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Large ovarian cystadenofibroma causing large bowel obstruction in a patient with Klippel–Feil syndrome—A case report

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ABSTRACT

INTRODUCTION: Ovarian cystadenofibromas (CAF) are epithelial tumors, which are fairly rare, mainly benign and asymptomatic. The Klippel–Feil syndrome (KFS) is a rare congenital anomaly which combines osseous and visceral development disorders. While bowel obstruction is a common complication in advanced ovarian cancer this condition is rarely described in cystic lesions such as CAF. We report the first case of large bowel obstruction due to a large benign ovarian CAF with an underlying KFS.
PRESENTATION OF CASE: A 60-year-old women with a KFS was admitted to the hospital with increasing abdominal girth and bowel obstruction. A CT scan revealed a massive intraabdominal cystic tumor of unclear origin, causing compression of the sigmoid colon with consecutive dilatation of the ascending colon and the small bowel. After successful conservative ileus treatment including nasogastric tube and iv fluids for a few days, we performed an explorative laparotomy, revealing a large cystic tumor originating from the left adnexe. After its removal including left adnexectomy, histological findings confirmed a benign cystadenofibroma. The hospital stay was uneventful and bowel obstruction symptoms resolved immediately.
DISCUSSION: Beside careful history taking and physical examination, diagnostic imaging is important to identify CAF. However, the distinction between benign and malignant lesions remains difficult even using MRI. Considering optimal preparation of the patient with successful ileus treatment, laparoscopy may have been the surgical approach of choice. Though, direct laparotomy seemed to be the appropriate approach, considering the unclear origin and dignity of the lesion. Considering the KFS and its inherent malformations interdisciplinary cooperation seems important in such rare cases.
CONCLUSION: Preoperative MR imaging and intraoperative frozen-sections can be helpful to determine the dignity of the neoplasm and avoid fertility impairing adnexectomy in young woman. Interdisciplinary management of such rare conditions seems of utmost importance.

1. Introduction

Bowel obstruction is a common complication in advanced ovarian cancer with a reported obstruction rate between 5–42% [1]. While ovarian teratomas [2] and endometriosis [3] can cause bowel obstruction too, this condition is rarely described in cystic lesions. We report the first case of bowel obstruction due to a large benign ovarian cystadenofibroma (CAF) with an underlying Klippel–Feil syndrome (KFS).

Ovarian cystadenofibromas (CAF) are epithelial tumors, which are fairly rare, mainly benign and asymptomatic [4,5]. It accounts for 1.7% of all benign ovarian neoplasms [6], occurring in women between 15 and 65 years [7]. Histological findings reveal fibrous stroma with an epithelial lining as the dominant components of the neoplasm.

The KFS is a rare congenital anomaly with an estimated incidence of 1/42,000 births. It combines osseous and visceral development disorders such as fusion of cervical vertebraes, low posterior hairline, brevicollis, basilar impression, atlanto-occipital fusion, scoliosis, facial asymmetry, torticollis, "Sprengel's deformity" and other genitourinary (e.g., kidney malposition) malformations, central nervous and cardiopulmonary anomalies. Defects of the notochord and its signalling are considered the main underlying cause [8].

Abbreviations: KFS, Klippel–Feil syndrome; CAF, ovarian cystadenofibromas.

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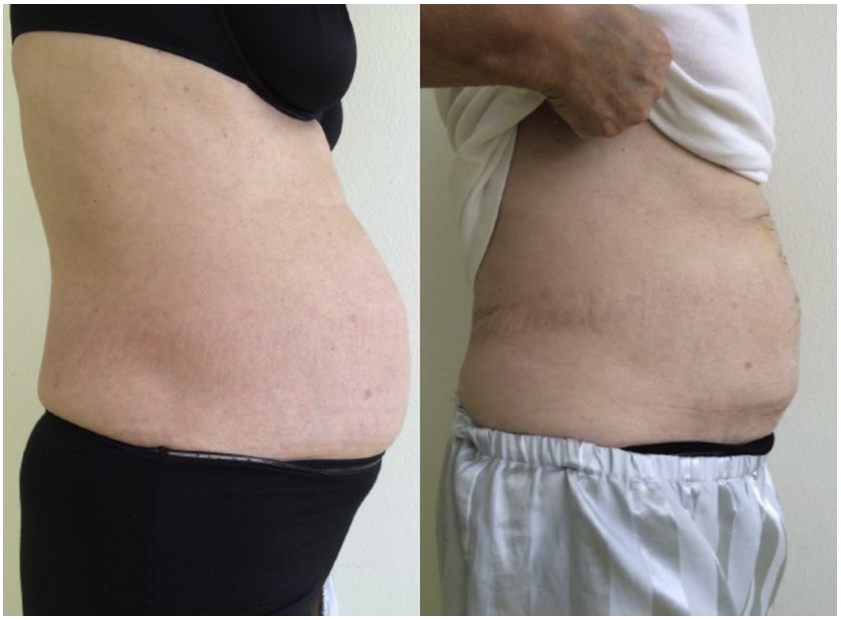

Fig. 1. Presented female patient. (a) Preoperative; (b) postoperative.

