

Successful Management of a Cholecystectomy in Ehlers-Danlos Syndrome

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ABSTRACT

Background/Aims: Ehlers-Danlos syndrome (EDS) is a heritable connective tissue disorder that affect the skin, joints, blood vessels and internal organs. Patients with EDS are at a higher risk of experiencing different surgical complications depending on the type and severity of the disease. We describe here a case of a patient with EDS Type III who underwent a successful laparoscopic cholecystectomy.

Case report: A 42 year old woman was evaluated for a long lasting and recurrent epigastric pain with a negative Murphy sign. Magnetic Resonance Imaging detected multiple gallstones within the gallbladder without biliary tree dilatation. An uncomplicated laparoscopic cholecystectomy was performed.

Conclusion: Surgical intervention in patients with EDS is a challenging topic. It should be performed by a multidisciplinary team aware of its specific needs and risks. Taking into account these concerns and the type of EDS, we successfully carried out a laparoscopic cholecystectomy in our patient.

Key words: Ehlers-Danlos, cholecystectomy, multidisciplinary management

CASE REPORT

A 42 year old woman was evaluated for a long lasting and recurrent epigastric pain with a negative Murphy sign.

The patient had been operated on for appendicular peritonitis, obesity with adjustable gastric band and left anterior cruciate ligament injury.

Laboratory tests remained within normal limits. Radiological studies with abdominal ultrasound and Magnetic Resonance Imaging detected multiple calculi within the gallbladder without biliary tree dilatation (*fig. 1*).

In order to definitely relieve the pain and avoid potentially life-threatening complications such as acute cholecystitis, obstructive cholangitis and pancreatitis, we planned a laparoscopic cholecystectomy.

Our patient had a well-known Ehlers-Danlos syndrome (EDS), Type III, and was wearing Murphy rings (*fig. 2*) to correct joint hypermobility and instability. We proceeded to a multidisciplinary perioperative management. A senior anesthetic was present during the surgery.

Nurses were also aware of the situation and put padding in place to reduce shear forces and external tissue pressure (1).

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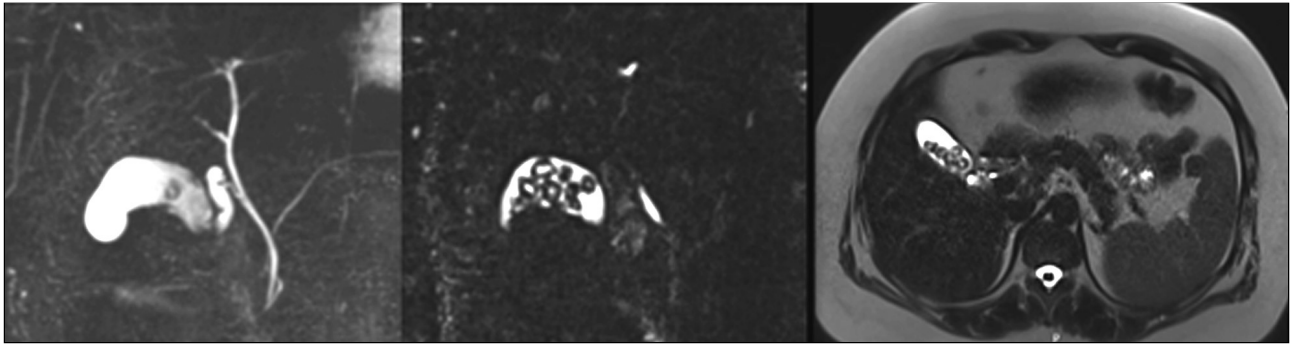


Figure 1 - Cholangio-IRM showing a gallbladder of normal size containing several gallstones



Figure 2 - Patient wearing Murphy rings. Cholecystectomy scars are visible without complications

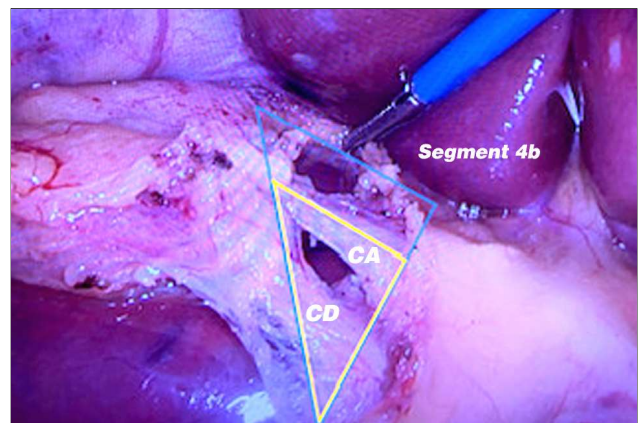


Figure 3 - Critical View of Safety (CVS)
In blue: hepatocystic triangle. In yellow: Calot's triangle
CA: cystic artery; CD: cystic duct

We created the pneumoperitoneum with an open laparoscopy instead of a Veress needle. Four trocars were inserted.

