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A SUCCESSFUL LAPAROSCOPIC APPENDECTOMY FOR AN ADULT MALE PATIENT WITH OSTEOGENESIS IMPERFECTA

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ABSTRACT

Background: Osteogenesis imperfecta (OI) is a genetic connective tissue disease defined by the loss of bone mass and density, which makes the bones more brittle and more likely to fracture over time. Bone deformity and articular instability are the subsequent symptoms.

Case report: This 25-year-old man had malformed lower limbs and trouble walking due to OI and dwarfism. He arrived complaining of fever, nausea, vomiting and diffuse peri-umbilical pain. During ultrasonography a blinded, oedematous lobe formation containing an appendicolith was discovered. Acute suppurative appendicitis was diagnosed, necessitating a laparoscopic appendectomy. Because the patient had previously undergone general anaesthesia, anaesthesia was thought to be attainable. Pneumoperitoneum and a 10 mm optical port inserted into the umbilicus were used in the surgical procedure. A diagnostic laparoscopy revealed faecolith obstruction and an acute suppurative appendicitis. After an hour, a laparoscopic appendectomy was performed effectively with little blood loss. Without experiencing any difficulties because of the surgery position, the patient was discharged.

Conclusion: We present a case of an OI dwarf patient with acute suppurative appendicitis. It highlights the possibility of performing laparoscopic surgery in general and a laparoscopic appendectomy in particular on OI patients.

KEYWORDS

Osteogenesis imperfecta, laparoscopic surgery, laparoscopic appendectomy, perioperative difficulties

LEARNING POINTS

- In rare instances involving OI, laparoscopic surgery in general and laparoscopic appendectomy in particular are practical and efficient options.

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INTRODUCTION

Osteogenesis imperfecta (OI) is a genetic connective tissue disease defined by the loss of bone mass and density, which makes the bones more brittle and more likely to fracture over time. Bone deformity and articular instability are the subsequent symptoms[1]. Across all racial and ethnic groups, the incidence is 1:20,000. An autosomal recessive type has also been observed, while the majority of cases are inherited as an autosomal dominant condition[2].

CASE DESCRIPTION

Our patient was a 25-year-old male adult with dwarfism and OI. He is barely 80 cm tall and weighs 16 kg. Before reaching adolescence, he suffered from long bone fractures that required surgery to correct using metallic plates *(Fig. 1)*. He is the lone case in his family and there is no family history. The patient's lower limbs were deformed and shortened, and he was walking with difficulty.

He complained of diffuse peri-umbilical pain, which had spread to his right lower abdomen and was accompanied by fever, nausea and vomiting when he arrived at our emergency room. Upon examination, the right iliac fossa had tenderness, rebound and rigidity, and the patient had a positive cough test and psoas sign. He had a WBC of 10.7 × 10^3/l and a 72.6% neutrophil percentage. A long, oedematous, blinded lobe structure was seen by pelvic-abdominal ultrasonography. The lumen of the structure was around 8 mm, and an appendicolith was visible inside, surrounded by thickened omentum *(Fig. 2 and 3)*.

When the patient was admitted to our hospital, it was concluded that a laparoscopic appendectomy was required, since all results pointed to an acute suppurative appendicitis diagnosis. Although there was a risk of hypoventilation due to thoracic deformity and restricted lung capacity *(Fig. 4)*, the anaesthesia team judged that general anaesthesia was possible in this case as well, as there was a history of general anaesthesia in the past with this patient. Laparoscopic surgery was preferred and selected because of the advantages of minimally invasive surgery, and the superiority of diagnostic laparoscopy for complete abdominal exploration in identifying the precise location of the inflamed appendix and the true cause of severe abdominal pain. An acute suppurative appendicitis with faecolith blockage near its base was found during a diagnostic laparoscopy *(Fig. 5)*. A laparoscopic appendectomy was completed successfully, and one pelvic drain was inserted at the end of surgery before wound closure. There was minimal blood loss, and the procedure took about an hour in total. On the fourth postoperative day, the patient was discharged from the hospital without suffering any complications linked to the surgical position. Acute suppurative appendicitis with faecolith was the histological diagnosis that was consistent.

DISCUSSION

Osteogenesis imperfecta, one of the most common skeletal illnesses, is a hereditary condition characterised by increasing bone lesions and varying degrees of symptoms related to connective tissue and bone fragility. Type I collagen (COL1A1, COL1A2), a key component of connective tissue, is typically the source of gene mutations that cause it[3].

