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Large Aggressive Angiomyxoma of the Liver

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Abstract

Aggressive angiomyxoma is an extremely rare mesenchymal tumor with a mixed component that occurs almost entirely from the pelvis and perineum. Although aggressive angiomyxoma arising from the liver is very rare, uncommon cases have also been reported in the larynx and lung. In the present study, a 37-year-old female who was admitted to our hospital with the desire of pregnancy was found to have a gigantic mass originating from the liver, and a right hepatectomy decision was made as a result of radiological examinations. Postoperative immunohistochemical findings of the resection material were compatible with aggressive angiomyxoma. Cure was achieved with resection and chemotherapy was not applied. No recurrence was detected in the follow-ups.

Keywords: Aggressive angiomyxoma, hepatectomy, immunohistochemistry, liver tumor

Introduction

Aggressive angiomyxoma (AAM) is an extremely rare mesenchymal tumor that usually arises in the vagina, vulva, pelvis, and perineal region of women.[1] It was first described by Steeper and Rosai in 1983 as a gynecological soft tissue tumor in women. The aggressive term emphasizes the infiltrative nature of the tumor and its tendency to local recurrence.[1,2] Apart from the liver, cases of AAM involving other rare regions such as the larynx and lungs have been reported.[3,4] In this report, we present the second largest known case of liver AAM.[5]

Case Presentation

A 37-year-old woman presented to the obstetrics and gynecology clinic for preconception counseling. Abdominal ultrasound and multi-detector computed tomography (CT) examinations were performed as she described swelling during her examination (Figure 1). Multidedector computed tomography (MDCT) showed a heterogeneous iso-hyperechoic gigantic mass measuring 17 × 15 × 12 cm in the liver; therefore, magnetic resonance imaging (MRI) was performed for further evaluation (Figure 2). Magnetic resonance imaging showed a lobular mass with cystic components and areas of hemorrhage in the right lobe of the liver, with no other site of involvement in the body. The mass was hypointense on T1-weighted images and hyperintense on T2-weighted images, with progressive heterogeneous enhancement and non-enhancing linear areas after the intravenous administration of gadolinium. Angiosarcoma was considered in the differential diagnosis. A right lobe hepatectomy was performed since the tumor almost completely filled the right lobe.

The macroscopic examination revealed a tumor measuring 21 × 16.5 × 15 cm with a central scar, conspicuous heterogeneity, and gray, cream, white, soft viscous shiny surface containing large and small nodular structures (Figure 3). Surgical margins were intact. The tumor was composed of cytologically bland, widely scattered small spindled to stellate-shaped cells with ill-defined cytoplasmic borders. The stroma was characterized by prominent myxoid change with fine collagen fibrils along with variably sized vessels ranging from small thin-walled capillaries and venules to larger muscular arteries. There were larger spindled cells with well-developed myoid features (Figure 4).

No mitotic figures, tumor cell necrosis, or pleomorphism were noted. Immunohistochemical analysis of the neoplastic cells showed positive staining of antibodies to desmin, smooth muscle

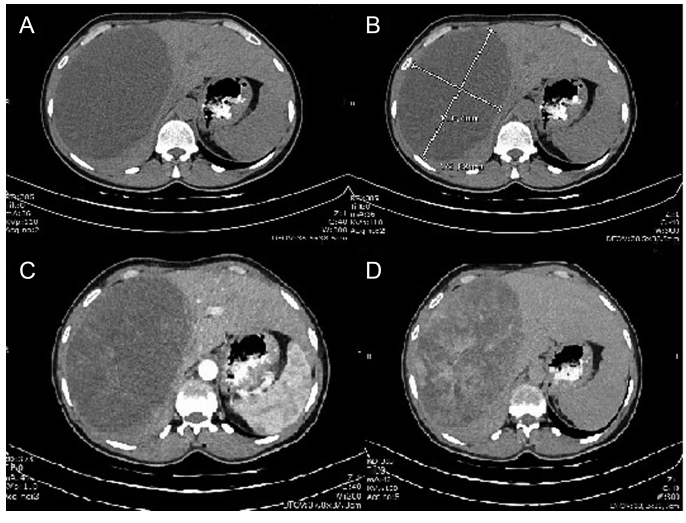

Figure 1. A-D. CT showed a large, regular, hypodense 17 × 12 cm mass in the right lobe of the liver (A–B: precontrast CT image). Blood vessels were observed in the tumor in the arterial phase (C: arterial phase), and the tumor was progressively unevenly enhanced in the delayed phase (D: delayed phase). CT, computed tomography.

