CASE REPORT

Appendiceal and rectal carcinomas with Krukenberg tumour mimicking primary ovarian cancer

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Abstract

Krukenberg tumour is an uncommon metastatic tumour of ovary. It accounts for 1-2% of ovarian malignancies. Pylorus of stomach is the most common site of primary tumour. We are presenting two case reports of patients who had Krukenberg ovarian masses. Case I; an appendiceal adenocarcinoma (AACa) which manifested clinically as primary ovarian cancer. Staging laparotomy revealed large right side ovarian tumour of clinical FIGO Stage III. Histological examination revealed a poorly differentiated adenocarcinoma which involved right ovary and appendix. Immunophenotypic analysis revealed positive expression of Cytokeratin (CK) 20 and CDX2 which was compatible with appendiceal primary and ovarian metastases. Case 2; 60 years female with right adnexal and rectal masses. Histology revealed a moderately differentiated adenocarcinoma of rectum, CK7 was negative. CK20 and CDX2 were positive. Large bowel was the primary tumour site. Distinction between ovarian, rectal and appendiceal primary malignancies is critical as the treatment modalities vary.

Keywords: Krukenberg Tumour, Ovarian Tumours, Metastatic Tumours.

Introduction

Krukenberg tumour is an uncommon metastatic adenocarcinoma of ovary.[1,2] They commonly occur in age group between 30-40 years and are rare after menopause. A large number of cases (80%) have bilateral ovarian involvement.[3,4] It accounts for 30%-40% of those tumours which secondarily metastasize to the ovary. Metastasis is from gastric adenocarcinoma, most commonly at pylorus 70% followed by 10% bowel,4% breast, 3% biliary system, 3% appendix and remaining from lungs, pancreas, bladder, renal pelvis and rarely 3% cervix.[1,4] Radiological evaluation may mimic other metastatic or primary ovarian tumours thus leading to difficulty in diagnosis.[1] We present two case reports where primary site of tumour was gastrointestinal tract and appendix with secondary metastatic involvement of ovaries hence classified as Krukenberg tumour.

Appendiceal malignancies are rare and they are diagnosed in only 0.9-1.4% of appendicectomy specimens.[5] It occasionally demonstrates ovarian metastases that are large and which dominate clinical and radiological presentations, leading to a misdiagnosis of an ovarian primary malignancy. The prognosis is poor, because it is usually found at an advanced stage due to a low threshold of suspicion and difficulties in diagnosis prior to surgery.[6]

Case-1

A 41 years old lady, parity four presented in emergency in November 2013 with generalized abdominal pain, vomiting and an abdominal mass for two months. Abdominal examination revealed a single, firm mass around 10x12cms with irregular boundaries below the umbilicus. Abdomino-pelvic ultrasound and Computed Tomography (CT) scan revealed complex solid cum cystic right adnexal mass around 13x8.6x12.6cms. No obvious appendiceal mass was evident (Figure).

She had elevated CA125 i.e. 39.8U/ml. Based on the clinical presentation, physical examination, tumour marker and radiological findings, a clinical diagnosis of an ovarian malignancy was made. Staging laparotomy revealed right large ovarian tumour size 13x10cm, frozen sections of which revealed an epithelial tumour. A 2x3cms mass was identified at base of appendix and clinical FIGO Stage III was assigned. Peritoneal fluid cytology revealed no malignant cells. Total abdominal hysterectomy, bilateral salpingoophorectomy, omentectomy, appendicectomy and right hemicolectomy at ileotransverse colon junction with end to end anastomosis were performed.

Histology report revealed right ovarian mass, morphological and immunohistochemical features favour metastatic adenocarcinoma compatible with appendix as primary site. Single focus of tumour in appendix around 4x2.5 cms present. Moderately to poorly differentiated adenocarcinoma in appendix, ascending colon and half transverse colon. Tumour was invading through muscularis propria into subserosal adipose tissues with

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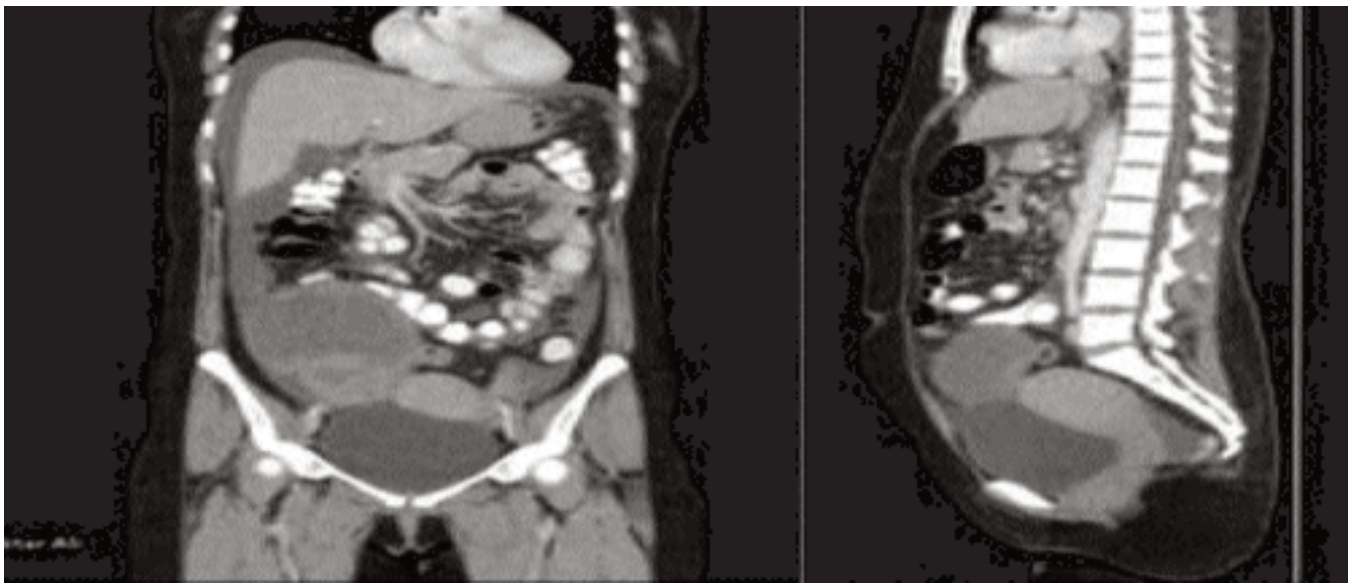

