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## **Case Report**

### **Clear cell carcinoma of ovary with associated mucinous cystadenoma and endometriosis**

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#### **Abstract**

A 45 year old woman presented with right sided ovarian mass with multiple omental deposits and liver metastases. The right ovary was enlarged and showed a partly cystic partly solid cut surface. Histological picture showed clear cell carcinoma with areas of mucinous cystadenoma and endometriosis. Clear cell carcinoma is known to be associated with endometriosis. To the best of author's knowledge, it's association with mucinous cystadenoma has been described only once in the literature, where clear cell carcinoma was shown to be associated with mucinous cystadenoma without any evidence of endometriosis.

#### **Introduction**

Clear cell carcinoma is a relatively uncommon type

of ovarian tumour which comprises 2.4% of ovarian epithelial neoplasms.<sup>1</sup> Although they were initially called mesonephromas and mesonephric carcinomas, their epithelial nature is now accepted due to their high association with pelvic endometriosis.<sup>2,3</sup>

We describe a case of clear cell carcinoma of the ovary in association with mucinous cystadenoma and endometriosis. To the best of our knowledge, only one such case has been published previously in the literature by N Dutt et al<sup>4</sup>, in which clear cell carcinoma was described associated with mucinous cystadenoma without any evidence of endometriosis.<sup>4</sup> This association was criticized by WG McCluggage as according to him the mucinous areas represented mucinous metaplasia of the endometriotic cyst of ovary.<sup>5</sup> However in our case, there were separate foci of mucinous cystadenoma and

endometriosis coexisting with areas of clear cell carcinoma, supporting the case of N Dutt et al.

### Case Report

A 45 year old woman presented to the clinician with abdominal pain and distension since one month. Ultrasound examination revealed a right-sided ovarian mass with multiple omental deposits and liver metastases. Per-operatively, in addition to the enlarged right ovary, the left ovary was also found to be enlarged, and the omentum studded with tumour deposits. Bilateral salpingo-oophorectomy was performed along with an omental biopsy which was sent for histological evaluation.

Grossly the right ovary was enlarged, measuring 9.5 x 9 x 6 cm. with ruptured capsule and a partly cystic, partly solid cut surface. The solid areas were pale brown, soft, necrotic and spongy. The cystic spaces were filled with mucinous secretions and included a haemorrhagic cyst. The

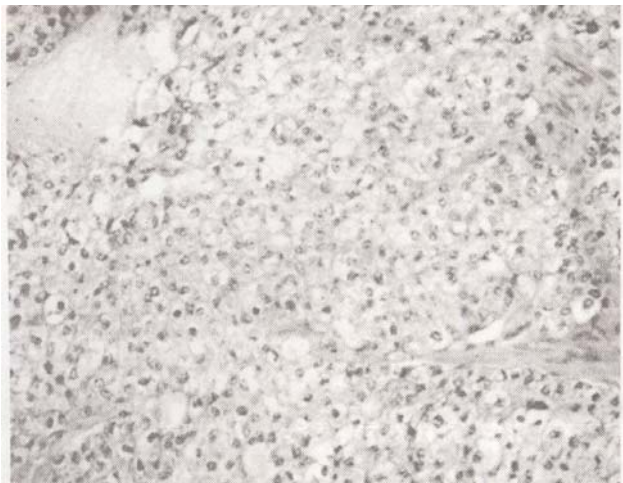


Figure 1. Clear cell carcinoma of ovary showing sheets of polygonal cells with clear cytoplasm. X40.



Figure 2. Ovarian tissue showing clear cell carcinoma (long arrow) with area exhibiting lining epithelium of mucinous cystadenoma (short arrow). X20.

left ovary measured 6 x 6 x 4 cm. and showed cystic spaces filled with dark brown haemorrhagic material.

