]. Nagakawa et al suggested a modification but still retained the 4 sub-groups [5]. The basic pathology is the impaction of a big stone at the infundibulum of the GB or cystic duct resting on the CHD and causing mechanical obstruction to the flow of bile. This results in intermittent or unremitting jaundice. Perforation and a cholecysto-choledochal fistula may then ensue, thus further complicating the picture. This syndrome may be symptomless or presents with features of frank obstructive jaundice mimicking atimes malignant obstruction as seen in our patient [6]. This is a rare complication of gallstones seen in about 1:10,000 cholecystectomies in United States of America with an equal sex distribution. The incidence of 0.7% in our cholecystectomies is not different from that in the literature [2].

Our patient fell into type IIa. The diagnosis was made on the operating table after direct choledochotomy to remove the gallstone. The definitive pre-operative diagnosis of M/S rests on endoscopic retrograde cholangio-pancreatpgraphy (ERCP) or percutaneous trans-hepatic cholangiography (PTC) – two modalities of investigation unavailable to us [7]. Ultrasound has its limitations but is expected to demonstrate an impacted stone in the GB with dilatation of intra-hepatic biliary canaliculi and the proximal common hepatic duct above the GB neck. There is supposed to be an abrupt change in the caliber of the CHD below this level [8]. The USS of our patient was reported as showing a huge stone in the GB with normal wall thickness and normal-sized bile duct. Operative findings confirmed the huge stone but a shrunken GB communicating with the right side of a grossly dilated duct. The other ancillary investigations that have been found useful include abdominal computerized tomography (CT) scan with contrast, magnetic resonance cholangio-pancreatography (MRCP), position emission tomography (PET) scan and recently, diagnostic laparoscopy [9].

If the pre-operative diagnosis is difficult, the surgical operation can be a nightmare. Surgical options for M/S type I are fairly straight forward – a cholecystectomy which may be total or partial, depending on the technical difficulty encountered. However, for types II(a), (b) or (c), various surgical options have been attempted [10,11,12] but the aim is to remove the stone, do a choledochoplasty when possible, using the remnant of the amputated GB or re-route the bile flow through a bilio-enteric anastomosis. In our patients, a choledochotomy was performed to remove the stone and this was followed by a partial cholecystectomy. A choledoplasty was then performed over a T-tube, using the remnant of the GB. There was an initial leak but this resolved within 12 weeks post-operatively to allow us to remove the T-Tube (fig 2 – T-tube cholangiogram). The role of therapeutic laparoscopic surgery is controversial. Most authors consider the dense fibrosis in Calot's triangle too hazardous for laparoscopic techniques and certainly ill-advised when there is a cholecysto-choledochal fistula [13-16]. In our patient with a previous upper abdominal operation, the fibrosis is better imagined. Endoscopic naso-biliary catheter drainage in conjunction with cholangioscopy and eletrohydrolic lithotripsy has also been reported.

Mirizzi Syndrome (M/S) is a rare complication of gall stones and is difficult to diagnose pre-operatively. The various surgical options available reflect the complex nature of the management of this syndrome. Unfortunately, the ancillary aids that are required for pre-operative diagnosis to aid in proper planning for appropriate surgical procedure are not available to us. This rare complication of cholelithiasis is found in our practice with equal frequency as in the environment with a high incidence of cholelithiasis.

The surgeon is therefore advised to develop a high sense of suspicion about this syndrome so as not to be caught unawares.

References

1. O. O. Akute and M. O. Obajimi. Cholelithiasis in Ibadan: An update WAJM 2002; 21: 128-131.
2. Hunt DR: Another paper on Mirizzi Syndrome? ANZ J Surg. 2004; 74: 826.
3. McSherry CK, Fersten H and Virship M. The Mirizzi Syndrome: Suggested Classification and Surgical Therapy. Surg. Gastroenterol 1982; 1:219-225.

4. Csendes A, Diaz JC, Burdiles P., Malenda F and Nava O. Mirizzi Syndrome and Cholecystobiliary Fistula: A unifying classification. Br. J. Surg. 1989; 76: 1139-1143.
5. Nagakawa T., Ohta T., Kayahara M, *et al.* A new classification of Mirizzi Syndrome from diagnostic and therapeutic view points. Hepatogastroenteroloy 1997;44:63-67.
6. Freeman ME, Rose JL, Forsmark CE and Vauthey J. Mirizzi Syndrome: A rare cause of obstructive jaundice. Dig. Dis 1999;17:44-48.
7. Morrsey KP and McSherry CK, Internal Biliary Fistula and Gall Stone Ileus. In: Surgery of the liver and Biliary Tract. Edn 2, ch.64 917-920. Ed.L.H. Blumgart, Edinburgh: Churchill Livingstone 1994.
8. Becker CD, Hassler H and Terrier F: Pre-operative diagnosis of the Mirizzi Syndrome: Limitations of sonography and computed tomography. Am.J.Roentgenol 1984; 142:591-596.
9. Berland LL, Lawson TL and Stanley RJ. CT appearance of Mirizzi Syndrome. JComput Assit Tomogr. 1984:8:165-166.
10. Baer HU, Mathews JB and Schweizer WP. Management of the Mirizzi Syndrome and the Surgical Implications of Cholecystocholedochal fistula. Br.J.Surg. 1990;77:743-745.
11. Safioleas MC, Evangelidakis E, Xypolitas N and Liosis A. Mirizzi Syndrome. Medical Annals 1999;12(2):545-547
12. Safioleas M, Stamaatakos M, Revenos C *et al.* An alternative surgical approach to a difficult case of Mirizzi: A case report and a review of the literature. World J. Gastroenterol 2006; 12(34): 5579-5581
13. Posta CG Unexpected Mirizzi anatomy: A major hazard to the Common Bile Duct during laparoscopic cholecystectomy. Surg. Laparos. Endosc. 1995, 5(5): 412-414.
14. England RE and Martin DF: Endoscopic Management of Mirizzi Syndrome. Gut 1997; 40:272-276.
15. Paul MG, Burris DG, McGuire AM, *et al.* H. Laparoscopic Surgery in the treatment of Mirizzi Syndrome. J. Laparoscopic Surg. 1992; 2:157-163.
16. Kelly MD. Mirizzi Syndrome. ANZ J. Surg. 2005, 75:501-503.
17. Binnie NR, Nixon SJ and Palmer KR. Mirizzi Syndrome managed by endoscopic stenting and laparoscopic cholecystectomy. Br. J. Surg. 1992; 79(7):647.
18. Binmoeller KF, Thonke F and Soehendra N: Endoscopic treatment of Mirizzi Syndrome. Gastrointest, Endosc. 1993; 39: 532-536.

Received: 03/05/11
Accepted: 28/11/12