A Case of Encapsulating Peritoneal Sclerosis Complicated by Malignant Peritoneal Mesothelioma

Genta KANAI1, Takatoshi KAKUTA2, Takashi HIRUKAWA3, Chizuko OKAMATSU4 and Masafumi FUKAGAWA1

1 Division of Nephrology, Endocrinology and Metabolism, Tokai University School of Medicine
2 Division of Nephrology, Endocrinology and Metabolism, Tokai University Hachioji Hospital
3 Department of Internal Medicine, Isehara Kyodo hospital
4 Department of Pathology, Tokai University School of Medicine

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We report a case of peritoneal mesothelioma discovered in a patient during peritoneal dialysis. The patient was a 55-year-old woman who had no history of asbestos exposure. Owing to end-stage kidney failure, she had been undergoing peritoneal dialysis for over 8 years, and she had been diagnosed with encapsulating peritoneal sclerosis. She was admitted to the hospital for intestinal obstruction. Three months later, she noticed an enlarging mass in the epigastric region. Computed tomography showed a 10-cm mass originating in the abdominal wall that had invaded the liver. It was diagnosed as malignant mesothelioma via biopsy. Cases of sarcoma-like mass-forming peritoneal mesothelioma are rare, and there are no prior reports of encapsulating peritoneal sclerosis complicated by malignant peritoneal mesothelioma. Thus, this unique case of peritoneal mesothelioma can provide us with important knowledge about this rare entity.

Key words: peritoneal dialysis, encapsulating peritoneal sclerosis, peritoneal mesothelioma

INTRODUCTION

Malignant mesothelioma is a malignant primary neoplasm that has a high degree of invasiveness in the tunica vaginalis, pericardium, peritoneum, and pleura. The widespread use of asbestos as a building material in the past led to an increase in the worldwide incidence of malignant mesothelioma. Peritoneal mesothelioma related to asbestos exposure is as aggressive as pleural mesothelioma, but low-grade peritoneal mesothelioma, which is unrelated to asbestos exposure, is diagnosed in many women [1-3].

In recent years, there have been reports that vascular endothelial growth factor (VEGF) is involved in epithelial-mesenchymal transition [4, 5], and this transition may have been implicated in the peritoneal failure experienced by peritoneal dialysis patients. Encapsulating peritoneal sclerosis that occurs as part of the end-stage course of peritoneal failure in long-term peritoneal dialysis patients has a poor prognosis because it leads to death due to intestinal obstruction and infection. However, its mechanism of development remains unclear. There are no prior reports of encapsulating peritoneal sclerosis complicated by malignant peritoneal mesothelioma. Here, we report a case we experienced of a rapidly progressive mass-forming type of malignant peritoneal mesothelioma in a peritoneal dialysis patient.

CASE REPORT

The patient was a 55-year-old woman. She had no history of asbestos exposure. At the age of approximately 20 years, she was diagnosed with hypertension and idiopathic aldosteronism. At the age of 33 years, she was diagnosed with chronic heart and kidney failure and began receiving outpatient treatment. At the age of 41 years, the disease had progressed to end-stage kidney failure and she began undergoing peritoneal dialysis. Because she developed pleuroperitoneal communication as a complication after starting continuous ambulatory peritoneal dialysis, she underwent thoracosopic surgery.

Eight years after starting peritoneal dialysis, the patient experienced repeated intestinal obstructions, and prednisolone administration was initiated for treating encapsulating peritoneal sclerosis. At the age of 50 years, because of reduced dialysis efficacy owing to decreased peritoneal function, she was placed on a course of once-per-week additional hemodialysis. At the age of 55 years, she required hemodialysis 3 times per week, and as a result, peritoneal dialysis was used only to exchange dialysate once per day for cleansing.

That same year the patient experienced abdominal tension. As ultrasonography did not reveal a lesion, she was placed on observation. Two months later, she was hospitalized for adhesive intestinal obstruction, but no mass was observed in the abdominal cavity. Five months later, a mass appeared in the epigastric region and rapidly enlarged. Bacteria were not detected in ascites. Computed tomography revealed a 10-cm mass originating in the abdominal wall with liver invasion (Fig. 1, 2). Biopsy results indicated malignant mesothelioma (Fig. 3). Biopsy results indicated malignant mesothelioma (Fig. 3).

Six months after first noticing symptoms, the patient was hospitalized and underwent chemotherapy. Initial chemotherapy consisted of 40 mg cisplatin and...
Fig. 1  Computed tomography (CT) scan of the abdomen. Contrast CT image shows the tumor with a liver invasion to uplift from the abdominal wall of the left hepatic lobe. It was a marginal irregular tumor with weak contrast effect in the $10 \times 4$ cm.

Fig. 2  Positron emission tomography (PET) scan of whole body. We performed PET/CT at the time of 1 hour and 2 hours after the administration of the fluoro-deoxy-glucose (FDG). Blood glucose level was 111 mg/dl. Strong accumulation of FDG was observed in the tumor of the liver surface from the anterior abdominal wall.

Fig. 3  Pathological specimen: (a) hematoxylin-eosin stain, and immunohistochemical analysis: (b) calretinin, (c) vimentin, (d) D2-40, (e) cytokeratin AE1/AE3, (f) CD146. Tumor cells are necrotic and tend to be large-nucleus atypical cells that proliferate. Mesothelioma shows a pattern reminiscent of the pleomorphic type.
mesothelioma. The tumor formed tubercles in the liver because of direct invasion in the diaphragm and local invasion in the peritoneum, and it formed a large number of tubercles in both lungs owing to vascular invasion in the epicardium and lymphangiofibrosis. There are very few cases of sarcoma-like peritoneal mesothelioma, as it is typically epithelial [3]. In the present case, because there were no tumor cells in any abdominal organs other than the liver, we believed it was a sarcoma-like mass-forming mesothelioma. Therefore, the symptoms of intestinal obstruction and pain were markedly worse than the ascites.

Wide areas of the peritoneum were sclerotic, and the abdominal organs were firmly adherent to the intestinal tract, forming a single mass. There was no marked hardening of the retroperitoneum. Histologically, the peritoneum underwent fibrotic changes including thickening because of hyalinization and calcification, which led us to suspect encapsulating peritoneal sclerosis. We observed necrosis in the tumor and organs due to circulatory collapse caused by hypotension prior to the patient's death. In particular, the fact that necrosis was observed over a larger area in the abdominal organs as compared to the thoracic organs was probably because the peritoneal sclerosis was not only an obstruction but also affected circulatory dynamics. The peritoneal mesothelioma was localized in the abdominal cavity and did not present with diffuse spreading as is often seen in pleural mesothelioma. Peritoneal sclerosis is the main feature of encapsulating peritoneal sclerosis, and neoplasm seeding was not indicated.

According to previous studies, the glycosylphosphatidylinositol-anchored cell-surface protein known as mesothelin is highly expressed in mesothelioma and is thought to contribute to tumor cell proliferation and adhesion [8]. It is believed that secreted mesothelin can be used as a marker because it has been reported that its blood levels increase as kidney function decreases [5]. Experimentally, it is known that decreased peritoneal function in peritoneal dialysis contributes to the promotion of the epithelial-mesenchymal transition of the mesothelium due to VEGF [4]. As it is known that malignant mesothelial cells express VEGF receptors, increased understanding of this mechanism may provide clues that allow us to elucidate the relationship between peritoneal failure and malignant mesothelioma.

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