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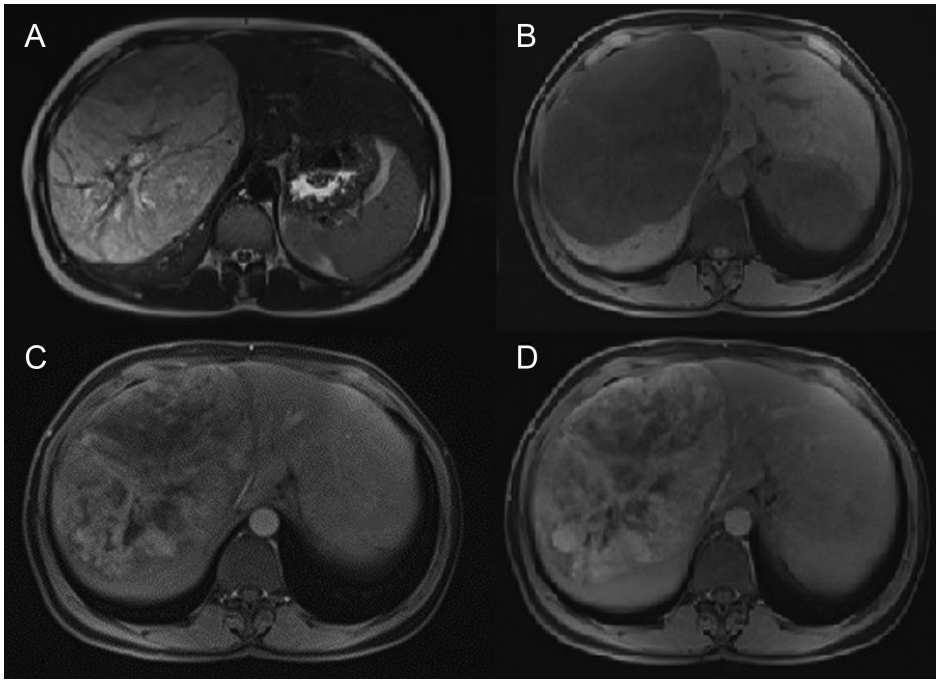

Figure 2. A-D. The lesion showed a slightly increased signal intensity on T2-weighted images (A: T2-weighted image) and slightly decreased signal intensity on T1-weighted images (B: precontrast T1-weighted image). A progressive uneven enhancement was observed after the infusion of contrast material (C: arterial phase; D: delayed phase).

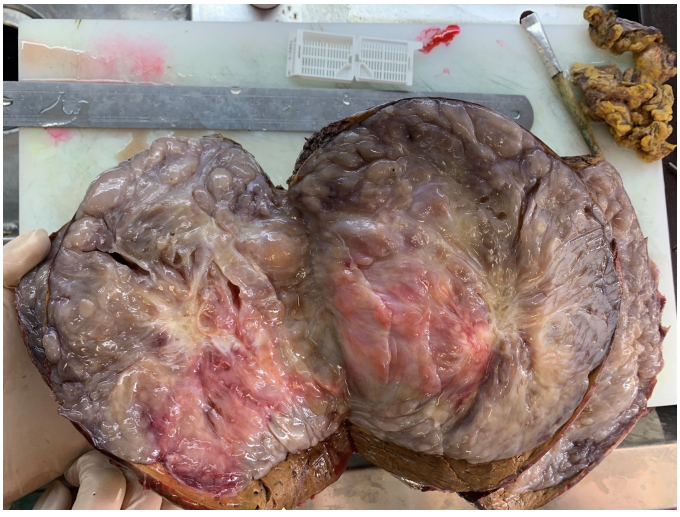

Figure 3. Macroscopic appearance of the tumor.

actin, and h-caldesmon. Vascular structures were stained with CD31 and CD34 but not tumor cells (Figure 5). Labeling for Ki-67 demonstrated a low proliferative index (<1% of tumor cells). Staining of tumor cells was positive for estrogen receptor (ER) and progesterone receptor (PR) in focal areas. The patient was subsequently diagnosed with primary AAM of the liver.

Adjuvant chemotherapy was not administered to the patient. There were no signs of recurrence or distant metastasis postoperatively after 18 months of follow-up. Informed consent was obtained from the patient for this study.

Discussion

Aggressive angiomyxoma is a rare, aggressive, and well-described tumor with both locally infiltrative and recurrent characteristics. The majority of patients are young adult women of reproductive age.[6] Aggressive angiomyxoma almost solely originates from the soft tissues of the pelvis and perineum. Only 4 cases of liver AAM have been reported in the literature yet since it is very difficult to identify this tumor by imaging

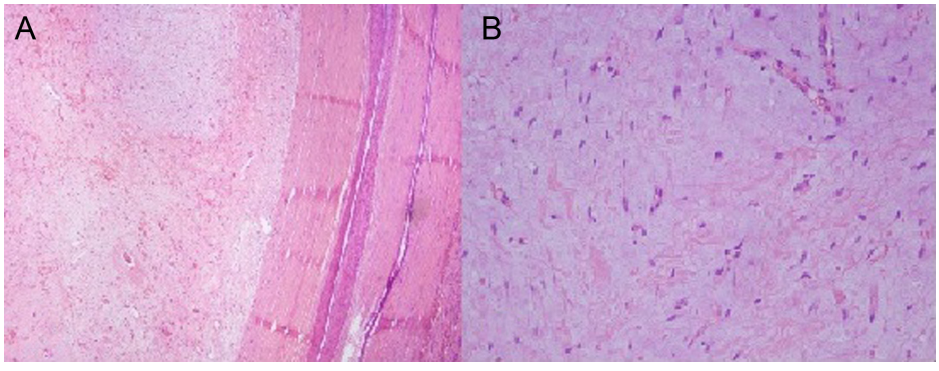

Figure 4. A, B. Tumor is composed of spindled to stellate-formed cells with prominent vascularity and myxoid stroma. (A: hematoxylin and eosin (HE) ×4, B: HE ×20).

