An unusual presentation of Krukenberg tumor in a young female patient: A case report

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ABSTRACT

Krukenberg tumor is an uncommon metastatic tumor of the ovary. It usually presents in female with an average age of 45 years and constitutes 1-2% of all ovarian neoplasms. 30% of all ovarian neoplasms occurring during childhood and adolescence are malignant. A 22-year-old unmarried lady presented with painless abdominal distension with amenorrhea. On examination, 18-week lump was palpable. This lump was firm to hard in consistency, non-tender, and mobile. On ultrasonography, bilateral ovarian tumors were reported, with peritoneal free fluid. Laparotomy with right salpingo-oophorectomy and enucleation of left ovarian mass was performed. Microscopic examination revealed bilateral signet ring cells carcinoma of the ovaries probably metastatic origin with intact capsule. Tumor cells exhibited strong, diffuse immunopositivity for cytokeratin (CK) 7 and CK 20. Diagnosis of Krukenberg tumor was made. Repeated endoscopic biopsy confirmed the diagnosis of signet ring cell carcinoma stomach. This case is reported because of its rarity in younger age group, diagnostic dilemma, and poor prognosis.

Key words: *Cytokeratin 7, Cytokeratin 20, Immunohistochemistry, Krukenberg, Signet ring cell carcinoma*

Ovarian carcinoma is the fourth most common malignant disease of women. Metastatic cancer to ovary accounts for 1-2% of all ovarian cancers [1,2]. In 1896, Fredrick Krukenberg described a new type of ovarian tumor. This tumor was later identified as a malignancy in the ovary from a primary lesion in the gastrointestinal (GI) tract, and named "Krukenberg tumor" after him. Krukenberg tumors refer to a malignancy in the ovary that metastasizes from a primary site, classically the GI tract and breast [3]. Krukenberg tumors are often (over 80%) found in both ovaries, consistent with their metastatic nature. Differentiating primary from metastatic ovarian carcinoma is often challenging. Treatment of patients with Krukenberg tumor is controversial. Until now, optimal treatment has not been established. We are reporting a case of Krukenberg tumor in a 22-year-old lady.

CASE REPORT

A 22-year-old unmarried lady presented to our hospital with a 2-month history of painless, progressive, and swelling in abdomen which was not an associated history of trauma. There was no history of contact with tuberculosis. Her menstruation history was normal. Blood examination showed mild anemia with a hemoglobin level of 10.5 g/dl and an erythrocyte sedimentation rate of 80 mm. Her tumor markers were measured, and the level of serum CA-125, serum carcinoembryonic antigen (CEA), and lactate dehydrogenase were 593.9 units/m, 5.41 μg/L, and 248 IU/L, respectively, whereas S-alpha-fetoprotein and β-chorionic gonadotropin were within normal limits. HIV serology was negative. Chest X-ray was normal and pelvic ultrasound demonstrated a heterogeneous pelvic mass in pelvis with moderate ascites. Magnetic resonance image pelvis revealed a large heterogenous solid cystic lesion 20 cm × 11 cm × 8 cm in pelvis extending above umbilicus probably arising from the right ovary along with a small cystic lesion in the left ovary with massive ascites (Fig. 1). The initial diagnosis of ovarian carcinoma was made, and laparotomy with right salpingo-oophorectomy and enucleation of left ovarian mass was performed. Histopathological examination of specimen revealed diffuse proliferation signet ring cells in ovarian stroma with features suggestive of signet ring cell carcinoma of both ovaries with intact capsule probably metastatic origin (Fig. 2). Immunohistochemistry (IHC) revealed diffuse positive of cytokeratin 7 (CK7) and CK20. The rest of the peritoneal cavity was completely unremarkable. Ascites for malignant cell was negative. A diagnosis of bilateral ovarian Krukenberg tumor was made. Postoperatively, upper GI endoscopy and colonoscopy were carried out and revealed features of linitis plastica of stomach. Biopsy from the stomach was inconclusive.