The two main methods used to diagnose OI are clinical and radiographic. The most defining feature of the clinical manifestations – which range from moderate forms to severe presentations – is bone fragility, deformity or limited development, and vulnerability to fracture from even minor stress. Hearing loss, blue sclera, dentinogenesis imperfecta and hypermobility of the joints are examples of extra skeletal symptoms. Since it is often difficult to place a case into a specific type of OI, classification based on severity and signs observed is still in use[4].

There are, to the best of our knowledge, just two published case reports of laparoscopic surgery for OI; the first involved

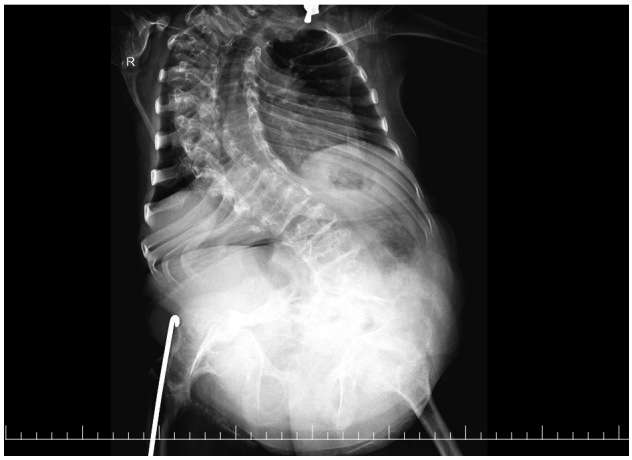

Figure 1. The patient suffered from long bone fractures that required surgery to correct, using metallic plates.

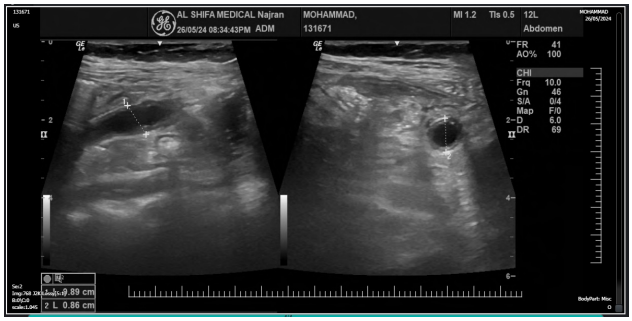

Figure 2. A long, oedematous, blinded lobe structure was seen by pelvic-abdominal ultrasonography.

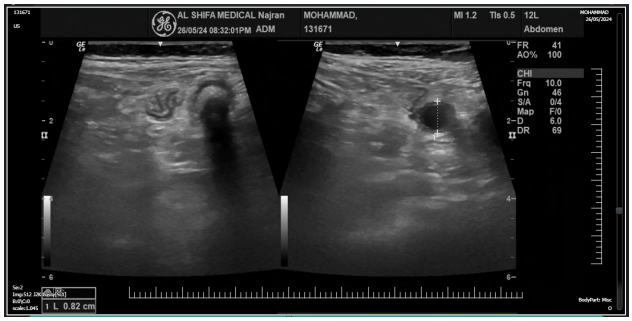

Figure 3. The lumen of the structure was around 8 mm and an appendicolith was visible inside, surrounded by thickened omentum.

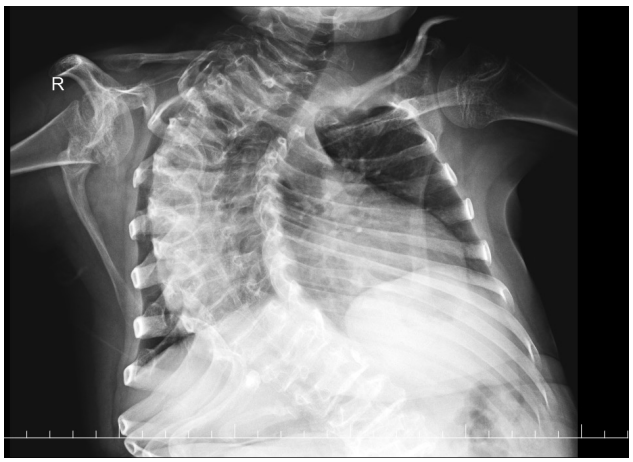

Figure 4. Thoracic deformity and restricted lung capacity.

