

**Case report****“PRIMARY SIGNET RING CARCINOMA OVARY-A CASE REPORT”**Tayal U<sup>1</sup>, Bajpai M<sup>2</sup>, Jain A<sup>3</sup>

1. Professor, Department of Pathology, Rama Medical College, Hospital and Research Center, Ghaziabad
2. Associate Professor, Department of Pathology, Rama Medical College, Hospital and Research Center, Ghaziabad
3. Assistant Professor, Department of Pathology, Rama Medical College, Hospital and Research Center, Ghaziabad

**Abstract:** Primary signet ring carcinoma ovary is an extremely rare tumor reported in the literature. We report a case of primary bilateral signet ring carcinoma in a 26 years old female. Krukenberg tumour usually is a secondary tumor of ovary originating from the stomach, gall bladder biliary tree, pancreas, appendix or colorectal organs. In the past few years there have been 13 cases reported as primary ovarian tumor with signet ring cells. In our case patient was presented with bilateral ovarian mass with the right ovary measuring 20x15 cm and left ovary 4x 2.5 cm. The patient was operated for ovarian tumor after detailed investigations in the month of April 2012. On last reporting to Rama Medical College Hospital and Research Center, Ghaziabad, in the month December 2012. The patient was recovering under the follow up treatment. **Conclusion:** Prognosis for Primary signet ring carcinoma of ovary is generally poor. It is important that correct identification and staging of the hidden primary is not delayed as timely treatment will definitely benefit the patient.

**Key words:** Adenocarcinoma, CK 20, Krukenberg tumor, primary signet ring, tumour antigens CA - 125.

**Introduction:** Krukenberg was the first to describe a signet ring cell carcinoma of the ovary<sup>[1]</sup>. Till date, 13 cases have been reported in the literature as primary signet ring tumor of ovary<sup>[2,3]</sup> While, most of these were benign, 3 cases were primary mucinous signet ring cell carcinomas of the ovary<sup>[4]</sup>.

In general, primary signet ring carcinoma of the ovary is an extremely rare tumor<sup>[5]</sup>. However, other cases of primary signet ring tumour have been reported<sup>[6]</sup>. It has been reported that SRCC (Signet ring cell carcinoma) were seen in younger patients. Differentiating primary from a secondary

krukenberg tumor is a diagnostic dilemma. The patients presenting signs and symptoms, clinical behavior and diagnostic evaluation are required to identify the primary source of the lesion.

**Case report:** A 26 year old female presenting with fullness of lower abdomen; difficulty in breathing and sudden pain of abdomen since 6 months was admitted in Rama Medical College Hospital and Research Center. On routine investigations, the patient was found to be anemic. Ultrasonography showed a bilateral ovarian mass with right ovary measuring 20x15 cm and left ovary 4x2.5cm along with bilateral

pleural effusion and ascitis. 2.5 lit of pleural fluid was aspirated. Pleural fluid as well as ascitic fluid was found positive for malignant cells. On further studies, cancer antigen-125 (CA-125) was found to be markedly raised (615 IU/L) but carcinoembryonic antigen (CEA) was normal. Cytokeratin 7 (CK 7) was found to be positive and Cytokeratin 20 (CK 20) was negative. The clinical presentation, radiological, laboratory reports and clinical diagnosis of a primary ovarian malignant tumour with metastasis was considered. The patient underwent an extended total hysterectomy. Both clinical and radiological findings indicated no gastrointestinal tract, pancreas, breast and biliary tree infections.

The ascitic fluid and pleural effusion regressed totally after the surgery. A repeat X-ray and ultrasound found no pleural effusion or ascitis after 2 weeks of surgery. General condition of the patient was improved on discharge after two weeks of hospital stay.