Computed tomography scan of the thorax and abdomen was done for postoperative evaluation, which shows diffuse thickening in the fundus and body of the stomach along with septated collection in bilateral adnexa and pouch of Douglas with mild ascites and right hydronephrosis. In view of histopathological report and imaging, gastric biopsy was repeated and it was also inconclusive. The case was discussed in tumor board and discussed for all possible treatment options and was decided to plan for deeper biopsy. Third gastric biopsy revealed signet ring cell carcinoma of stomach (Fig. 3). After proper explanation and counseling, possibly thinking it to be metastatic disease with poor outcome, she was offered palliative chemotherapy, but the patient party decided not to take any active oncological management. After 4 months of diagnosis, the patient came to us with feature cachexia, massive ascites, right hydronephrosis, and uremia. At this time only, palliative treatment was offered. The patient was discharged on request.

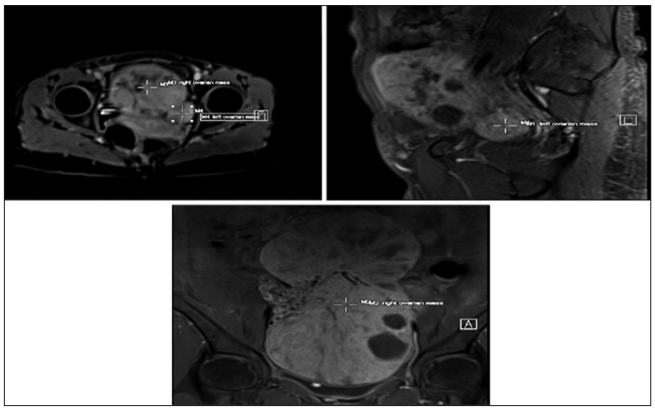

Figure 1: Contrast magnetic resonance imaging of pelvis showing large heterogeneously enhancing predominantly solid lesion in pelvis extending to the abdomen with a smaller lesion in left adnexa

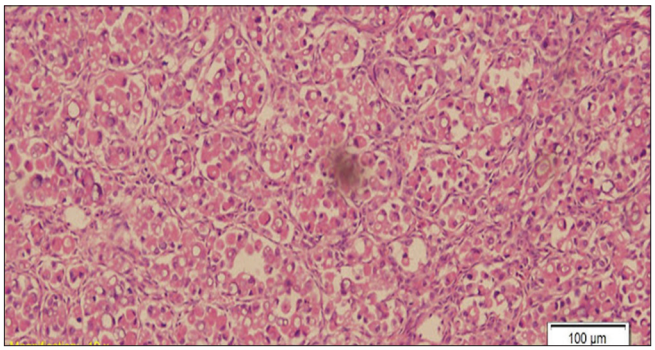

Figure 2: Sections studied form bilateral ovarian tumor shows sheet-like diffuse proliferation of signet ring cells in ovarian stroma with features of signet ring cell adenocarcinoma

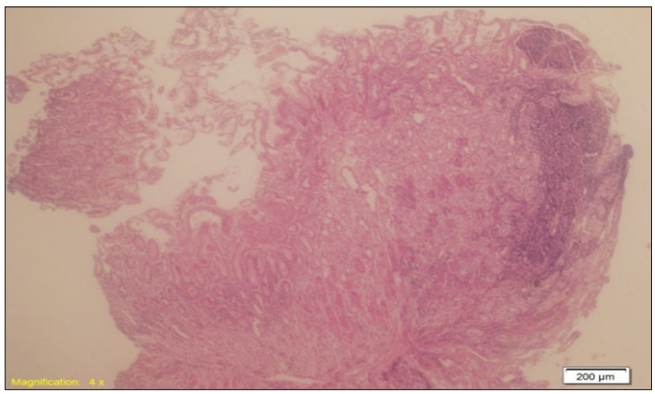

Figure 3: Tumor involving stomach showing signet ring cells with hyperchromatic nuclei ×40 magnification

DISCUSSION

Ovarian carcinoma is the fourth most common malignant disease of women. Types of ovarian carcinoma, including serous, mucinous, endometrioid, and transitional carcinoma, differ from each other with respect to morphology, genetic alterations, and in their clinical course.

Krukenberg tumor is a metastatic signet ring cell adenocarcinoma of the ovary, accounting for 1-2% of all ovarian tumors. Stomach is the common primary site in Krukenberg tumors (70%). Carcinomas of colon, appendix, and breast (mainly invasive lobular carcinoma) are the next most common primary sites [1]. Review of literature indicates that 35-45% of the patients reported were <40 years, with an average range of 40-46 years. It is rarely seen in younger age group and only a few such cases have been reported. Gupta et al. and Khurana et al. reported Krukenberg tumor in women aged 20 and 13 years, respectively [4,5]. Majority of cases are synchronous, but 20-30% occur as metachronous lesion after the removal of primary.

Our patient was originally a case of signet ring cell carcinoma of stomach with synchronous Krukenberg tumor. Here, patients underwent the removal of ovarian lesion without the knowledge of having a gastric lesion. After the histopathology had come as signet ring cells, she was further investigated to have signet ring cell gastric carcinoma after repeated biopsy.

Distinguishing between primary and metastatic tumor is fundamental to the patient's management and can be differentiated on the basis of clinical features, morphological, and pathological findings. The presence of signet ring cells is one of the most important morphological features of metastatic mucinous carcinoma of ovary, which are rare in primary ovarian mucinous tumors [6]. The features favoring secondary mucinous carcinomas are surface tumor deposits, nodular growth pattern, and lymphovascular permeation. The features more in favor of primary carcinoma of ovary are unilateral tumor, lower tumor staging, and background of adenofibroma or cystadenoma. The IHC may help in diagnosing Krukenberg tumor from primary ovarian neoplasm but needs to be applied with discretion [7,8]. The tumors that are immunoreactive for CEA or CK20 and negative for CK7 are more likely to be of colorectal in origin. However, the tumors that are immunoreactive to CK7 and CK20 are more likely to be of gastropancreatobiliary in origin. CK7 and CK20 are usually not reactive in primary carcinoma [9]. Special AT-rich sequence-binding protein 2 is a nuclear matrix-associated transcription factor, expression of which is restricted to glandular lining of the lower GI tract [10].

In the present case, the tumor cells were immunoreactive to CK7 and CK20. These findings of immunochemistry favor and further strengthen the diagnosis of metastatic signet ring cell carcinoma of ovary from gastropancreatobilliary system. The route of spread of this tumor is still not well established. As the tumor is usually well encapsulated and rarely shows any ovarian surface involvement, theory of peritoneal seeding from primary lesion is questioned. Rich lymphatics draining gastric mucosa and submucosa initiating retrograde lymphatic spread to ovary are mostly accepted theory.

In addition, ascites is a common presentation in the Krukenberg tumor and usually reveals malignant cells [1]. However, there has been a reported case of bilateral Krukenberg tumors with benign ascites and right hydrothorax that revealed no malignant cells. This is known as pseudo-Meigs syndrome, in contrast to the Meigs syndrome, which presents with a triad of benign ovarian tumor, ascites, and right-sided hydrothorax [11]. In our case also, ascites is present but does not reveal any malignant cell. The rarity of this case is due to the rarity of gastric cancer in young women. Indeed, only 0.4-0.5% of gastric cancers occur in women aged <30 years [12].

The prognosis of a patient with Krukenberg tumor is extremely poor with average survival time between 3 and 10 months. Only 10% of patients survive more than 2 years after diagnosis [13]. There is no specific guideline for treating Krukenberg tumor, but existent literature favors operative removal of Krukenberg tumor along with primary tumor if there is no other dissemination.

CONCLUSION

Krukenberg tumors are very rare in younger age group. Their poor prognosis emphasizes the importance of early diagnosis and treatment. Awareness of the diagnostic manifestations of the tumor leads to the correct diagnosis and prevents tumor misclassification, thus avoiding improper clinical management.

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