Histological examination of the right ovary revealed an infiltrating neoplasm composed of solid sheets and nests of large polyhedral clear cells having pleomorphic nuclei with clumped chromatin and prominent nucleoli (Figure 1). In some areas multiple layers of these cells were seen lining the cystic spaces with hobnail appearance focally (Figure 2). Tumour cells showed cytoplasmic glycogen on special staining and immunohistochemically, they were shown to be positive for cytokeratin CAM 5.2 and AE1/AE3 and were negative for alphafetoprotein. Cystic spaces lined by a single layer of tall columnar mucinous cells were seen in close proximity to the tumour. There was no cytological atypia or complexity of architecture in these areas. There were foci of endometriosis in the adjacent ovarian tissue. Based on the features, described a diagnosis of clear cell carcinoma of the ovary associated with benign mucinous cystadenoma and endometriosis was made. The right ovary showed features consistent with mucinous cystadenoma along with areas of endometriosis. Deposits of clear cell carcinoma were identified in the omental biopsy.

### Discussion

Clear cell carcinoma was first described in 1939 by Schiller, who called it a "mesonephroma" of ovary due to its presumptive origin from the mesonephric rests in the female genital tract.<sup>3,4</sup> In view of their association with pelvic endometriosis and endometriotic cysts, the frequent admixture with typical endometrioid carcinoma, and ultrastructural similarities with mullerian epithelial cells, their epithelial origin is now generally accepted.<sup>2,6</sup> Clear cell carcinoma comprises 2.4% of ovarian epithelial neoplasms and 7.4% of ovarian carcinomas.<sup>1</sup> The patients are usually in their fifth or sixth decades of life.<sup>1,7</sup> Interestingly, they are the most common epithelial ovarian neoplasms to be associated with paraneoplastic hypercalcaemia.<sup>1</sup> Clear cell carcinoma has an incidence of associated pelvic endometriosis in 50% and endometriosis in the same ovary in 24% of the cases.<sup>2,4,8</sup> Some authors believe that this incidence of association may be much higher if ovarian clear cell carcinomas are extensively sampled because of the possible obliteration of endometriosis by the tumour.<sup>5,8</sup> Benign and borderline clear cell tumours of ovary are very rare and most are malignant with a 5 year survival rate ranging from 37-47%.<sup>4,7,9</sup> Histologically different patterns have been described out of which solid and tubulopapillary are most common.<sup>1,7,9,10</sup> The cells are polygonal with abundant clear cytoplasm separated by delicate fibrovascular septae with papillary cores often exhibiting hyalinization. In the tubulopapillary pattern, the cells show a hobnail appearance. Cytoplasmic

glycogen in the favour of PAS positive, diastase resistant hyaline globules is seen.<sup>1,7</sup> Immunohistochemically, the tumour cells are positive for cytokeratins, epithelial membrane antigen and vimentin and are negative for alpha-fetoprotein. The differential diagnosis includes yolk sac tumour, dysgerminoma and metastatic renal cell carcinoma.<sup>1</sup>

The presence of intracytoplasmic lumina with mucinous inclusions are described in clear cell carcinoma which, according to some authors, support the hypothesis that clear cell carcinoma should be regarded as an end stage transformation, which may arise from any of the other epithelial tumours.<sup>4</sup> Association of clear cell carcinoma with mucinous lesions has been described only infrequently. One such case was described by N Dutt et al. where clear cell carcinoma was seen associated with mucinous cystadenoma.<sup>4</sup> This association was criticized by WG McCluggage, according to whom the mucinous areas were in fact reproductive mucinous metaplasia of the endometriotic cyst of ovary.<sup>5</sup> According to him, mucinous metaplasia in endometriotic cysts can be so extensive that a diagnosis of mucinous cystadenoma may be considered.<sup>5</sup> However, in our case there were separate foci of mucinous cystadenoma and endometriosis

coexisting with areas of clear cell carcinoma. There was no continuity between the areas of endometriosis and mucinous cystadenoma in our case. This favours the case of N Dutt et al who suggested the origin of clear cell carcinoma in a background of mucinous cystadenoma.

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