CASE REPORT

Exceptionally Rare Krukenberg Tumour in a Patient With Dextrocardia and Situs Inversus Totalis

Vickneswaren Thever Ramasamy, Yong Chee Meng

Gynae-Oncology Unit, Department of Obstetrics & Gynaecology, Ampang Hospital, Jalan Mewah Utara, Taman Pandan Mewah 68000 Ampang Jaya, Selangor, Malaysia

ABSTRACT

Krukenberg tumour is a rare clinical entity and accounts for about 1-2% of all ovarian tumours. It is a metastatic ovarian tumour in which accurate diagnosis requires proper endoscopic evaluation and histopathological confirmation to exclude primary ovarian tumours. Dextrocardia with situs inversus totalis is another unusual clinical entity, with an incidence of 1 in 10,000 of the general population. Many patients with this unusual condition are unaware of their structural abnormality and mostly are diagnosed incidentally when they present with other medical conditions. Herein, we report a case of 47 years old woman who presented with bilateral ovarian tumour and incidental diagnosis of Dextrocardia with situs inversus totalis. She underwent surgery and histopathological examination confirms Krukenberg tumour. Both conditions existing together, an exceptionally rare clinical entity.

Keywords: Krukenberg tumour, Dextrocardia, Situs inversus totalis

Corresponding Author:
Vickneswaren Thever Ramasamy, MMed
Email: vickneswaren6074@gmail.com
Tel: +60342896000, +60122191375

INTRODUCTION

In clinical practise diagnosis of Krukenberg tumour has always been a challenge. Being a metastatic ovarian tumour, most patients would undergo extensive evaluation to determine the primary site of the tumour only when there is clinical suspicion. Unfortunately, at times this condition is only known following surgery done for suspected primary ovarian tumour. This make things extremely difficult, to treat a patient when the primary site is still unknown.

Apart from this, the clinical challenge becomes even more crucial when there is presence of a rare anatomical abnormality such as Dextrocardia and situs inversus totalis. Therefore a holistic approach in terms of endoscopic evaluation, radiological assessment, histological confirmation and most importantly a good surgical anatomical knowledge are very much required to make an accurate diagnosis and to plan the treatment promptly.

CASE REPORT

47 years Para 3 with no previous medical illness presented with gradual abdominal distension over a year which was painless. There were no compressive symptoms or constitutional symptoms. Upon clinical assessment the mass was suspected to be a huge pelvic mass with tumour makers CA125 was marginally elevated and normal CEA levels. She had a CT Thorax, Abdomen and Pelvis done which revealed bilateral multiloculated ovarian mass with thick septation suspicious of malignancy (Fig. 1), multiple uterine fibroid, right iliac fossa colo-colic intussception with bowel mass as leading point and also Dextrocardia with situs inversus totalis. Patient underwent a colonoscopic assessment which showed a polypoidal tumour (Fig. 2) at the descending colon (65 cm from the anal verge), tumour was prolapsing distally causing intussusception, the stalk of the tumour identified and tattooed with Indian ink: unfortunately the procedure was abandoned due to pain.