Following the concept of culture safety in cholecystectomy (2), we achieved the critical view of safety (3) after releasing peri vesicular adhesions. We correctly identified the two elements of the hepatocystic triangle: the cystic duct and cystic artery (*fig. 3*). As proposed by Crane (4), we used the Hem-o-lok® ligature system to adapt to the weakness and friability of the tissues (*fig. 4*). The cystic duct and artery were divided between Hem-o-lok® clips, and an uneventful retrograde cholecystectomy was performed with progressive electro dissection of the gallbladder bed.

After the extraction of the gallbladder, we did not notice biliary leak or hemorrhage. Consequently, no drain was left in place. Non absorbable sutures were used to close the wounds.

The patient recovered completely and was discharged to home on the first postoperative day. And, when seen twelve days after the surgery, she was doing well. The follow up at one year was unremarkable.

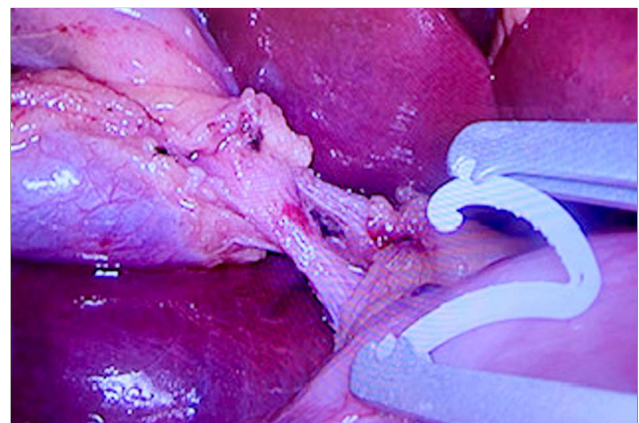


Figure 4 - Hem-o-lok® being placed

The histological studies showed a calculous cholecystitis.

DISCUSSION

Most EDS subtypes are caused by defective collagen type I, III and V (5). And, when the patients with EDS

need surgery, the poor quality of their connective tissue (1,6,7,8) may present some challenges.

The most common surgical issues are related to fragile blood vessels, wound healing, spontaneous rupture of hollow organs, joint instability and subluxation and hernias (1,5).

In our patient, to avoid the skin bruising, we used good padding as suggested by Weisemann (1). And, considering the risk of atlantoaxial dislocation and temporomandibular dysfunction, a senior anesthesiologist performed the endotracheal intubation without any incident (1).

For the creation of the pneumoperitoneum, we think that open laparoscopy was more safer than the use of Veress needle. It allowed us to prevent a possible iatrogenic intestinal lesion because of the history of appendicular peritonitis. And if ever a bleeding had occurred, it would have been easier to control it in open approach laparoscopy.

The case described by Crane (4) was a Type IV, the vascular type with arterial and hollow organs fragility or spontaneous rupture (5). That was the main reason for them to put a drain and left it in situ for several days. Our patient had a Type III, the hypermobile form with probably less vascular or hollow organs fragility. So, we decided not to place a drain.

The other aspect to be aware of is the poor healing capacity of our patient.

In 2000, seventeen years before the diagnosis of EDS, she had been operated on for left anterior cruciate ligament injury. Absorbable sutures were used which led to wound dehiscence.

Therefore, this time, we used ETHIBOND Excel® (green braided non-absorbable suture) for the closure of aponeurosis and fascia. And, the skin were closed with ETHILON®Nylon suture. No dehiscence was observed.

CONCLUSION

In patients with EDS, a multidisciplinary approach is advised with a team familiar with the disease and its complications. And, a laparoscopic cholecystectomy may be managed successfully by anticipating the common issues related to the connective tissue frailty.

To avoid an uncontrollable bleeding or intestine perforation, open laparoscopy should be preferred to Veress needle puncture approach.

Hem-o-lok ® might be well fit to control possible biliary leak or cystic artery bleeding.

A drain might be useful in type IV in order to monitor bleeding, biliary leak or hollow organs perforation.

Wound dehiscence occurs in patients with EDS and non absorbable sutures should be used to reduce the risk.

Conflict of interest: none.

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Ethical approval

Ethical approval was not required for this case report.

Acknowledgment

Published with written consent from the patient.

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