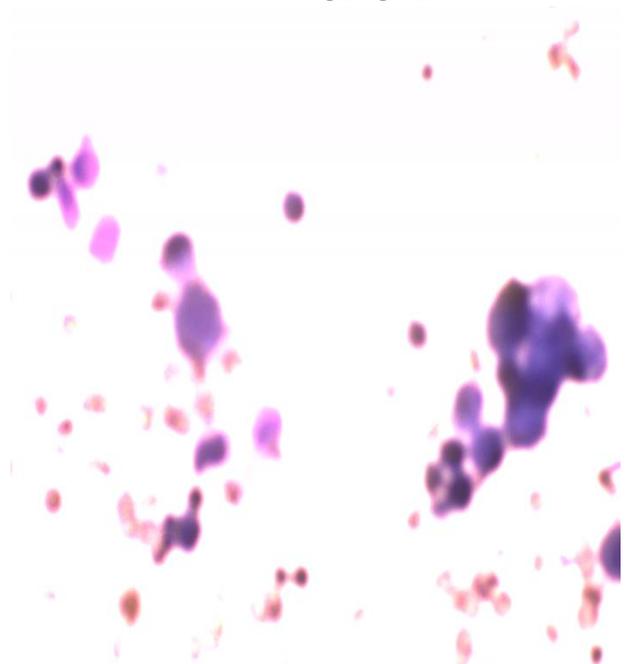
**Gross Examination:** The specimen of uterus, ovary, and bilateral adnexal structures consist peritoneal fold, right and left pelvic lymph nodes and pre and para aortic lymphnodes were received in the pathology department[Fig-1].



**Fig 1: Specimen of Uterus**

The gross examination revealed a bilateral ovarian tumour. Right ovary tumour mass, measured 15x10x8 cm and weighed about 1.5 Kg. Left ovary mass measured 3.5x3 cm. Both ovaries showed a smooth glistening, bosselated appearance. The cut surface was lobulated, firm, grey white with 2-3 cystic areas at the periphery. The uterus and cervix were normal in appearance. All lymph nodes were sampled and measured 0.5 to 1 cm in size with smooth and homogenous cut surface.

**Microscopic examination:** Sections from right as well as left ovary tumour mass showed signet ring cells in groups with peripheral crescentic nuclei. Signet cells were also seen infiltrating the stroma. Microemboli of the tumour cells were seen in peritoneal lymph node, left fallopian tube and peritoneum. Cytoplasm of these tumour cells varied from granular eosinophilic to pale vacuolated appearance and was found positive for PAS and Mucicarmine staining[Fig-2].



**Fig 2: Microscopic examination**

With these histopathological findings, diagnosis of bilateral primary signet ring cell carcinoma of ovary was confirmed. Radiographic and endoscopic exploration of the digestive system did not reveal any primary tumour; in thorough investigation to locate the hidden tumour. After the total hysterectomy patient was relieved of pleural effusion and repeat X-Ray after two weeks showed no pleural fluid after 2 weeks.

**Discussion:** Primary signet ring carcinoma ovary is a rare. Diagnosis is responsible for the most frequent diagnostic confusion with secondary ovarian tumor. Krukenberg is a metastatic signet ring cell adenocarcinoma of the ovary, accounting for 1-2% of all ovarian tumour. The interval between the diagnosis of primary carcinoma and the subsequent finding of ovarian involvement is usually 6 months or less, but longer periods have been also reported. A history of a prior carcinoma of stomach or any other organ can be obtained in only 20% to 30% of the cases<sup>[7]</sup>. In many cases the primary tumour is very small and can escape detection. In the present case meticulous radiographic and endoscopic investigation did not reveal any primary carcinoma.

Distinction from other primary ovarian tumour is of great importance as classification of secondary Krukenberg tumor as a primary ovarian tumor may lead to suboptimal treatment of the patient. Major tumour in this group is sertoli-leydig cell tumour particularly if intracellular mucin is not evident on routine staining. Differentiation can be made on the basis of their characteristic histologic features<sup>[8]</sup>. Due awareness of the primary signet

ring carcinoma ovary and careful clinical history can help to minimize the errors.

In our patients the preoperative serum CA-125 level in the patient was increased to a high level and subsequently decreased after tumors resection. However CEA was normal. Results of CK20 and CK7 also indicated towards ovarian origin of the carcinoma. The finding of our case matched with result of the other authors<sup>[9,10,11]</sup>. Immunohistochemical study in such cases is said to be of limited value because of overlapping of immunophenotype among primary ovarian signet ring carcinoma and secondary from stomach, pancreas, biliary tree, appendix, or colorectum, the most common primary sites. In the present case, patient has no symptoms of GIT.

**Conclusion:** Prognosis is generally poor, it is vital to differentiate these tumours from secondary ovarian tumours. It is also important that correct identification and staging of a hidden primary is not delayed as timely treatment will definitely benefit the patient. In the present case extensive search for the primary was done radiologically, serologically, Immunologically, including CEA and CA125, but no primary foci was detected. Hence the case is considered to be a Primary signet ring Krukenberg carcinoma ovary.

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**Address for correspondence:**

Dr. Uma Tayal  
Professor, Department of Pathology,  
Rama medical college, Hospital & research center, Ghaziabad  
Email: umatayal25@gmail.com