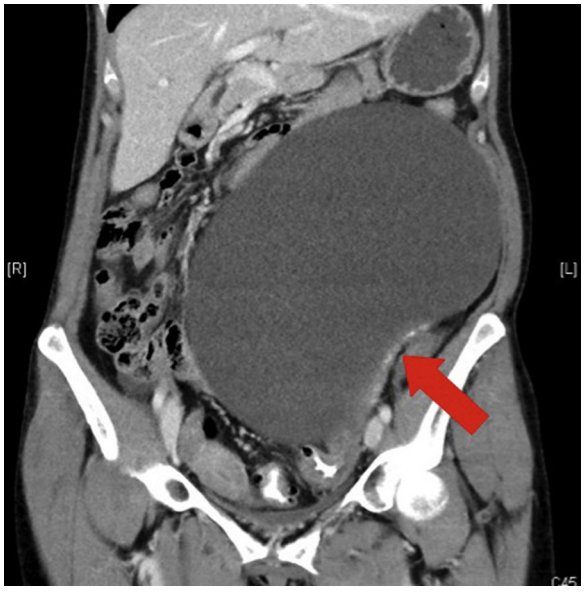

Fig. 2. Compression of the descending colon (red arrow).

2. Presentation of case

A 60-year-old, postmenopausal female patient with a KFS and an associated left pelvic kidney admitted herself to the emergency department with increasing abdominal girth (Fig. 1a) over a period of the past 4 weeks. She complained about alterations of her bowel habits with diarrhea alternating with constipation, pencil-shaped stools and episodes of perianal fresh blood discharge. A colonoscopy 3 years ago was unremarkable, especially no signs of obstruction of inflammatory processes were identified. A recent gynecological examination few months ago revealed no abnormalities.

On clinical examination the abdomen was heavily bloated without tenderness or pain. A median laparotomy scar due to explorative laparotomy 20 years ago did not show any conspicous features. A large resistance on the left lower abdomen was palpable and bowel sounds were dull. Digital rectal examination was unremarkable. All blood test including tumor markers were normal.

A CT scan showed a large left sided intraabdominal cystic tumor with unclear origin and dignity, causing subtotal compression of the descending colon (Fig. 2). On the admission day, the patient developed increasing abdominal discomfort with symptoms of bowel obstruction including nausea and vomiting. A decompressive therapy was initiated with iv fluids and insertion of a nasogastric tube, draining 4 l of reflux in two days. Cardiology specialists cleared the patient for surgery concerning her underlying KFS. Subsequently we performed an explorative laparotomy three days after hospital admission.

Intraoperative findings revealed a massive $20 \times 10 \times 20$ cm thin-walled cyst and an additional smaller cyst measuring 4 cm originating from the left adnex. The mass almost completely occupied the abdominal cavity with displacement of the small bowel and parts of the colon in the right upper abdomen and total entrapment of the sigmoid colon between the cystic lesion and the pelvis. The absence of adhesions of the cystic mass to any abdominal organs suggested a benign character of the tumor. Large bowel loops were distended and the rectum was collapsed. The small bowel was only slightly distended due to the preoperatively performed decompressive therapy. No other obstructing reason was found. During extraction from the abdomen, the cyst showed a restiform origin from the left adnex, directly distal to the left ovary (Fig. 3). Both cysts were resected at their restiform origin using ligations. However, due to the unclear dignity of the cystic mass and the unusual extent of the mass a left total adnexectomy was performed. The intraoperative and postoperative course was uneventful. The patient completely recovered and preoperative bowel symptoms resolved immediately, with return to normal abdominal girth (Fig. 1b). the patient was discharged 5 days after surgery. A follow-up 6 weeks after surgery was unremarkable. Pathological examination revealed a cyst covered by flat to cuboidal epithelium (Fig. 4a), with partially thickened cyst wall (maximum diameter 12 mm, see Fig. 4b and c). A sex cord stromal tumor was excluded by negative immunohistochemical staining for alpha-Inhibin. Taking all findings together, the diagnosis of a benign cystadenomfibroma (CAF) was made. This case report has been reported in line with the CARE criteria [9].

