Sarcoma-like mural nodule in a borderline mucinous tumor of the ovary: A rare entity

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ABSTRACT

Sarcoma-like mural nodule (SLMN) is a very uncommon and misleading benign entity which may be associated with benign, borderline or malignant mucinous neoplasm of the ovary. It should be distinguished from other malignant mural nodules with sarcoma, carcinosarcoma or anaplastic carcinoma for proper management. We report a rare case of SLMN in a borderline mucinous tumor of the ovary in a 30-year-old lady. In spite of having confusing histopathological features the final diagnosis was made depending on the younger age of the patient, well circumscription of the nodule, absence of vascular invasion and immunohistochemical profile.

Key Words: Mural nodule, mucinous, ovary, sarcoma-like

INTRODUCTION

Mucinous cystic tumors of the ovary rarely contain one or more solid mural nodules in which the histological features differ markedly from the background of benign, borderline or malignant mucinous neoplasms.[1] Four varieties of mural nodules have been described, including true sarcomas, sarcoma-like mural nodules (SLMN), foci of anaplastic carcinoma and mixed type.[2] SLMN tend to occur in younger women and are characterized by smaller size, sharp demarcation, a heterogeneous cell population, and no serious impact on the prognosis.[3] It should be differentiated from true sarcomatous mural nodules and foci of anaplastic carcinoma with areas of sarcomatoid differentiation since both of these lesions carry a graver prognosis.[4] We report a very rare case of SLMN in a borderline mucinous tumor of the ovary.

CASE REPORT

A 30-year-old unmarried female patient presented with progressive abdominal enlargement for last 7 months. She also complained of mild lower abdominal pain and discomfort for last 2 and half months. A cystic swelling of about 18–20 weeks size reaching just below the level of umbilicus was felt on physical examination. Her menstrual cycle was normal. Ultrasonography revealed a left ovarian cystic space-occupying lesion (SOL) of 12 cm × 9.6-cm size with internal echogenicity and septations. Uterus was normal in size and right adnexa were unremarkable. Her routine laboratory tests were otherwise normal. Left ovarian cystectomy was performed. Grossly the cystic ovarian mass [Figure 1a] measured 13.8 cm × 12 cm × 8 cm. The outer surface was smooth and grayish whitewith wide areas of congestion. The cut section revealed multilocular cyst containing reddish-brown thick mucoid fluid. An elevated firm, well-defined nodular area measuring 4.8 cm × 3.6 cm × 2.5 cm was noticed on the inner surface of the largest locule. Microscopic examination displayed the histopathological features of a borderline mucinous tumor showing complex cribriform architecture with mild to moderate cellular atypia along with an adjacent circumscribed nodular area [Figure 1b and d]. However, no recognizable glandular structures infiltrating into the deeper part of the nodule were identified. The nodular area was predominantly composed of pleomorphic spindle cell...
cells with hyperchromatic nuclei and occasional prominent nucleoli arranged in a vaguely fascicular pattern. The pleomorphic cells focally demonstrated smooth muscle and rhabdomyoblastic differentiation [Figure 2a and b]. Frequent mitotic figures including atypical forms admixed with histiocytes and inflammatory cells especially eosinophils were noticed. Conspicuous areas of coagulative necrosis [Figure 2c] and hemorrhage were also observed. Immunohistochemistry for cytokeratin was focally positive and vimentin was diffusely positive in the pleomorphic spindle cells of the nodule. The atypical mucinous glands displayed strong cytokeratin positivity [Figure 2d]. Overall, the histopathological features of the nodule closely mimicked that of a sarcoma except the presence of circumscription and absence of vascular invasion. True sarcoma, atypical inflammatory myofibroblastic tumor, carcinosarcoma were considered as differential diagnoses. However, considering the circumscription of the nodule in macroscopy as well as in microscopy along with corroborative histopathological and immunohistochemical findings, the diagnosis of a borderline mucinous tumor with SLMN was made.

DISCUSSION

In 1979, Prat and Scully[5] reported 14 cases of SLMNs, seven of them from the previous literature, and pointed out their favorable clinical behavior. They described three types of morphology in SLMNs namely pleomorphic and epulis type, pleomorphic and spindle cell type and giant cell-histiocytic type. Sarcomatous nodules can exhibit a variety of patterns, including fibrosarcoma, rhabdomyosarcoma, and undifferentiated sarcoma. Mixed nodules may feature carcinosarcoma or a mixed anaplastic carcinoma and sarcoma-like nodule.[2] The present case displayed the morphology similar to the pleomorphic and spindle cell type with areas of smooth muscle and rhabdomyoblastic differentiation.

In contrast to SLMN, the sarcomatous nodules and foci of anaplastic carcinoma which can also present as mural nodule, tend to occur in older patients and are characterized by larger size, poor circumscription, a monotonous spindle cell population in the former, evidence of carcinomatous differentiation in the latter, and aggressive behavior.[3] Anaplastic carcinomatous nodules generally have spindled and rhabdoid cells with unequivocal cytological features of a high-grade malignancy, and are associated with intraepithelial or invasive mucinous carcinoma in 72% of cases.[4] Atypical inflammatory myofibroblastic tumors may resemble SLMNs possibly due to a florid reaction to intramural hemorrhage or spilling of mucinous material in cystic ovarian tumors. In contrast to SLMN that occur in association with an epithelial tumor, inflammatory myofibroblastic tumors have a predilection for relatively younger patients, appear de novo, and are larger and less well circumscribed.[7] Although various hypotheses have been proposed to explain the histogenesis of SLMN, it probably represents a reactive and self-limited phenomenon within a neoplasia. SLMN usually strongly express vimentin and focally or weakly express cytokeratin, similar to true sarcomas yet distinct from anaplastic carcinomas that usually have strong/diffuse cytokeratin expression.[8] Thus even immunohistochemistry failed to differentiate SLMN in our case from true sarcoma. Finally the age of the patient, well circumscription of the nodule and lack of vascular invasion were the only parameters that led us to the correct diagnosis. In the recent past, Agarwal et al.[9]...
and Chakrabarti et al.\textsuperscript{10} from India reported the cases of SLMNs in benign mucinous tumor of ovary.

The outcome of mucinous tumors with SLMNs is the same as the corresponding category of mucinous tumors without the nodule.\textsuperscript{[1]} But SLMNs should be regarded with caution as their histologic appearance is worrisome and clinical follow-up data are very limited.\textsuperscript{[3]}

Hence, careful and meticulous examination of this rare lesion within a mucinous cystic tumor is essential for reassuring the patient of a favorable clinical outcome and for excluding the similar looking aggressive lesions having poorer outcome.

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