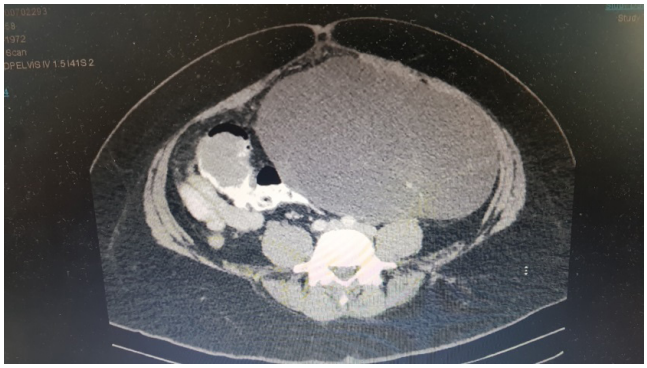

Figure 1: Adnexal mass with colonic mass on CT

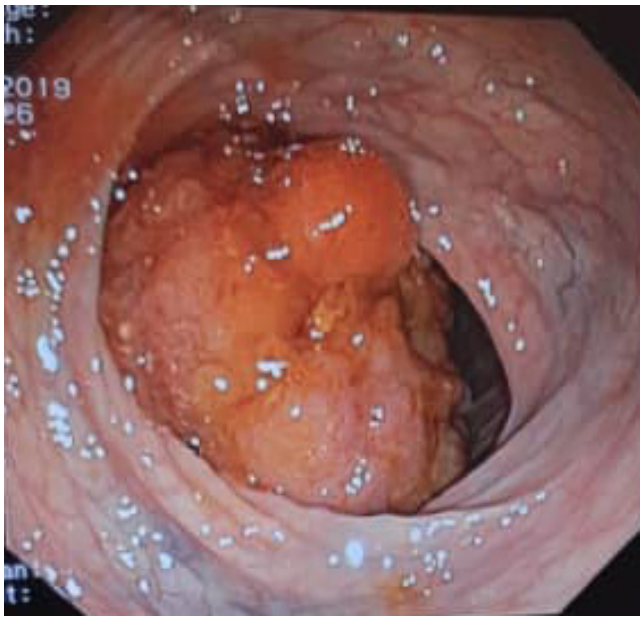

Figure 2: Colonoscopic view of the polypoidal growth at the sigmoid colon

Thereafter she underwent a Laparotomy with Total Abdominal Hysterectomy, Bilateral Salpingoopherectomy (Fig. 3), sigmoid colectomy (Fig. 4) with end to end anastomosis, Omentectomy, Left pelvic lymph node dissection, Appendicectomy and Upper abdomen peritonectomy. She recovered well following surgery and her histopathological examination showed Adenocarcinoma with mucinous component of the Sigmoid colon with metastasis to bilateral Ovaries, anterior abdominal wall/peritoneum and Pouch of Douglas. She was diagnosed to have Krukenberg tumour. Patient underwent another colonoscopic examination following surgery which showed no recurrence of disease at anastomotic site (25cm from anal verge). At present she has been subjected for chemotherapy by the Oncology team.

DISCUSSION

Dextrocardia with situs inversus is characterized by abnormal position of the heart as well as the internal organs (1). Situs inversus totalis is a unique phenomenon in which there is reversal of the organs in the chest and abdominal cavity in a mirror image pattern (1). Situs

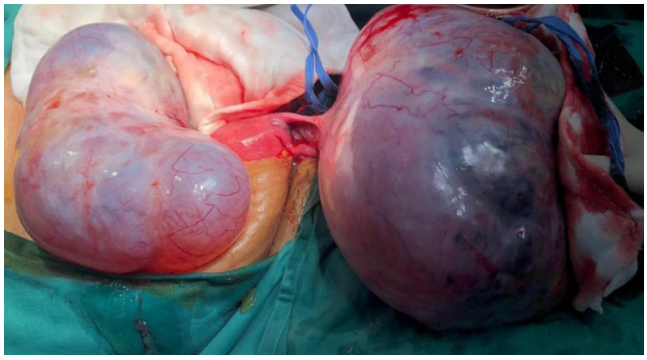

Figure 3: Bilateral ovarian tumour

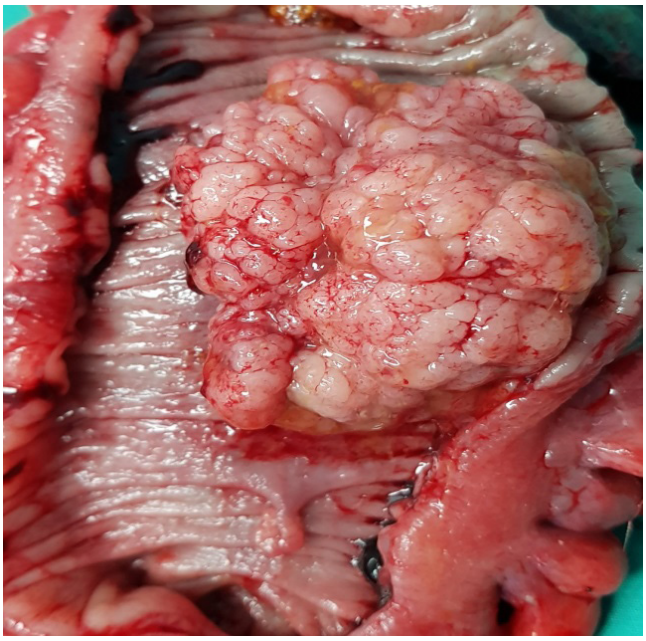

Figure 4: Primary tumour of the sigmoid colon

inversus is often seen with other congenital abnormalities affecting the spleen, kidneys, lungs and the vascular system as well (2). Situs inversus totalis coexisting with primary ciliary dyskinesia is called as Kartagener syndrome (2). There are some cases of situs inversus totalis with various types of cancers and surgeons had difficulty during surgery due to the abnormal anatomy. However there is no etiological relationship between situs inversus totalis and cancer reported before.