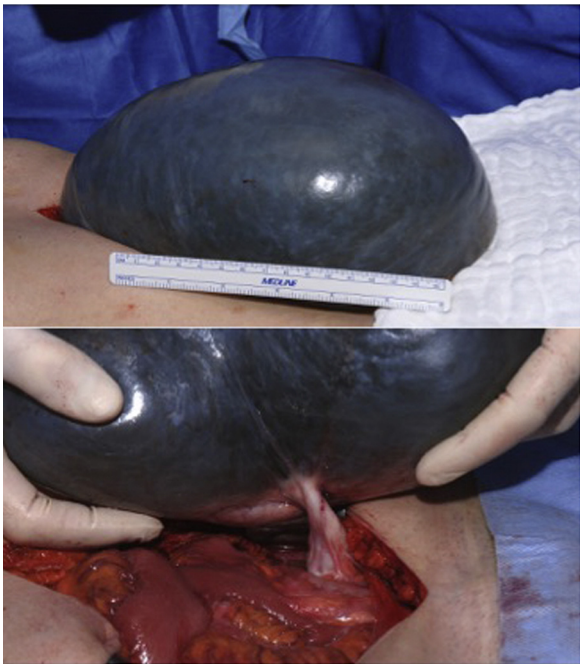

Fig. 3. Intraoperative Situs.

3. Discussion

To our knowledge this is the first reported case of a large ovarian cystadenofibroma (CAF) causing bowel obstruction in a patient with a KFS. Their coincidental occurrence is remarkable.

Beside careful history taking and physical examination, diagnostic imaging is important to identify the lesion. Several authors suggest MRI to be the method of choice for diagnosis of multiloculated cystic mass with solid fibrous component such as cystadenofibromas (low-signal intensity on T2-weighted sequences and a "black sponge" aspect) [4]. Nevertheless, the distinction between benign, borderline or malignant variation seems to remain difficult even in MRI scans due to the solid portions, which might mimic malignant neoplasm [2]. However, due to the emerging patients symptoms of bowel obstruction and underlying KFS we performed an abdominal CT scan, which identified the cystic mass and the nearly complete sigmoid obstruction (Fig. 2). Considering the KFS and its inherent malformations careful interdisciplinary patient evaluation excluding concomitant disorders (e.g., ventricular septal defect) [10,11] seems of utmost importance.

Although some authors performed laparoscopy for cystic ovarian lesions [12] direct laparotomy seemed to be the appropriate approach for this size of mass with unclear origin and dignity, minimizing the risk of intraoperative ruptur and spillage during laparoscopy. Interdisciplinary cooperation seems important in such rare cases, which we accomplished by consulting cardiological and gynaecological experts before and during surgery. We abstained from frozen-sections as suggested by some authors [7] because it would not have changed the extent of the resection in our postmenopausal patient. Accordingly, left sided adnexectomy was subsequently conducted.

4. Conclusion

Due to unclear origin and dignity of this space consuming cystic mass causing large bowel obstruction, we consider explorative laparotomy as the surgical approach of choice. It allows safe resection of the mass with careful exploration of the abdomen. In younger, fertile woman preoperative MRI imaging may alternatively be used for imaging diagnostics. Intraoperative frozen-sections can be considered in younger woman to avoid

