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## **Case Report**

# **Ovarian Primary Neuroendocrine Carcinoma of Non-Small Cell type: report of an extremely rare Neoplasm**

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### **Abstract**

Paraffin block sections of a uterus and ovarian mass from a 31 year old female were sent for second opinion to the Pathology Laboratory of Aga Khan University.

Histologic examination and immunohistochemical features gave a diagnosis of primary neuroendocrine carcinoma of non-small cell type admixed with benign mucinous cystadenoma.

This is a rare tumour with only eight being reported in literature.

### **Introduction**

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.<sup>1</sup> Only a handful of cases have been reported worldwide. All reported cases are associated with a surface epithelial-stromal neoplasm which may be benign, borderline or malignant.<sup>2</sup> Histologically, these neoplasms show neuro-endocrine differentiation. They are always unilateral and age range is between 22 and 77 years.<sup>3</sup> Prognosis is extremely poor with metastases to peritoneum, liver and other abdominal organs.<sup>4</sup> We report a case of this extremely rare neoplasm.

### **Case Report**

We received four paraffin blocks for second opinion. The previous histopathology report was attached. The patient

was a 30 year old female. According to the gross description on the report, the specimen consisted of a uterus with a large ovarian mass. This measured 15x12x11 cms and on sectioning showed a solid grey cut surface with necrotic areas. In addition, multilocular cystic foci were present which were filled with mucinous material. The ovary of the other side was also attached and was unremarkable. Histopathology report was carcinoid tumor with mucinous cystadenoma in one ovary. The other ovary was histologically normal.

The four blocks received by us for second opinion were all from the ovarian mass. On histologic examination, these showed a malignant neoplastic lesion composed of nests, trabeculae and sheets of large tumor cells with pleomorphic hyperchromatic nuclei, prominent nucleoli and abundant cytoplasm. Numerous mitotic figures were seen and foci of necrosis were noted. (Figures 1 and 2).

Immunohistochemically the tumor cells showed

by a single layer of benign mucinous epithelium. No nuclear pleomorphism, stratification of epithelium or invasion into underlying stroma was seen (Figures 3 and 4).

Based on the histological and immunohistochemical

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Figure 1. Section showing trabeculae and sheets of large tumor cells. H&E (X 10).

Figure 2. Section showing large tumor cells showing large vesicular nuclei, prominent nucleoli, pink cytoplasm and mitoses. H&E (X 20).

strong positivity for the following markers: cytokeratin MNF, Chromogranin and Synaptophysin.

In one of the sections, a mucinous component was identified. This was composed histologically of a cyst lined

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Figure 3. Section showing the presence of surface epithelial (mucinous) component and neuroendocrine component. H&E (X10).

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Figure 4. Section showing mucinous surface epithelial component H&E (X 20).

features, a diagnosis of primary neuroendocrine carcinoma of non-small cell type admixed with benign mucinous cystadenoma in the same ovary was made. The presence of the mucinous component supports the diagnosis of the former.

### Discussion

Only eight cases of this distinct neoplasm have been reported in world literature.<sup>1-5</sup> Grossly, these tumors are similar in appearance to other malignant ovarian tumors with grayish cut surface along with hemorrhagic and necrotic areas.<sup>1</sup> Histologically, these tumors are composed of solid sheets, islands, nests and cords of tumor cells. The cells are large having big, pleomorphic nuclei. The nuclei are hyperchromatic or vesicular, some having prominent nucleoli. Cytoplasm is variable in amount ranging from scanty to abundant. Mitotic activity is variable although it is usually brisk and abnormal mitoses are present.

scanty to abundant. Mitotic activity is variable although it is usually brisk and abnormal mitoses are present. Immunohistochemically, the tumor cells show positivity with cytokeratins, neuroendocrine markers chromogranin, synaptophysin, neuron specific enolase and serotonin.<sup>2-4</sup>

The tumor may morphologically resemble carcinoid tumor of the ovary but cells are larger with much greater cellular and nuclear pleomorphism. In addition, the presence of surface epithelial-stromal component helps to differentiate between these tumors. It is very important to distinguish between the two since prognosis of neuroendocrine carcinoma of non-small cell type is much worse. In the series of five cases reported by Eichhorn et al, three patients died in a period of eight to thirty-six months postoperatively in spite of the fact that two of these patients received chemotherapy and the third one received radiation therapy. In the case reported by Jones et al<sup>4</sup>, the patient developed peritoneal implants and liver metastases and died of disease within ten months of initial presentation. This poor prognosis is seen in spite of the fact that six of the eight reported cases were stage I tumors at the time of initial presentation.<sup>1</sup>

The size of the tumor cells and the presence of surface epithelial stromal component distinguishes this neoplasm from ovarian carcinoma of small cell pulmonary

type.<sup>3</sup> However, the distinction between these neoplasms is not important prognostically. The presence of surface epithelial stromal component confirming the ovarian origin differentiates this tumor from metastatic small cell carcinomas to the ovary.<sup>1,5</sup>

Of the eight reported cases, surface epithelial-stromal component was mucinous in seven. One patient had an endometrioid carcinoma.<sup>1</sup> Of the five cases reported by Eichhorn et al<sup>2</sup>, two patients had mucinous adenocarcinoma while two other patients had borderline mucinous neoplasms with foci of invasion. Our patient had a benign mucinous cystadenoma. The case reported by Jones et al<sup>4</sup> also had a mucinous cystadenoma.

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