Well-Differentiated Papillary Mesothelioma of The Peritoneum: A Pathological Analysis.

Abstract:

Well-differentiated papillary mesothelioma of peritoneum (WDPM) is a rare subtype of peritoneal epithelioid mesothelioma which typically has low malignant potential. It most commonly occurs in young females. A 35-year-old-female with complaints of right lower abdominal pain underwent diagnostic workup and subsequent surgery revealed a flat abdominal mass loosely adherent with peritoneum. The surface of the mass was smooth with firm consistency.

Histopathology, immunohistochemistry and special stain confirmed the diagnosis of well differentiated peritoneal mesothelioma. WDPM in females is frequently asymptomatic and associated with indolent course and usually diagnosed incidentally.

Key words: WDPM; peritoneum; mesothelioma; immunohistochemistry

Introduction

Well differentiated papillary mesothelioma (WDPM) is a very uncommon neoplasm of peritoneum. Most frequently it appears like little numerous nodules on peritoneal surface, omentum or mesentery. WDPM is characterized by indolent course and is diagnosed in most cases incidentally during operations for other reasons. Sometimes differentiation between WDPM and other mesothelial lesions may be difficult. In case of simultaneous well-differentiated papillary mesothelioma (WDPM) and carcinoma involving other organs the surgeon may be mistaken by considering peritoneal implants of WDPM as tumor metastasis. This situation may result in overtreatment of the patient. Thus a thorough pathologic examination of the specimens taking care not to miss any areas of invasion, and utilizing immunohistochecmical analysis when necessary are important to avoid such mistakes. Well-differentiated papillary mesothelioma (WDPM) of peritoneum is a rare subtype of peritoneal epithelioid mesothelioma which typically has low malignant potential. It most commonly occurs in young women lacking a history of asbestos exposure.

WDPM is a rare and unusual mesothelial tumor characterized by a lack of deep invasion and associated with an indolent clinical course and long survival. For these reasons, WDPM is best considered as a specific clinico-pathologic entity distinct from conventional diffuse malignant mesothelioma.

Case history

A female patient of 35 years hailing from Mymensingh was admitted to a local clinic with complaint of pain in right iliac region. She was operated upon with suspicion of acute appendicitis. Incidentally, during operation a smooth surfaced flat mass was partially seen in peritoneal cavity. A small biopsy was taken from the mass and sent for histopathological examination to local histopathological laboratory. The diagnosis was adenocarcinoma. The patient came to BSMMU, Dhaka, for further confirmation of her diagnosis and better management. CT-guided FNAC was done from the mass and the cytological diagnosis again was adenocarcinoma. Then the patient was operated upon to remove the mass. Peroperatively, a large flat mass loosely attached with peritoneum was found and was removed smoothly without much difficulty. Other organs were found to be normal. The whole mass was sent for histopathological examination to the department of pathology, Banga Bandhu Sheikh Mujib Medical University (BSMMU), Dhaka. On gross examination, the tissue was a flat piece, already sectioned, with smooth surface and gray white in colour. The consistency of the tissue was firm and cut surface was solid. On histopathological examination, the tissue showed many papillary fronds lined by columnar epithelium with moderate amount of cytoplasm and bland-looking nuclei pushed to the apices of the cells. No mitosis was seen. Areas of fibrosis were also seen. The differential diagnoses were mesothelioma, reactive mesothelial proliferation and adenocarcinoma. With the available facilities, confirmed diagnosis of the tissue could not be made. The paraffin blocks, H & E stained slides and clinical and operative findings were sent to Singleton Hospital, UK for confirmatory diagnosis. They performed special staining and immunohistochemistry to make the diagnosis. Mucin stain was negative. The cells were negative for CEA (Carcino Embryonic Antigen) and BerEP4 and strongly positive for CK (Cytokeratin) and calretinin. Staining for EMA (Epithelial Membrane Antigen) was not helpful. The final diagnosis they reported was a variant of mesothelioma called WDPM.
Discussion:

Well differentiated mesothelioma remains a rare, enigmatic entity possessing low malignant potential but requiring long term surveillance. As a result, the identification of a mesothelial-based papillary proliferation mandates cautious pathologic examination in order to prevent both under “diagnosis” (yielding consequence such as failure to treat the patient) or “over diagnosis” (resulting in aggressive treatment for malignant mesothelioma generally not warranted for this rare variant). WDPM cases have been reported in both men and women, involving the pleura, peritoneum, and tunica vaginalis. Occasionally, well differentiated papillary mesothelioma may involve two cavitary surfaces simultaneously most often the pleural and pleural and peritoneal surfaces. In one study, twenty-six of the 32 patients for whom microscopic findings were reported had predominantly well developed papillary architecture with branching or coarse papillae and central vesicular nuclei. The remaining six patients had a prominent tubulopapillary pattern. The surfaces of the papillae were covered with a single, uniform layer of cuboidal mesothelial cells. Because a specific marker for mesothelioma has not yet been identified, the accuracy of the diagnosis relies on the use of battery of immunohistochemical markers.

From practical stand point, a current recommended protocol for differentiating epithelial mesotheliomas from peritoneal serous carcinomas involves the use of two markers for mesothelioma and adenocarcinoma: calretinin and cytokeratin 5/6 and CA 19-9 and MOC-31 respectively. In our case, the cells were strongly positive for calretinin and cytokeratin and negative for carcinoembryonic antigen. These immunoprofile supports the diagnosis of mesothelioma rather than adenocarcinoma. Given the rarity of the disease, it is crucial that we continue to learn about WDPM from isolated reports and small case series.

References: