Small cell carcinoma of bilateral ovary-pulmonary type: A Rare Case Report

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Abstract: Small cell carcinoma of ovary is a rare relatively aggressive tumor of ovary. Here we present a case report of 60 year old postmenopausal women. Microscopically, the tumor showed solid growth of small cells arranged in sheets and closely packed nests separated by a fibrous stroma. The tumor cells had hyperchromatic nuclei with inconspicuous nucleoli and scanty cytoplasm. Immunohistochemical staining showed positivity for synaptophysin, chromogranin and cytokeratin. The immunohistochemical features confirmed the diagnosis of pulmonary type of primary small cell carcinoma of the ovary.

Keywords: Immuno-histochemistry(IHC), ovary, pulmonary type, small cell carcinoma.

INTRODUCTION

Small cell carcinoma of ovary was first described in details by Dickersin and Scully in the 1980s.¹ It mostly occurs in young women and adolescents, with a mean age of 23 yrs. (range 1 - 46) at diagnosis. About 50% of patients present with an advanced stage of disease.² Small cell carcinomas of the ovary are of two types. The hypercalcemic type is most common, which is usually unilateral and present with variable IHC findings. The second is Pulmonary type which is similar to the homonymous lung tumor which can be pure or associated with endometrioid carcinoma or other patterns.³ ⁴ On IHC, there is reactivity for keratin, neuron specific enolase (NSE), EMA and rarely for chromogranin and Leu 7.⁵ Prognosis is very poor because of frequent extraovarian spread.⁶

CASE REPORT

A 60 year old female who presented with fullness and pain in abdomen since 1 year. CT scan and ultrasound was done and it showed large pelvic abdominal mass measures 20x14cm. It showed multiple septations, solid and cystic areas, areas of calcification with no lesion in any other organ including lung. The impression was of carcinoma ovary. Clinically, laboratory investigations did not revealed hypercalcemia. Ca 125 was 52.5 u/ml (normal is <35 u/ml). Total abdominal hysterectomy was done and sent for histopathological examination. Grossly, both ovaries were approximately of same size measuring 14.5x10x 8 cm with outer surface ruptured. Cut surface showed soft solid tumor with areas of necrosis and hemorrhage (Figure 1).

Figure 1: gross photograph showing solid tumor with some areas of necrosis and hemorrhage
Microscopically, there was solid growth of small cells arranged in sheets and closely packed nests with insular arrangement separated by a fibrous stroma. (Figure 2) The tumor cells were pleomorphic, small (round to oval) and with scanty cytoplasm. Nuclei were hyperchromatic with inconspicuous nucleoli (Figure 3). The capsule of the ovaries showed infiltration by tumor cells. No lymphocytic infiltrate or granulomas were seen in the stroma. IHC for cytokeratin, synaptophysin, chromogranin, neuron specific enolase, CD 20, calretinin, MIB-1 and mic 2 was done. The results showed diffuse positivity for cytokeratin, synaptophysin, chromogranin and Neuron specific enolase whereas CD 20 and calretinin were negative. Over 80% of the tumor cells showed strong reactivity for MIB-1. Hence, histopathology and IHC confirmed the diagnosis as bilateral ovarian tumor with features of small cell carcinoma-pulmonary type.

**Figure 2:** Small cells arranged in sheets and closely packed nests with insular arrangement separated by a fibrous stroma (H&E, x10)

**Figure 3:** H&E, 40x, pleomorphic tumor cells with scanty cytoplasm.
DISCUSSION

SCCO is a very rare, malignant, undifferentiated neoplasm with a very poor prognosis. The cell lineage remains not clearly defined.\(^8\)\(^9\) It occurs in young females and is nearly always bilateral.\(^10\) The differential diagnosis includes sex cord tumors, dysgerminoma, ovarian metastasis of lymphoma and melanoma.\(^11\) Out of the two types hypercalcemic type and pulmonary type of small cell carcinoma of ovary, pulmonary type is still more rare, very rarely published in the literature.\(^12\) Eichhorn J et al\(^6\) reported a series of 11 primary small cell carcinoma of ovary. They did not classified the tumors into pulmonary type and hypercalcemic type. Out of these 11 cases, 5 were bilateral and 5 cases were combined with other tumors of ovary. On IHC, 7 cases showed NSE positivity and 6 cases were positive for keratin. A case of pulmonary type of small cell carcinoma, reported by Mebis J et al in a 54 year old lady, was in combination with endometrioid adenocarcinoma of the left ovary and brenner tumor in the right ovary.\(^13\) The present case showed no combination with other ovarian tumors and presented with short history without hypercalcemia or metastasis.

CONCLUSION

Small cell carcinoma of ovary is rare, aggressive tumor with poor prognosis. A limited number of published studies are available, mainly of hypercalcemic type which usually occurs as a unilateral tumor only in the ovary and in young women. Pulmonary type of small cell carcinoma ovary is more rare and has to be differentiated from metastasis of small cell carcinoma of the lung mainly by clinical history and imaging findings. The present case is a pulmonary type of small cell carcinoma which was confirmed by microscopy and IHC. There was no significant history nor any features of lung tumor in the same patient.

REFERENCES