NEUROENDOCRINE CARCINOMA OF OVARY IN ASSOCIATION WITH MUCINOUS CYSTADENOMA

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ABSTRACT

A case of Neuro-endocrine carcinoma of ovary has been discussed. This thirty years old patient presented with mass in lower abdomen which was diagnosed to be an ovarian tumour .She had laparotomy and biopsy report was neuroendocrine carcinoma. She had two courses of chemotherapy and then did not come for follow up.

Key Words: *Ovarian Cancer, Neuro-Endocrine Carcinoma, Non-Small Cell Variant.*

INTRODUCTION

Neuro endocrine tumours are a heterogeneous group of separate clinicopathological entities, having a common characteristic i.e expression of endocrine differentiation potential.[1]

Non-small cell neuroendocrine carcinoma of ovary is a rare aggressive neoplasm, characteristically arising in association with a surface epithelial tumour.[2] It is an extremely rare ovarian tumour; only ten cases have been reported in literature so far. All these cases were associated with a surface epithelial stromal component.[3]

We report a case of Neuro-endocrine carcinoma of ovary in a thirty years old patient.

CASE HISTORY

A thirty-year-old lady presented to Gynae out patient dept with history of pain and mass in the lower abdomen for the last four months, which had worsened for the past two weeks. Her urinary and bowel habits were normal. She had lactational amenorrhoea and had no previous history of menstrual irregularity. She had delivered four children normally without any complications. Her last baby was delivered fourteen months ago.

On physical examination, she was normotensive, afebrile, of medium built weighing 70 kg, with no lymphadenopathy.

On abdominal examination there was a mass in lower abdomen corresponding to 20 weeks gestation. It was non-tender and fixed. There was no clinical evidence of other visceral involvement and ascites.

On vaginal examination, cervix was regular; a fixed non-mobile mass was felt in the pelvis corresponding to 20 weeks gestation. Uterus was not felt separately from the mass. There was fullness in both fornices.

Sonography revealed a large complex mass in pelvis 7.3 - 5.3 cm, with the probability of tubo-ovarian mass. There was evidence of mild ascites. The rest of abdominal and pelvic viscera were normal with no lymphadenopathy.

Ca-125 was reported as 137U/ml. Renal and liver function tests were normal. Exploratory laparotomy was planned.

Operative findings were haemorrhagic ascitic fluid, about 10 to 12cm multilobulated solid cystic, right ovarian, fixed mass, with adhesions to gut. Right tube, left ovary and tube were normal. Uterus was of normal size, but with a friable surface. The surface of liver was smooth, with normal pelvic and para-aortic lymph nodes. There was no evidence of involvement of gut and omentum. The ovarian mass was removed followed by total abdominal hysterectomy and removal of both adnexa and omentectomy. Cut section of the ovarian mass showed solid, cystic irregular areas with chocolate colored fluid. Postoperative period was uneventful and patient was discharged on 6th postoperative day.

The results of histology were reported as:- “Histological and immunohistochemical features favour Neuroendocrine carcinoma of non-small cell type”. The sections were stained with a panel of

monoclonal antibodies using envision system. There was positive staining for cytokeratin, chromogranin, and synaptophysin.

***Microscopic description*:** Fragments of cellular neoplasm composed of solid sheets, nests and cords and trabeculae. The tumour cells are medium to large having centrally placed nuclei. The nuclei show moderate pleomorphism, coarse chromatin and brisk mitotic activity. Abundant apoptosis also noted. One section shows cyst wall composed of ovarian stroma and lined by tall columnar epithelium.

Comments: The presence of the surface epithelial stromal component confirms the ovarian origin and differentiates it from metastasis small cell tumor to the ovary. The histology results were confirmed from other laboratory as well, which reported carcinoid tumour with mucinous cystadenoma within one ovary. Urinary excretion of 5-hyroxy indole acetic was reported as 7.5mg/24 hour.

The lady was referred to Institute for Chemo-radiation, where she had two courses of chemotherapy only and then did not turn up for follow up.

DISCUSSION

Primary neuroendocrine carcinoma of non small cell type in the ovary is an extremely rare neoplasm. It is distinct histologically from ovarian carcinoma of the small cell pulmonary type, which are composed of small cells with scanty neoplasm and oval to spindle shaped nuclei.[1]

Ovarian carcinoids develop in pure form or in association with the other tumours, mainly teratomas. Ovarian neuroendorine carcinomas belong most probably to surface epithelial neoplasms, which express endocrine pathway of differentiation. The neuroendocrine carcinoma of non-small cell type is characterized by the presence of islands, sheets and trabeculae with little intervening stroma and cellular homogeneity.[1] To date, ten ovarian neuroendocrine carcinomas have been reported in world literature.[1,4] Aga khan University in Karachi has reported one case.[5] In a series of five reported cases, three presented in stage one, one in stage 2, and one in stage 3.[6] By In eight cases, this was mucinous adenoma carcinoma, one patient had endometriod adenocarcinoma, and one patient has mucinous cyst adenoma. The patient reported by Aga Khan University also had mucinous cyst adenoma.[5] Our patient had mucinous cystadenoma as well. The age of the patients ranged from 22-71 years.[3]

This composite tumor is highly aggressive. Prognosis is extremely poor with metastases to peritoneum, liver and other abdominal organs[7] of eight cases with follow up information, all had died of disseminated tumour, six within 10 months, and two in 19 months and 3 years respectively, after diagnosis. Only the neuroendocrine carcinoma component was found in metastatic sites.[6] One case of neuroendocrine carcinoma with mucinous cystadenoma was reported to be having uneventful clinical course ten months after surgery with chemotherapy.[8]

The value of CA 125 as tumour marker in the follow up of the patients with neuro endocrine carcinoma to judge treatment response is not helpful as reported The level of CA 125 decreased despite the presence of a clinically progressive tumour, while the level of urinary 5-hydrooxyindoleacetic acid increased 16 weeks before pelvic recurrence was detected. This indicated that it was the neuroendocrine component, and not the mucinous component of the tumour, that was not responsive to the chemotherapy.[9]

Our patient had raised level of Ca 125 with normal urinary 5-hydrooxyindoleacetic acid. The therapeutic protocols including chemotherapy and irradiation have not been established yet with most ovarian neuroendocrine carcinomas. Our patient received two courses of chemotherapy only and then did not come for follow up.

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