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Teaching cases

Well-differentiated papillary mesothelioma manifesting in a hernia sac

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ABSTRACT

Well-differentiated papillary mesothelioma (WDPM) is a tumor of uncertain malignant potential that usually occurs as a multifocal lesion of the female peritoneum, and is incidentally found at the time of surgery. We present here a multifocal case that had arisen from the lining of a hernia sac. To our knowledge, only four cases of this event have been previously described. A review of the five cases reported, including our case, revealed that the mean age of the patients was 56.6 ± 8.35 years. There was predominance in men (4:1). In four cases, the lesion was incidental. Most tumors were found in inguinal hernias. Four cases presented with gross abnormalities in the hernia sac. All the five patients were alive with no evidence of WDPM after a mean follow-up of 38.6 months. Extensive sampling of this rare lesion helps to rule out an epithelial malignant mesothelioma and prevents overtreatment.

Introduction

Malignant epithelial tumors presenting within hernia sacs are rare, occurring in less than 0.5% of surgically excised hernia sacs [14].

Well-differentiated papillary mesothelioma (WDPM) is a primary tumor of the serosal membranes; it is rare and distinctive, with a tendency toward superficial spread without invasion. This neoplasm arises principally in the peritoneum of women of reproductive age [7], but also occurs in the pleura, pericardium and tunica vaginalis testis. WDPM can occur as single [10] or multiple lesions usually of small size. Localized WDPMs invariably pursue a benign course.

The majority of tumors consisting of multiple small nodules behave in a benign or indolent fashion. However, occasionally, WDPMs may pursue an aggressive course, resulting in death of patients [4,9]. Therefore, this neoplasm is currently considered a lesion of uncertain malignant potential.

A WDPM presenting as a finding at the time of clinical or pathological examination of a hernia sac is an exceptional event. As far as we are aware, only four cases of this occurrence have been previously reported [2,7,13,15]. We report herein a new case of this rare event.

Case report

A 66-year-old male with good general status presented with bilateral reducible inguinal hernia. The right one was indirect and the left one direct. He had no history of exposure to asbestos or occupation in the construction industry.

Both sacs were resected, and Lichtenstein hernia repair was performed using polypropylene graft. Grossly, two irregular gray yellow membranous tissue fragments measuring 6.5 cm × 4 cm × 2 cm and 7 cm × 2.5 cm × 1 cm were received. The first fragment corresponding to the right hernia did not show any abnormalities. The second fragment, corresponding to the left hernia, showed multiple reddish excrescences ranging in size from 0.1 cm to 2 cm. Histopathologic study revealed a multicentric neoplasm composed of pedunculated (Fig. 1A) or broad-based (Fig. 1B) elevated lesions of broad papillae with fibrous, hyaline or edematous cores lined by uniform, bland, cuboidal cells (Fig. 1C) showing moderate acidophilic cytoplasm and central nuclei with fine chromatin (Fig. 1D). There was no evidence of mitosis. Invasion of the submesothelial connective tissue was not present. An immunohistochemical positive staining was obtained for cytokeratin 5/6, calretinin (Fig. 2), and WT-1. Stains for epithelial membrane antigen (EMA), desmin, GLUT-1, IMP3, and p53 were negative.