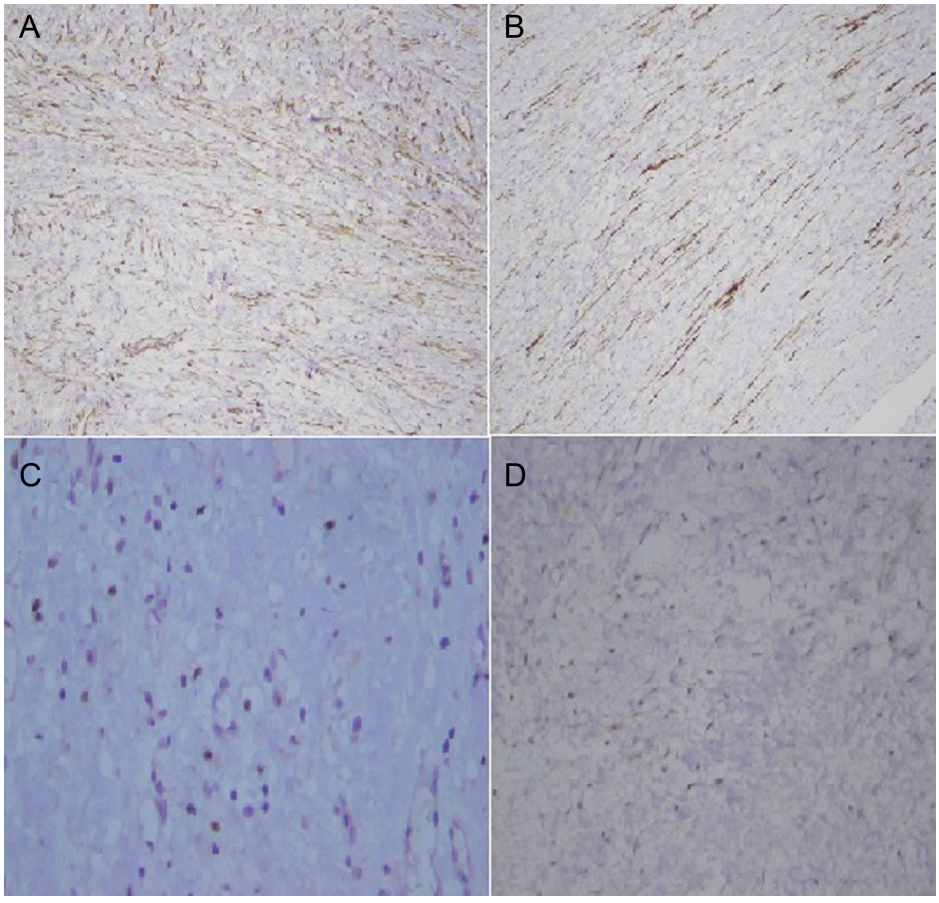

Figure 5. A-D. Tumor cells showed positive immunoreactivity for SMA ×100 (A), desmin ×100 (B), ER ×200 (C), PR ×100 (D). SMA, smooth muscle actin; ER, estrogen receptor; PR, progesterone receptor.

modalities.[3,4,6,7] However, there are some radiological diagnostic clues. The most important one is the dense collagen fibrils contained in the myxoid tumor tissue, which contribute to the typical radiological features of AAM. The characteristic MRI findings of AAM include low signal intensity on T_1-weighted MRI, that is, the same signal intensity as the skeletal muscle, and high signal intensity on T2-weighted images. These appearances are likely to be related to the loose myxoid matrix and the high water content of angiomyxoma. Variable hyperintense and hypointense linear areas appear on T2 and post-contrast T1-weighted images (Figure 2). Immunohistochemically, AAM is positive for vimentin, desmin, CD 34, ER, progesterone receptor (PgR), and α-smooth muscle actin and is negative for MSA and S-100.

The treatment is wide surgical excision due to the high risk of local extension and the high local recurrence rate (up to 70%).[3,8] Although recurrence is seen especially in the first 2 years of follow-up, AAM may rarely recur in the following years.[8] Moreover, hormone therapy has been shown to be useful for the treatment of ER- and PR-positive cases. Gonadotropine releasing hormone (GnRH) agonists may be of value in managing cases of AAM, either primary or recurrent, which are not amenable to surgical excision.[8] Radiation therapy and chemotherapy should not be recommended for AAM because the mitotic index of AAM is usually low.

In conclusion, the present study reports the fifth case of AAM originating from the liver, which is an uncommon location for this rare tumor. Aggressive angiomyxoma should be kept in mind in the differential diagnosis of solid liver lesions. The MRI and MDCT clues are valuable for the diagnosis of AAM.

Informed Consent: Prior to the submission of this article, verbal and written informed consent was obtained from the patient whose clinical photographs are included in this publication.

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