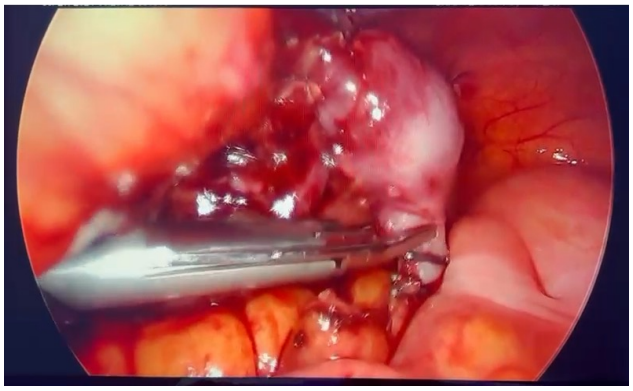

Figure 5. The appendicular faecolith and the narrow operative field due to low pneumoperitoneum pressure used.

the laparoscopic drainage of a tubo-ovarian abscess in a 33-year-old Japanese woman[5], and the second involved the laparoscopic sleeve gastrectomy of two teenage patients, one from the UK and the other from the USA[6]. We are presenting the first instance of an adult patient with OI who had an acute suppurative appendicitis that was successfully treated with a laparoscopic appendectomy.

When performing surgery on patients with OI, however, there were worries about a variety of intraoperative difficulties, thus the anaesthetic strategy, patient position and surgical technique needed to be taken into account[7].

The anaesthesia team judged that general anaesthesia was possible in this case as well, as there was a history of general as in the past with this patient. Regarding the surgical position, and due to the patient weight and height, the main difficulty was that we were dealing with what was in effect a 25-month-old infant instead of a 25-year-old adult patient, and the ease of fracture. So, we decided that there was no need to apply any restraint over the patient for fixation and to avoid putting the patient in the Trendelenburg position. In terms of the surgical technique, pneumoperitoneum was initiated, and a 10 mm optical port was placed into the umbilicus. Prior to inserting the remaining two 5 mm ports, pneumoperitoneum pressure was first performed at a lower pressure (10 mmHg). After that, we switched to an even lower insufflation pressure (8 mmHg).

We were able to successfully perform this surgery by carefully evaluating the anaesthetic technique, surgical position and surgical procedure. The procedure was completed without any special complications despite taking some time, and it was challenging to use the forceps in a supine posture and with a lower pneumoperitoneum pressure than usual.

CONCLUSION

We describe a case of acute suppurative appendicitis in patient known with OI that was treated with laparoscopic surgery. During the literature search, we were unable to locate any cases of laparoscopic surgery for appendectomy that were performed on patients who had OI. Consequently, we anticipate that surgeons who come across situations similar to this one in the future will find this report useful.

REFERENCES

1. Neri Morales C, Silva Amaro A, Cardona JD, Bendeck JL, Cifuentes Gaitan K, Ferrer Valencia V, et al. Osteogenesis imperfecta: a case series and literature review. *Cureus* 2023;**15**:e33864.
2. Edelu B, Ndu I, Asinobi I, Obu H, Adimora G. Osteogenesis imperfecta: a case report and review of literature. *Ann Med Health Sci Res* 2014;**4**:S1–5.
3. Valadares ER, Carneiro TB, Santos PM, Oliveira AC, Zabel B. What is new in genetics and osteogenesis imperfecta classification? *J Pediatr (Rio J)* 2014;**90**:536–541.
4. Uda T, Kudo M, Ohara Y, Yamazaki H, Ishizuka Y, Ihira K, et al. Laparoscopic surgery in osteogenesis imperfecta (OI) patient with tubo-ovarian abscess (TOA): a case report. *Japanese Journal of Gynecologic and Obstetric Endoscopy* 2018;**34**:128–133.
5. Zani A, Ford-Adams M, Ratcliff M, Bevan D, Inge TH, Desai A. Weight loss surgery improves quality of life in pediatric patients with osteogenesis imperfecta. *Surg Obes Relat Dis.* 2017;**13**:41–44.
6. Palomo T, Vilaça T, Lazaretti-Castro M. Osteogenesis imperfecta: diagnosis and treatment. *Curr Opin Endocrinol Diabetes Obes* 2017;**24**:381–388.
7. Masroor T, Chappell AG, Shahzad F. Anesthetic and surgical considerations in osteogenesis imperfecta: a case report of mandible fracture management. *Eplasty* 2022;**22**:e12.