Clear-Cell Myomelanocytic Tumor of Ligamentum Teres Hepatis

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Abstract

Clear-cell myomelanocytic tumor (CCMT) of falciform ligament/ligamentum teres hepatis is a very uncommon lesion described as a variant of perivascular epithelioid cell tumor (PEComa), which includes smooth muscle cells, blood vessels, and express melanocytic and smooth muscle markers. Most patients are asymptomatic or have nonspecific gastrointestinal symptoms, and the tumors are usually found incidentally. The origin and definitive diagnosis of the CCMT can be made by histopathological examination of the specimen after the surgical excision. PEComas express HMB45, HMSA-1, Melan A, Mart 1, microphtalmia transcription factor (Mitf), actin, and less desmin. In this case report, we presented our first experience with a CCMT of ligamentum teres hepatis.

Keywords: Clear-cell myomelanocytic tumor, HMB45, ligamentum teres hepatis, perivascular epithelioid cell tumor

Ligamentum Teres Hepatisin Berrak Hücreli Myelomelanositik Tümörü

Öz

Falciform ligament ve ligamentum teres hepatis'in berrak hücreli miyomelanositik tümörü (BHMT), düz kas hücrelerini, kan damarlarını içeren ve melanositik ve düz kas belirteçlerini eksprese eden perivasküler epiteloid hücre tümörünün (PEComa) bir varyantı olarak tanımlanan çok nadir görülen bir lezyondur. Çoğu hasta asemptomatiktir veya spesifik olmayan gastrointestinal semptomlara sahiptir ve tümörler genellikle tesadüfen bulunur. BHMT'nin orijini ve kesin tanısı, cerrahi eksizyon sonrası piyesin histopatolojik incelemesi ile konulabilir. PEComa HMB45, HMSA-1, Melan A, Mart 1, mikroftalmi transkripsiyon faktörü (Mitf), aktin ve daha az desmin ifade eder. Bu vaka sunumunda ilk kez karşılaştığımız ligamentum teres hepatisin BHMT'ünü sunduk.

Anahtar Kelimeler: Berrah hücreli myelomelanositik tümör, HMB45, Ligamentum teres hepatis, Perivaskuler epiteloid hücreli tümör

Clear-cell myomelanocytic tumor (CCMT) of falciform ligament/ligamentum teres hepatis is a very uncommon lesion defined first in 2000 [1]. It is described as a variant of perivascular epithelioid cell tumor (PEComa) that includes smooth muscle cells, blood vessels, and express melanocytic and smooth muscle markers [1-3]. PEComas consist of different clinical forms, such as angiomyolipoma, lymphangioleiomyomatosis, clear-cell sugar tumor of the lung, CCMT of the ligamentum teres hepatis, and other rare clear tumors [4].

PEComas are most common in young women and found in multiple organs, including uterus, vulva, liver, rectum, heart, urinary bladder, abdominal wall, pancreas, skin, and bone [1, 5, 6]. All PEComas, except the pancreatic and uterine ones, have been shown to be associated with tuberous sclerosis complex [1]. They may cause pain owing to compression effect of mass, but they are often recognized incidentally [4].

CCMT of ligamentum teres hepatis is a rare tumor, and as far as we know, only 12 cases have been reported in the literature. Because of this rarity, little is known about CCMT's behavior. Herein, we present our first experience with a CCMT of ligamentum teres hepatis.

Case Presentation

A 28-year-old woman admitted to general surgery department with epigastric pain and nausea. There was no personal or family history of tuberous sclerosis. Physical examination showed tenderness and palpable intra-abdominal mass in the epigastrium. Tumor markers were in the normal ranges. Computed tomography (CT) and magnetic resonance imaging showed a 10 × 9 × 8 cm mass adjacent to the left lobe of the

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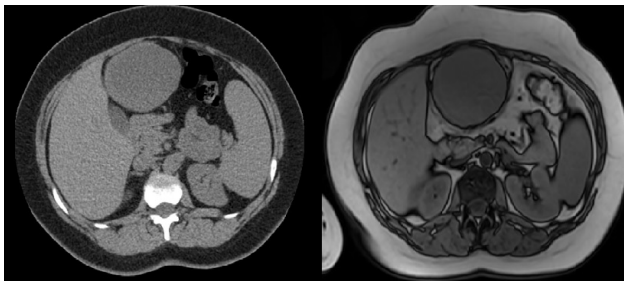

Figure 1. CT and magnetic resonance images of the mass adjacent to the left lobe of the liver