Clinically when there are presences of any ovarian metastatic lesions, Krukenberg tumour should be suspected and the usual site of primary malignancy is often from the gastrointestinal tract (3). Most patients are asymptomatic or having mild and nonspecific symptoms in early stages. But in later stages more specific symptoms appear as the size of the tumour becomes larger. For the diagnosis of Krukenberg tumour a histopathological confirmation is mandatory, which includes the presence of stromal involvement, mucin producing neoplastic signet ring cells and ovarian stromal sarcomatoid proliferation (4). Common sites of primary are stomach and colon, other sites includes; breast, biliary system and appendix. In most cases the ovarian involvement is bilateral; however unilateral involvement is not uncommon as well. These metastatic ovarian tumours are generally solid and well defined. Presences of bilateral solid cystic ovarian masses are not specific for Krukenberg tumour as many ovarian primaries have this morphology. Three possible pathway of metastases have been identified; lymphatics, haematogenous and transcoelomic metastases (5). It has been postulated that functioning ovaries which are rich in blood supply are prone to hematogenous metastases; the incidence is observed more in premenopausal than postmenopausal women (5).

In many instance ovarian mass is the first sign that appears before identifying the primary tumour. Therefore when Krukenberg tumour is suspected, apart from the routine imaging for investigating an ovarian tumour, an endoscopic evaluation of the gastrointestinal tract is very important to look for presence of primary tumour. Occasionally the primary tumour is never discovered which worsens the prognosis.

No optimum treatment plan has been recommended for treating Krukenberg tumour. It is important to determine the primary tumour in order to plan the appropriate treatment. Krukenberg tumour has a poor prognosis with average life expectancy ranging from 14 months to 28 months. In recent years, combination of surgery and chemotherapy has led to improvement in prognosis but it still remains poor. The assistance of palliative care is very much required in managing patient with Krukenberg tumour.

Dextrocardia with situs inversus totalis is a clinical challenge both diagnostic and surgically, due to the anatomical variation faced by the surgeons and makes it difficult in orientation of the organs. Familiarity with this anatomical variation would be an advantage however being a rare condition some surgeons might not have seen such variations before, therefore the important key point is for surgeons to spend a little more time to identify the location of important surgical landmarks for the proper surgical orientation prior to any intervention and confirmation by another fellow surgeon would be ideal. Meanwhile, the surgical technique did not differ from the usual manner. As for diagnosis is concern in terms of anatomical variation, being confusing at first but however with clinical awareness and combination of clinical with radiological approach diagnosis would be feasible.

CONCLUSION

These conditions are rare entity and with limited literature available. Therefore it is crucial that surgeons and radiologist to be aware of such anomaly pre operatively. Proper endoscopic evaluation is warranted pre operatively and histological confirmation is very important to exclude ovarian primary.

ACKNOWLEDGEMENTS

Special thanks to all clinicians and nursing staffs involved in the management of this patient.

REFERENCES

1. Yilmaz, S.; Demirtas, A.; Tokpinar, A. & Acer, N. Dextrocardia and situs inversus totalis in a turkish subject: a case report. Int. J. Morphol., 2019;37(3):900-902.
2. Kumar A, Singh MK, Yadav N. Dextrocardia and asplenia in situs inversus totalis in a baby: a case report. J Med Case Rep. 2014;8:408
3. Muthukrishnan S, Naganathbabu OL, Murugesan SD, Srinivasan UP, Amudhan A, Rajendran S. Krukenberg tumours from gastrointestinal cancers—analysis from a tertiary care centre in India. J Gastrointest Oncol 2018;9(6):1164-1167.
4. Moore RG, Chung M, Granai CO, Gajewski W, Steinhoff MM. Incidence of metastasis to the ovaries from nongenital tract primary tumours. Gynecol Oncol. 2004;93: 87-91
5. Kubeček O, Laco J, pačekJ, Petera J, Kopecky J, Kubečkovб A, Filip S. The pathogenesis, diagnosis, and management of metastatic tumors to the ovary: a comprehensive review. Clin. Exp. Metastasis 2017;34:295-307.