Figure: CT Scan images complex solid cum cystic right adnexal mass around 13x8.6x12.6cms.

peritoneal extension and pelvic lymph node involvement. Endometrium, myometrium and cervix were unremarkable. Left ovary was involved by metastatic tumour. Left fallopian tube was normal. Omentum was tumour free. Pathological TNM (Tumour, Lymph Nodes and Metastasis) stage was T3, N2, M1. Immunohistochemical staining with CK7 and CK20 were positive. Patient is on follow-up for the past eighteen months with no recurrence.

Case-2

A 60 years old lady, parity three presented in April 2016 in emergency with complaint of per rectal bleeding, abdominal mass and abdominal pain for one year. Abdominal examination revealed a single firm mass with irregular boundaries around 12x12cms arising from lower abdomen. Abdominopelvic ultrasound, CT scan and Magnetic Resonance Imaging (MRI) revealed a large heterogenous soft tissue mass in right adnexa 15.1x11.8x10 cms with internal septa and mild ascites. Mural soft tissue thickening of rectosigmoid area was also noted which was approximately 10.2cms.

She had elevated CEA 136ng/ml and normal CA125 i.e. 8.30U/ml. Ascitic fluid cytology was negative for malignancy. Based on the clinical presentation, physical examination, tumour marker and radiographic studies, a clinical diagnosis of secondary ovarian and primary rectal malignancy was made. Exploratory laparotomy, diverting colostomy, total abdominal hysterectomy, and bilateral salpingoophorectomy, removal of right ovarian mass and left DJ stenting was performed. Clinical FIGO Stage III was assigned. Peritoneal fluid cytology revealed no malignant cells. No peritoneal, omental and liver metastasis were found. Histology revealed a moderately differentiated adenocarcinoma of rectum, CK7 was negative and CK20 and CDX2 were positive. Immunohistochemical profile showed a large bowel as primary tumour site which was in favour of metastasis from gastrointestinal tract rather than primary ovarian tumour. Uterus and both fallopian tubes were normal. Patient is on follow-up for twenty four months with no recurrence.

Discussion

Paget in 1854 first discovered Krukenberg tumour. Krukenberg tumour is an uncommon metastatic signet ring cell tumour of ovary that originates primarily in the stomach.[1] They contribute only 30%-40% i.e. much less than metastases from other ovarian cancers. They commonly present with symptoms of ascites, bloating pelvic pain and sometimes with menstrual irregularities .Only 20-30% have prior history of stomach or colon cancer.[7] Our cases had unusual presentation with ascites at 40 to 60 years without any symptoms to direct us in its early detection.

Appendiceal malignancies are rare and they are diagnosed in only 0.9 to 1.4% of appendicectomy specimens.[5] Ovarian metastases are encountered in 16.7 to 37 % of AACa.[8] The analysis of CK7 and CK20 is useful in such cases.[6] The ovarian carcinomas are CK7 positive /CK20 negative and the AACa as are CK7 negative /CK20 positive. CDX2 is a sensitive marker for colorectal carcinoma which metastasizes to the ovary. The

differential diagnosis is clinically important, since the therapeutic approach is totally different for ovarian and appendiceal cancers. An optimal surgical debulking is advocated for ovarian tumours and chemotherapy with taxol and carboplatin is effective whereas the clinical utility of radical tumour debulking and chemotherapy remains unknown for appendiceal cancers.[5,8] However, an aggressive cytoreductive surgery and a perioperative intraperitoneal chemotherapy with Mitomycin-C and 5-fluorouracil could be used for appendiceal cancers with peritoneal disseminations.[5] The appendix should always be carefully examined during explorations for ovarian masses and prophylactic appendicectomy should be considered as a part of the treatment of ovarian carcinomas.

Gross and microscopic findings can differentiate between Krukenberg tumours and other primary tumours of ovaries. Both chemotherapy and radiotherapy have no significant role in management.[1]

Conclusion

Krukenberg tumour is a rare clinical entity. It is essential to rule out other ovarian malignancies to avoid misdiagnosis and management of the Krukenberg tumour.[9] Serum CA-125 level can help to predict the prognosis.

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