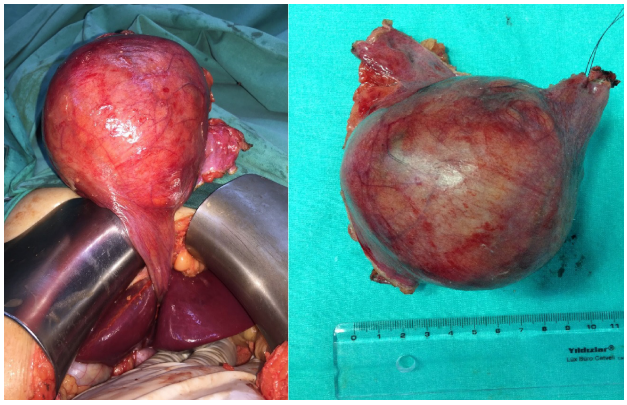

Figure 2. Intraoperative and postoperative images of tumor

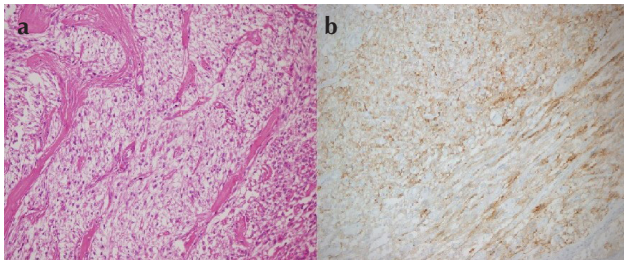

Figure 3. a, b. (a) The tumor is composed of spindled to epithelioid cells with predominantly clear cytoplasm (HEX100). (b) Tumoral cells are immunopositive with HMB45 (HMB45X200)

liver, but not related to the liver or stomach (Figures 1). Patient underwent laparotomy. During exploration, a 12-cm-diameter mass was seen originating from the ligamentum teres hepatis, and it was extending toward the umbilicus near falciform ligament (Figure 2). The mass was excised with negative surgical margins. Pathological diagnosis was CCMT of the ligamentum teres hepatis (Figure 3). Immunohistochemically, the tumor was positive with antibody HMB45 and smooth muscle actin and negative for S-100 protein and pancytokeratin. She was discharged on the fourth postoperative day and followed up with ultrasound (US) every 6 months and CT every year. No recurrence or metastasis was detected during the 5-year follow-up. A written informed consent was obtained from the patient for the study.

Discussion

CCMT, a PEComa variant, shows a predilection for the falciform ligament and teres hepatis. The reason for this anatomical preference is still unclear, but it is considered that they may be differentiated owing to origin from the smooth muscles of the umbilical vessels [1]. Most patients are asymptomatic or have nonspecific gastrointestinal symptoms, and the tumors are found incidentally but also may be presented with an acute abdominal pain [4, 5]. Radiological views of PEComas are nonspecific. Although US and CT show cystic or solid components of the mass and its relationship with adjacent structures, they are insufficient in differential diagnosis. The origin and definitive diagnosis of the CCMT can be made by histopathological examination of the specimen after the surgical excision [2, 4].

Although the pathological processes of PEComas are not fully clear, according to a hypothesis, they may have been caused by neural crest cells that can express melanocyte and smooth muscle phenotype. PEComas are characterized by the proliferation of epithelioid cells around dilated vascular structures. It is typical that most of the cells settled in the perivascular area have epithelial structure and spindled cells that resemble smooth muscle cells located far from the vessels [3, 4]. PEComas express HMB45, HMSA-1, Melan A, Mart 1, microphtalmia transcription factor (Mitf), actin, and less desmin, and do not express S-100 protein. The structure of PEComas includes microfilament bundles with electron-dense condensation, numerous mitochondria, and membrane-bound dense granules [1, 6]. The mass in our report had similar immunohistochemical properties.

The differential diagnosis of PEComas includes clear-cell sarcoma of tendon and aponeuroses, angiomyolipoma, and leiomyosarcoma. Although most of PEComas reported are benign, cases with metastases to lung, pancreas, and bladder in the late-stage follow-up have also been reported [1, 7]. The malignancy criteria of PEComas have not yet been clarified. However, the lesion is more than 5 cm and infiltrative, of high nuclear grade, and have mitotic activity higher than 1/50 high power fields, and vascular invasion and coagulation necrosis are considered to be high risk findings in terms of malignancy [3]. Although in our patient the size of the tumor was greater than 5 cm and had high mitotic activity, it did not metastasize during 5 years of follow-up.

CCMT of ligamentum teres hepatis is very rare, and its definitive diagnosis is made by histopathological examination. Considering that it may have malignant character, total surgical resection of the mass should be performed and patients should be followed up for a long time.

Informed Consent: Written informed consent was obtained from patient who participated in this study.

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