Report of a rare case of carcinosarcoma Stage IV

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Introduction:

Carcinosarcoma (CS) or malignant mixed müllerian tumor is a bizarre gynecological neoplasm with diverse origin and histopathological characteristics with few reported cases in the literature. With an incidence of 2/100.000 and 5-years survival rate at stage I of 50% and at stage IV of 0% [1]. The most common location is the uterine corpus and the classical symptoms are postmenopausal bleeding and abdominal pain [2]. Because of its aggressive nature, it mostly presents distant metastasis at diagnosis. The optimal management is controversial and required multidisciplinary approach which includes surgery, chemotherapy, radiotherapy and hormonal therapy [1].

Case Report:

A 46 year old Thai woman, GI/PI, was referred to our clinic with two weeks history of nocturnal pyrexia, moderate abdominal pain for 4 weeks, cough, vertigo and obstipation. An abdominal CT scan showed multiple nodules and tumor masses distributed over the entire abdominal cavity concentrated at the pelvic region (Fig.1). In the meantime, blood results indicated severe bacterial sepsis and a positive influenza B blood test. Blood cultures were negative. Serum levels of CA125 and CA15-3 were increased.

As laparoscopy revealed extensive peritoneal carcinomatosis, moderate ascites, Fitz-Hugh-Curtis syndrome and massive adhesions enable uterus exposure a representative specimen was taken and submitted for histological examination with the suspicion of genital tuberculosis. A QuantiFERON-TB Gold test was performed with a negative result. An inconclusive histology showed features of chorioncarcinoma, which would not correlated to negative hCG levels and missing tumor cells in ascites.

The patient underwent laparotomy with omentectomy tumordebulking and resection of disintegrating tumor masses at mesenterium and terminal ileum with ileostomy. A VAC dressing was applied and due to instability the patient was transferred to the intensive unit care. Histologically, there was a biphasic neoplasm with poorly differentiated mixture of mostly epithelial and sarcomatoid elements with necrotic areas and polymorphonuclear cells. There was a strong suspicion of carcinosarcoma or chorioncarcinoma. Because of still inconclusive results the sample was send to further immunohistochemistry diagnostic to a reference pathology center.

At that point, the condition of the patient was continuously deteriorating. Despite care and regular change of VAC the abdominal pressure was increasing daily. The patient passed away four weeks after being admitted to the hospital despite all the treatment due to multiple organ failure. After dead, the definitive histology revealed a mostly poorly differentiated spindle cell carcinoma with anaplastic components. It was presented the following immunohistochemistry findings: positive reaction for estrogen and progesterone receptors, p63, SAMARCA4 and SMARCB1; the p-53 immunoreactivity was negative such as the reactivity for desmin and caldesmon. These features were categorized with a high probability as a highly aggressive carcinosarcoma with extremely poor prognosis.

Fig. 1. CT Scans in coronal plane showing multiple mesenteric, retroperitoneal and intraperitoneal nodules and tumor masses distributed over the entire abdominal cavity concentrated at the pelvic region.

Discussion:

Histologically, CS is a biphasic malignancy consisting of malignant epithelial and mesenchymal components. Several hypotheses regarding its histogenesis have been proposed. However, there is now enough evidence to consider CS as metaplastic carcinomas in which the sarcomatous elements derive from the carcinoma during the evolution of the tumor [3]. Here we present a rare case of a highly aggressive carcinosarcoma at Stage IV in a patient without risk factors and with unusual morphological and immunohistochemically features showing a predominant carcinomatous component with a marked degree of anaplasia and a less proportion of poorly differentiated sarcomatous component. These unusual features may have hindered the properly diagnosis and treatment. A large multicenter trial is necessary and should be encouraged in order to reduce the incidence rate of this disease and improve its management.

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