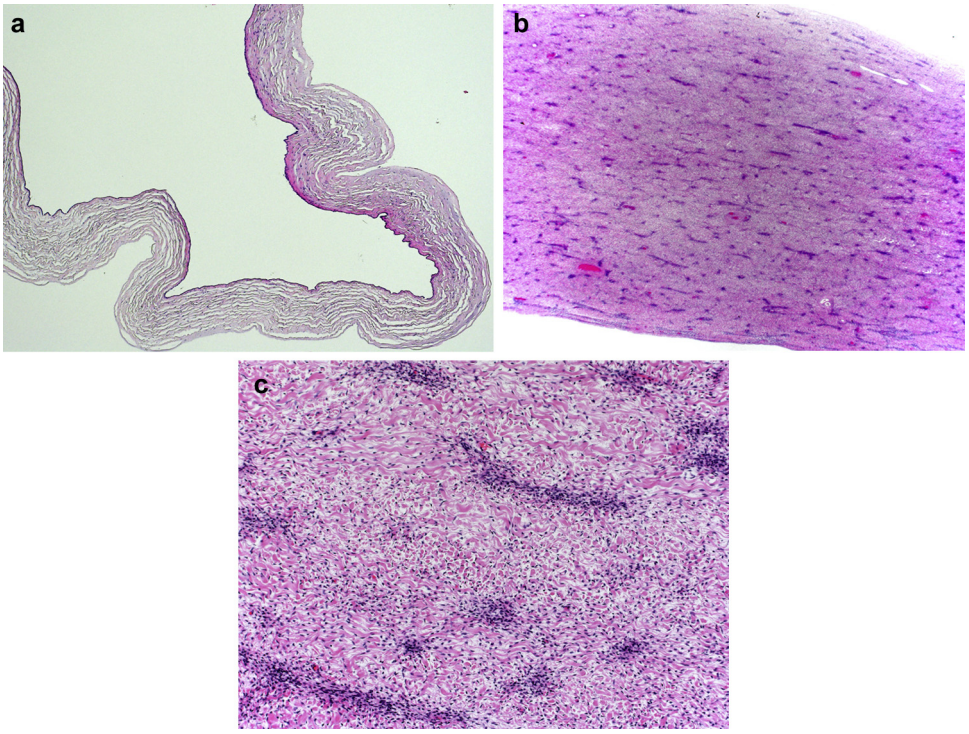

Fig. 4. Pathological examination. (a) Part of the thin cystic wall lined by flattended epithelium. No mucinous epithelium or cellular atypias are seen (HE-staining, 40×). (b) Overview of the thickened cyst wall with stroma cells grouping around vessels (HE-staining, 12.5×). (c) Detailed view of the thickened wall. No smooth muscle cells or elements of a stromal sex cord-stromal tumor as well a atypia of the stromal cells can be detected (HE-staining, 100×).

extensive fertility impairing surgery. Interdisciplinary management seems mandatory in these patients and the role of the pathologist is crucial to determine the dignity and origin of this tumour.

Conflicts of interest

The authors declare that there is no conflict of interest.

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Ethical approval

No ethical approval necessary for a case report. The patient's consent was obtained.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Author contribution

Robert Mechera carried out the writing of the manuscript. Thomas Menter was the treating pathologist and added the pathological images and information. Daniel Oertli was reviewing and correcting the manuscript. Henry Hoffmann supported Robert Mechera during the writing process and designed the final artwork. All authors have approved the final article.

Guarantor

Robert Mechera.

References

[1] S.J. Mooney, M. Winner, D.L. Hershman, J.D. Wright, D.L. Feingold, J.D. Allendorf, et al., Bowel obstruction in elderly ovarian cancer patients: a population-based study, Gynecol. Oncol. 129 (April (1)) (2013) 107–112.
[2] R. Al-Harfoushi, E.-H. Abdulaziz, S.I.H. Andrabi, B. Patterson, M. Whiteside, Ovarian teratoma presenting as small bowel obstruction in an elderly lady—a case report, Int. J. Surg. Case Rep. 2 (January (1)) (2011) 6–8.
[3] S. Lanitis, M. Korontzi, C. Karaliotas, Acute bowel obstruction in a premenopausal woman, Gastroenterology 144 (January (1)) (2013) e5–e6.
[4] P.-F. Montoriol, A. Mons, D. Da Ines, N. Bourdel, L. Tixier, J.M. Garcier, Fibrous tumours of the ovary: aetiologies and MRI features, Clin. Radiol. 12 (December (68)) (2013) 1276–1283.
[5] T.S. de Silva, A. Patil, R.N. Lawrence, Acute presentation of a benign cystadenofibroma of the fallopian tube: a case report, J. Med. Case Rep. 4 (January) (2010) 181.
[6] S.-M. Cho, J.Y. Byun, S.E. Rha, S.E. Jung, G.S. Park, B.K. Kim, et al., CT and MRI findings of cystadenofibromas of the ovary, Eur. Radiol. 14 (May (5)) (2004) 798–804.
[7] D.H. Lee, A case of mucinous cystadenofibroma of the ovary, Case Rep. Obstet. Gynecol. 2014 (January) (2014) 130530 (Fig. 1).
[8] K.M. Kaplan, J.M. Spivak, J.A. Bendo, Embryology of the spine and associated congenital abnormalities, Spine J. 5 (5) (2005) 564–576.
[9] The CARE Guidelines, available from: http://www.care-statement.org/ (accessed 21.12.15).
[10] M.R. Tracy, J.P. Dormans, K. Kusumi, Klippel–Feil syndrome: clinical features and current understanding of etiology, Clin. Orthop. Relat. Res. (July (424)) (2004) 183–190.
[11] W.B. Moore, T.J. Matthews, R. Rabinowitz, Genitourinary anomalies associated with Klippel–Feil syndrome, J. Bone Joint Surg. Am. 57 (April (3)) (1975) 355–357.
[12] E.S. Sills, C.R. Kaplan, M. Perloe, M.J. Tucker, Laparoscopic approach to an uncommon adnexal neoplasm associated with infertility: serous cystadenofibroma of the fallopian tube, J. Am. Assoc. Gynecol. Laparosc. 10 (November (4)) (2003) 545–547.