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Case Report

Mixed Ovarian Tumor Associating a Carcinoid Tumor and A Borderline Mucinous Tumor with Microinvasion: About A Case

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Abstract

Carcinoid tumors of the ovary may be primary or metastatic. Primary carcinoid tumors are rare and the majority of tumors occur in association with a mature cystic teratoma, but a considerable number occur in a pure form. They may also arise in a solid teratoma or mucinous tumor. Histologically, according to WHO, there are four variants: insular, trabecular, strumal and mucinous. They can be mixed with a combination of pure types; most often insular and trabecular. Immunohistochemistry is necessary for confirmation of the diagnosis. Most tumors are seen in perimenopausal women. Two thirds of primary carcinoid tumors are localized and have a good prognosis. Surgery is the treatment of choice based on total hysterectomy with bilateral adnexectomy. The present case report describes a carcinoid tumor associated with endocervical-like mucinous borderline tumor with microinvasion of the ovary in a 49 year old woman.

Keywords: Ovary, carcinoid tumor, mucinous borderline tumor, microinvasion.

INTRODUCTION

Primary carcinoid tumors of the ovary are rare representing 1% of all carcinoid tumors and less than 0.1% of ovarian tumors. There are 4 types: insular, trabecular, strumal and mucinous, of which the insular type is the most es

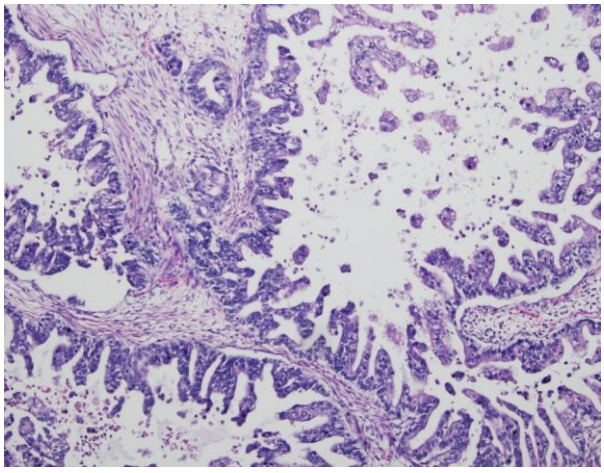

Fig-1: Endocervical-like mucinous borderline tumor with microinvasion. Papillary arrangement with short vegetations (HEx10).

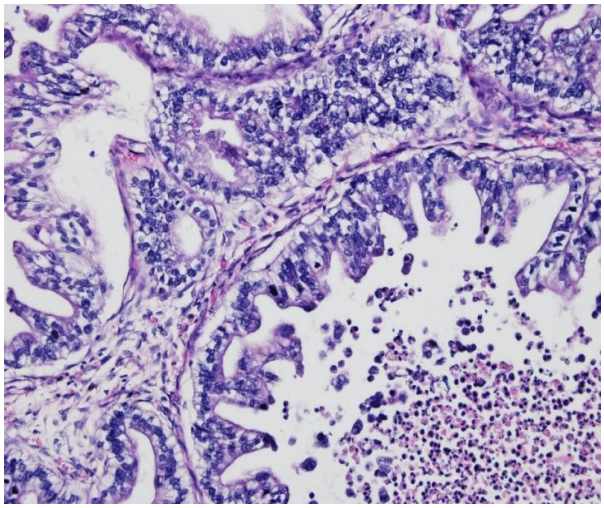

Fig-2: Borderline mucinous tumor showing pluristratifiction and cytonular atypies with microinvasive foci (HEx20)

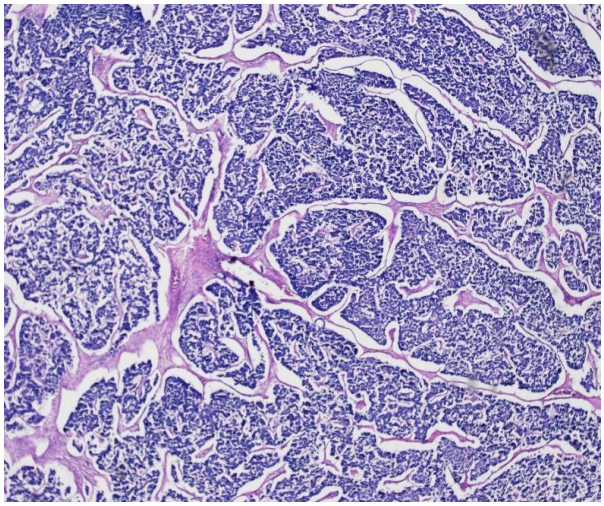

Fig-3: Tumor proliferation of cord and insular architecture in a vascular stroma (HEx10)

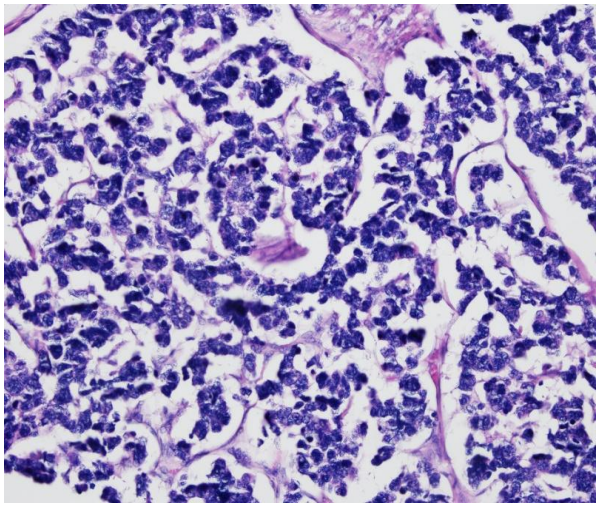

Fig-4: Monotonous cells with granular salt and pepper chromatin with some mitosis (HEx20)

