A Rare Case Of Well Differentiated Papillary Mesothelioma Of Peritoneal Origin

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Abstract: Solitary well differentiated papillary mesothelioma is an unusual variant of epithelial mesothelioma. Most of them exhibit either benign or indolent behavior. Making the differential diagnosis between this rare tumor and serous papillary carcinoma can be problematic. We report here a case of a 24 year-old unmarried female with a well differentiated papillary mesothelioma of peritoneal origin.

Key Words: Mesothelioma; Ovary; well differentiated papillary mesothelioma.

I. Introduction

Well differentiated papillary mesothelioma (WDPM) is an unusual variant of epithelial mesothelioma. It occurs mainly in the peritoneum, and this tumor is most commonly seen in women who have no history of asbestos exposure. It is often found incidentally at laparotomy that is done for other indications.¹,²,³ WDPM rarely occurs at other sites, including the ovary,³,⁴ pericardium,⁵ and the tunica vaginalis.⁶,⁷ The tumor is occasionally found as a frozen sectioning because surgeons suspect this tumor to be another malignant tumors, such as diffuse malignant mesothelioma, or peritoneal dissemination from other tumor sites or ovarian serous papillary carcinoma.¹ Most WDPMs exhibit either a benign or indolent behavior.¹,⁸ Making the differential diagnosis between this rare tumor and serous papillary carcinoma can be problematic. WDPM must be distinguished from the significantly more aggressive lesions to avoid unnecessary treatment.¹,⁸ We report here a case of a 24 year-old unmarried female with a well differentiated papillary mesothelioma of peritoneal origin.

II. Case Report

Miss X aged 24 years was brought to DR.PSIMS & RF with pain of lower abdomen since 1 month duration. She has no h/o menstrual irregularities or history of asbestos exposure. On examination her vitals are stable. P/A is soft. She is non co-operative on per rectal examination. On gynecological US Rt ovary – 3.8 x 1.8 cm. A heterogenous lesion measuring 6.8 x 3.9 cm seen in right adnexa in close approximation with anterosuperior border of right ovary peripheral and internal vascularity noted within the lesion.

Her CA 125 – 472 u/ml; CEA – 4.5 ng/ml; αFP – 2.1 IU/ml; CA – 19.9 – 3.8 IU/dl.

On CT scan right ovary- measures approximately 2.8 x 1.9 cm. Evidence of complex cystic lesion seen in Right adnexal region measuring approximately 6, 8 cm in transverse diameter & 4.6 cm anteroposteriorly. The lesions is in close contact with superolateral border of urinary bladder and superior to fundus of uterus. The lesion is in contact with right ovary. Lesion is showing nodularity in continuity with adjacent of cyst on post contrast study. At least two small cysts seen adjacent to the adnexal region on either side measuring 2 cm on each side possibly arising from respective ovarian tissue.

Laprotomy done - An irregular mass of size 5 x 4 cm seen adjacent to the superior border of Right ovary from peritoneum seen. Mass is friable and easily removed. Grape like vesicles in appearance. mass removed & sent for frozen section.

On frozen section – well differentiated papillary mesothelioma of peritoneal origin.

On histopathological examination - tumour showing papillary structures with columnar epithelial lining. The papillae have abundant stroma. Focal areas showing a complex papillary structures lined by cuboidal to columnar cells with a few cells showing eosinophilic and few cells have clear cytoplasm. Few cells exhibiting pleomorphism. At one focal area there is epithelial dipping into stroma forming tubules. There are necrotic foci with nuclear debris.

Patient has been referred to oncologist for further evaluation where it has been diagnosed as well differentiated papillary mesothelioma of peritoneal origin which has benign and indolent course.

The patient has remained well after a follow-up of seven months.
Pic 1: Irregular mass of size 5 x 4 cm seen adjacent to the superior border of Right ovary from peritoneum seen.

Pic 2: Grape like vesicle in appearance

Pic 3: Evidence of complex cystic lesion (20 – 30 HU0 seen in Rt adnexal region measuring approximately 6.8 cm in transverse diameter & 4.6 cm anteroposteriorly
III. Discussion

WDPM is considered to be a benign subtype of mesothelioma that predominantly affects the peritoneum of women in the third and fourth decade. The histologic appearance of WDPM is that of thin fibrovascular papillae lined by relatively uniform cuboidal mesothelial cells. Psammoma bodies are occasionally encountered. WDPM must be differentiated from metastatic peritoneal adenocarcinoma, serous papillary adenocarcinoma, diffuse malignant mesothelioma and reactive mesothelial hyperplasia.

The most important point in the differential diagnosis of this case was to rule out serous papillary adenocarcinoma of ovary. The absence of multilayering, cell stratification, cytologic atypia and mitotic activity supported a diagnosis of WDPM. Calretinin is a 29-kDa calcium binding protein that is expressed in central and peripheral neural tissues, and in a limited number of nonneural tissues including normal and neoplastic mesothelium. Calretinin was previously identified to be the most sensitive (88%) and specific (100%) mesothelial marker to differentiate diffuse peritoneal mesothelioma in women from serous papillary ovarian carcinoma and serous papillary peritoneal carcinoma. This case showed reactivity for calretinin and cytokeratin, but any reaction for CEA was negative. The immunoreactivity for CEA and CD15, and another glycoproteins, such as Ber-EP4, TAG-72, and MOC-31 is quite useful in supporting a diagnosis of adenocarcinoma.

Diffuse malignant mesothelioma (DMM), in which well-differentiated papillary elements are prominent, can also be confused with WDPM. DMMs are usually bulkier than WDPMs and they demonstrate diffusely infiltrative cytologically atypical cells. Abnormal mitoses and bizarre cells are also found in DMM. Patients with WDPM generally have a far better prognosis than those with DMM.

WDPM must also be distinguished from reactive mesothelial hyperplasia. Papillary structures may occur in mesothelial hyperplasia. However, in comparison the conspicuous papillary pattern of WDPM, such changes are rare in with hyperplasia. The lack of inflammation, the accompanying reactive changes in the adjacent serosal tissue and the relatively uniform or monotonous tumor cells are essential points for the diagnosis of WDPM.

Pic 4: Fibrous papillae show a single layer of cuboidal mesothelial cells The tumor cells stain positively for calretinin (Immunohistochemical stain).
IV. Conclusion

It is important to diagnose this tumor in order to refrain from an aggressive therapeutic approach. The histological findings that include the cytologic features and the immunohistochemical study results are important for making the correct diagnosis.

References