Original Article
Aggressive angiomyxoma of the liver: a case report

Shengnan Qi1, Bingcheng Li1, Jifeng Peng1, Ping Wang1, Wanyu Li1, Yunzhao Chen1,2, Xiaobin Cui1,2, Chunxia Liu1,2, Feng Li1,2

1Department of Pathology, Shihezi University School of Medicine and The Key Laboratories for Xinjiang Endemic and Ethnic Diseases, Chinese Ministry of Education, Shihezi 832002, Xinjiang, China; 2Department of Pathology, The First Affiliated Hospital, Shihezi University School of Medicine, Shihezi 832002, Xinjiang, China

Received July 12, 2015; Accepted September 10, 2015; Epub September 15, 2015; Published September 30, 2015

Abstract: Aggressive angiomyxoma (AAM) is a rare benign mesenchymal tumor that occurs almost exclusively in the soft tissues of the pelvis and perineum. Very few cases of AAM occurring outside these regions have been reported. The present report presents a case of AAM originating from the liver of a 50-year-old female patient. Tumor resection was performed, and pathological examination revealed microscopic features that were characteristic of AAM. Histopathological examination showed that the tumor was composed of scattered spindle- or stellate-shaped cells with thick-walled blood vessels lying in a myxoid stroma. Immunohistochemically, the tumor cells stained positively for CD34, vimentin, and actin. In this paper, we also discuss the differential diagnosis of AAM. To the best of our knowledge, this study is the first to report a case of AAM originating from the liver.

Keywords: Aggressive angiomyxoma, liver, immunohistochemistry

Introduction
Aggressive angiomyxoma (AAM) is a rare benign mesenchymal tumor with myxoid and vascular components that usually affects the pelvic-perineal region. This tumor predominantly occurs among adult female of reproductive age and was first described as a separate histopathological entity in 1983 [1]. Since then, less than 250 cases have been reported in the literature, mostly in the form of small case series or isolated case reports [2]. AAM is a slow growing tumor, but the risks of local extension and postsurgical local recurrence (i.e., 36%-72%) justify surgical excision of the neoplasm [3]. However, uncommon cases in which the neoplasm has features consistent with AAM and occurs in the head, neck, and lung region have been reported [4, 5]. To the best of our knowledge, this study is the first to report a case of AAM originating from the liver.

Case report
A 50-year-old woman who had a history of biliary calculi complained of discontinuous pain of the middle and right upper abdomen for over one day with accompanying symptoms including fever, chills, nausea, and vomiting. Computed tomography (CT) and magnetic resonance imaging showed dilatation of the intrahepatic bile ducts, particularly in the left lateral lobe, with a patchy or nodular density shadow and obvious abnormal strengthening of the hepatic peripheral parenchyma (Figure 1). Left lateral lobe and gallbladder resections were performed. During the intraoperative period, a tumor was found in the left lateral lobe.

The results of macroscopic examination showed a tumor (2.0 cm × 2.0 cm × 1.0 cm) with a whitish-grey soft and glistening external surface. Microscopically, the tumor revealed spindle- or stellate-shaped cells and variably sized thick-walled blood vessels lying in a myxoid stroma and infiltrating with a small amount of lymphocytes mast cells (Figure 2). Cellularity was universally low to moderate, and cellular atypism or mitotic figures were not apparent.

Immunohistochemically, the tumor was positive for vimentin; thick-walled blood vessels were positive for CD34 and SMA and the tumor cells
were negative for desmin, S-100, Ki-67, EMA, ER, PR, CD99, CD10, CAM5.2, and CK19 (Figure 3). Pathological examination confirmed the diagnosis of intrahepatic AAM.

The patient was postoperatively followed up for six months and remained free of apparent tumor recurrence and metastasis.

Discussion

To the best of our knowledge, this report is the first study to document a case of AAM occurring in the liver. Physical examination of other regions of the patient in this case yielded unremarkable results. Hence, we believe that the neoplasm primarily originated from the liver. Although the immunohistochemical features of the tumor were inconsistent with those of AAM, its histological features were completely consistent with those of AAM. The therapy administered to patients with AAM is complete wide surgical excision.

Myxoid or fibrous tumors with numerous vascularity occur infrequently in the liver, and differential diagnosis of these tumors includes myxoid neurofibroma and angiomysfibroblastoma.
Aggressive angiomyxoma of the liver

In myxoid neurofibroma, tumor cells reveal wavy or buckled nuclei and show diffuse immunoreactivity for S-100 protein. The lesion lacks the characteristic vascularity of AAM. AMF represents a rare benign soft-tissue tumor that most frequently affects the vulvo-vaginal region of women aged between 20 and 50 years. AMF is composed of myofibroblastic cells and prominent thin-walled blood vessels within a fibromyxoid stroma. AMF is also characterized by the expression of vimentin, desmin, and CD34, which suggests an undifferentiated mesenchymal tumor with preferential myofibroblastic differentiation.

In conclusion, the clinical and morphological features of the tumor reported here were consistent with those of AAM. To the best of our knowledge, this report is the first study to document a case of AAM in the liver, which is an uncommon location for this particular tumor. Reporting a large series of these tumors may lead to a better understanding of how AAM may occur outside the pelvic-perineal region.

Acknowledgements

This work was supported by grants from the National Natural Science Foundation of China (Grant Nos. 81160322 and 81460404).

Disclosure of conflict of interest

None.

Address correspondence to: Drs. Chunxia Liu and Feng Li, Department of Pathology, Shihezi University School of Medicine, Shihezi, Xinjiang 832002, China. E-mail: liuliu2239@sina.com (CXL); lifeng-7855@126.com (FL)

References