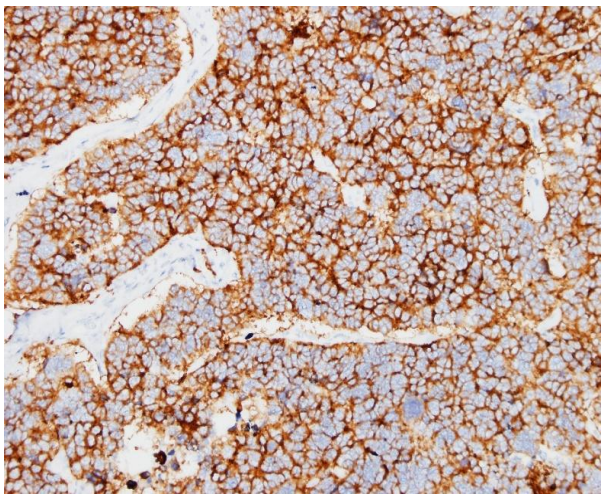

Fig-5: Positive staining for Synaptophysin (HEx20)

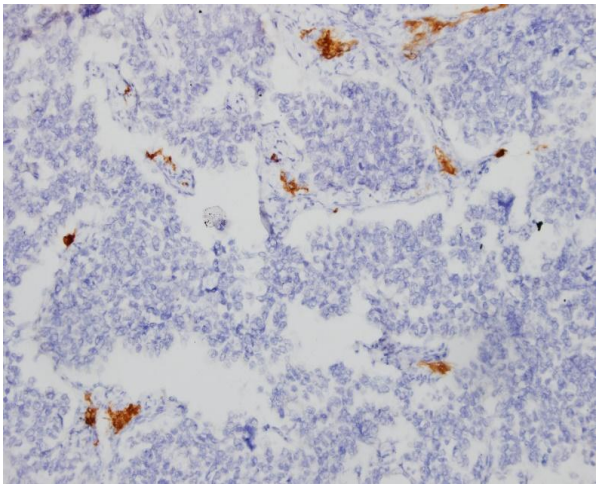

Fig-6: Negative staining for Inhibin (HEx20)

DISCUSSION

Carcinoid tumors are well-differentiated neuroendocrine tumors of low malignancy, located primarily in the gastrointestinal tract and bronchial tree [1]. Carcinoid tumors of the ovary can be primary or metastatic. Primary carcinoid tumor of the ovary accounts for less than 1% of all carcinoid tumors and less than 0.1% of all ovarian neoplasms [2]. Most primary ovarian carcinoids are thought to be teratomatous in origin. Often, the teratoma contains rnucinous cysts or a mucinous cystadenoma [3], but a considerable number of tumors present in a pure form [4]. Histologically, according to WHO, there are four main variants of primary ovarian carcinoid: insular, trabecular, struminal, mucinous and mixed (insular and trabecular) [2]. These carcinoid tumors of the ovary were first described by Stewart et al. in 1939, who reported two cases of an insular carcinoid tumor arising in an ovarian teratoma [4]. A common observation among nearly 200 cases of insular and trabecular ovarian carcinoids were their intimate association with mucinous epithelium; in most cases, the mucinous epithelium consisted of short bands or cysts [3]. Primary island carcinoid tumors of the ovary are usually found in association with, or as part of, a mature cystic teratoma, but they may also arise from a solid teratoma or mucinous tumor [4]. These tumors may also develop in association with a glandular component of the müllerian type (mucinous type tumors, endometrioid carcinoma, and carcinosarcoma) [5]. The age incidence of patients with ovarian carcinoids is highly variable, but most patients are postmenopausal [4], and usually present with a heterogeneous ovarian mass, or as an incidental finding on abdominal radiology [2]. Macroscopically, they form a solid nodule within a cystic teratoma or, when pure, as a yellow-gray solid mass, and vary from microscopic tumors to large tumors measuring more than 20 cm in their largest diameter [4]. Microscopically, the neuroendocrine component grows into the background of a surface epithelial tumor, but the two components

can still be recognized as distinct [5]. Immunohistochemistry is crucial in the diagnosis by the presence of two or more specific markers. Synaptophysin and chromogranin are the classic markers. CD56 can be used, but it is not specific. Estrogen and progesterone receptors are always negative, unlike adenocarcinomas [2]. Two-thirds of these tumors manifest as a localized lesion, while about one-third show distant spread [2]. Primary ovarian carcinoids only occasionally metastasize and should be treated as ovarian tumors with low malignant potential [4]. The appropriate treatment is not well defined, but surgery remains the primary treatment, with chemotherapy reserved for tumors with a high proliferative index, as measured by Ki67 [2].

CONCLUSION

Primary carcinoid tumor of the ovary is a rare entity with an indolent evolution and a good prognosis in localized cases. The diagnosis is histological with immunohistochemical confirmation. Total hysterectomy with bilateral adnexectomy remains the treatment of choice for primary ovarian carcinoid tumors.

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