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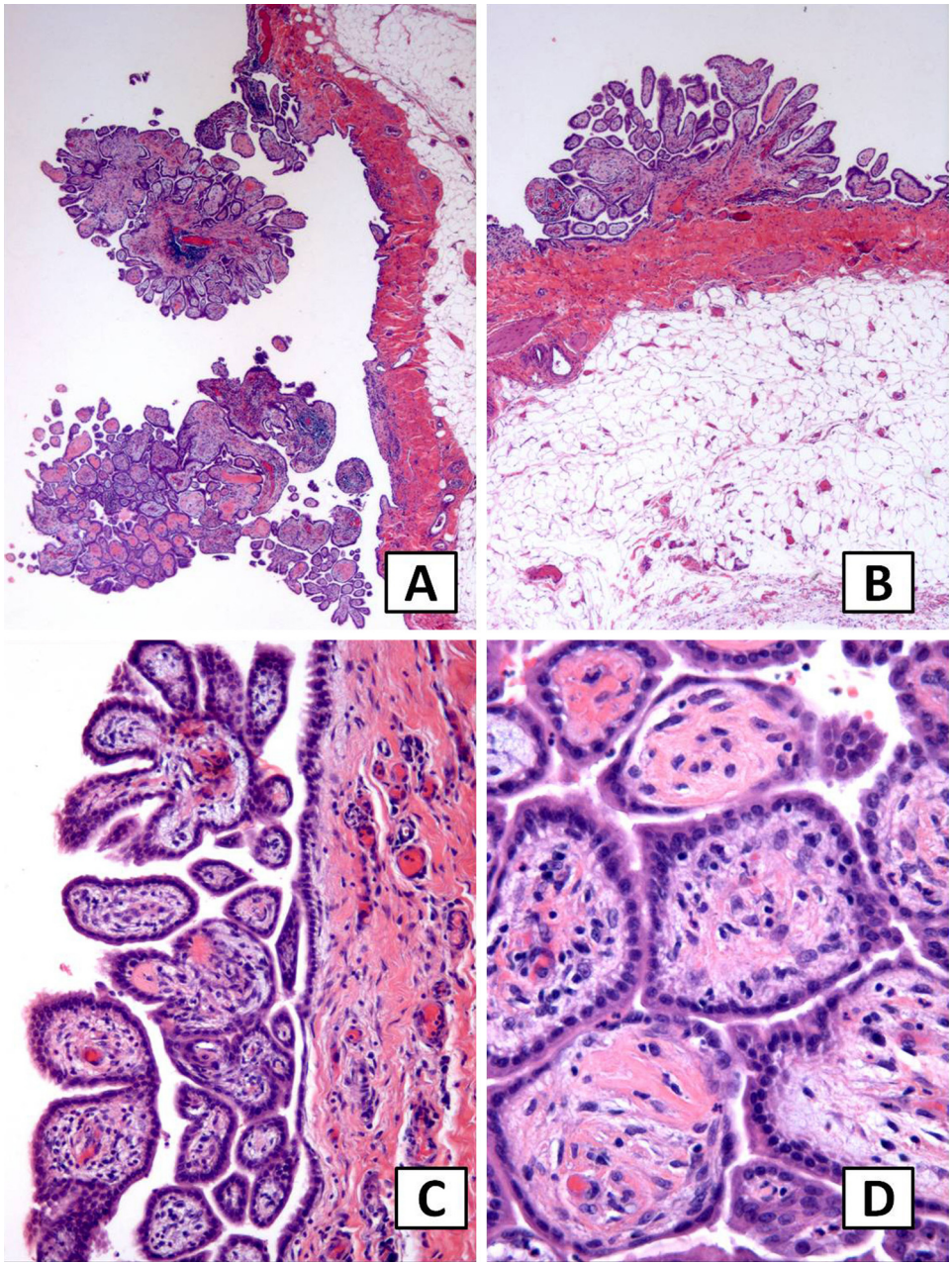

Fig. 1. Well-differentiated papillary mesothelioma in a hernia sac. (A) Multicentric tumor showing characteristic stalks. (B) Broad-based elevated lesion. (C) The tumor shows coarse papillae with fibrous, hyaline, or edematous cores covered by a single row of cuboidal cells. (D) These cells are bland and uniform.

One month later, following the diagnosis of WDPM, the patient was further investigated with a computed tomography (CT) scan of the abdomen and chest which revealed a slight effusion around liver, spleen, intestine loops and in Douglas cul-de-sac. There were no nodules in the mesentery or omentum. Chemotherapy was not instituted. He was advised to undergo a regular follow-up. After an interval of 103 months, the patient is asymptomatic and well with no evidence of disease.

Discussion

WDPM is regarded as a tumor predominantly affecting the peritoneum of women in the third and four decade. However, the tumor may occur over a wide age range and in male patients. In most patients, the WDPM is an incidental finding at laparotomy for a benign or malignant process [12]. On the other hand, these lesions may be associated with chronic abdominal pain, cramping, constipation, weight loss, bloating, ascitis, hemoperitoneum and even acute abdomen [4,7,8,13]. Occasionally, patients have a history of asbestos exposure [4].

WDPMs are usually multifocal, each individual lesion ranging in size from 0.1 cm to 2 cm [13]. Microscopically, the tumors show a papillary architecture that may be accompanied by a tubular, glandular, or trabecular pattern. Scant psammoma bodies may be occasionally seen. Tumor cells are uniformly bland not showing large nucleoli. Mitotic figures are sparse or absent and necrosis is not present. The mesothelial nature of the cuboidal tumor cells can be confirmed with immunohistochemical staining. These cells are reactive for broad-spectrum cytokeratin, cytokeratin 5/6, calretinin, thrombomodulin and WT1. Weak multifocal staining can be obtained with PAX8 [12].