Case Report

Undifferentiated Embryonal Sarcoma of the Liver in an Adolescent with Mesocolon Involvement Mimicking a Colon Tumor with Liver Invasion

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Key Words
Undifferentiated sarcoma;
Liver tumor;
Mesocolon invasion

Undifferentiated embryonal sarcoma (UES) of the liver is an uncommon and highly malignant tumor of mesenchymal origin. It almost exclusively occurs in childhood with a peak incidence between 6-10 years of age[1] but rare occurrences in adults have also been reported. The synonym was first described by Stocker and Ishak in 1978[2] as a distinct clinicopathologic entity. In reviews of pediatric liver tumor, UES was found to be the third most common malignant liver tumor accounting for 13% of hepatic malignancy.[1,2] UES patients often show nonspecific symptoms including fever, abdominal pain, and RUQ palpable mass with normal laboratory study and no reliable tumor marker has been confirmed. Imaging study often reveals large, heterogenous, and hypovascular lesion. Tumors are thought to be aggressive with very poor prognosis initially but long term survival following complete resection with neo- and/or adjuvant chemotherapy has been reported in recent years.[3-5] We report a case of UES with invasion to mesocolon in an 18-year-old young adult who was treated at our institution with combined liver resection and right hemicolectomy.
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An 18-year-old young adult presented with palpable mass lesion, abdominal fullness, body weight loss, and intermittent passage of tarry stool for 2 months. He initially visited a local hospital where abdominal CT scan showed a huge, well-demarcated, heterogenous, multilobular cystic tumor over hepatic flexure colon with suspicion of hepatic involvement (Fig. 1). He was then referred to our institution for further evaluation. The results of laboratory studies were within the normal ranges except for mildly elevated alanine aminotransferase. Tumor markers including alpha-fetoprotein, carcinoembryonic antigen, and CA-199 were normal. The angiography showed a huge tumor in the right abdomen with blood supply from branches of SMA with patent main portal vein and its major branches (Fig. 2). Gastrointestinal stromal tumor from hepatic flexure colon was suspected based on a comparison with a previous abdominal CT scan. At laparotomy the tumor appeared well encapsulated arising from S5-6 of liver with encasement of mesocolon (Fig. 3). To achieve en-bloc resection margin, right hemicolectomy with hepatic bisegmentectomy

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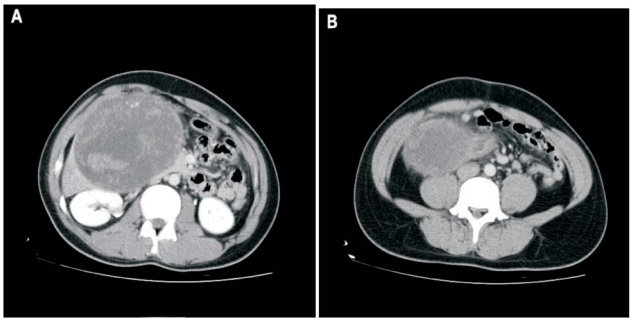

Fig. 1. (A) Abdominal CT scan shows a huge, heterogenous, multilobular cystic tumor in right lobe of liver. (B) The tumor adheres to hepatic flexure colon mimicking colon tumor.

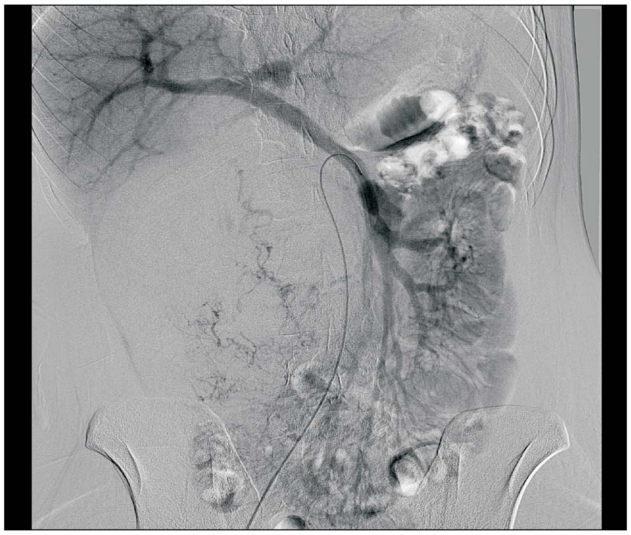

Fig. 2. Angiography shows a large tumor in right abdomen with blood supply from branches of SMA.

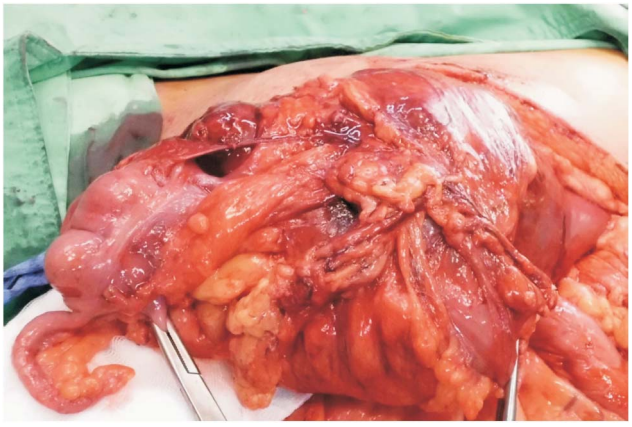

Fig. 3. A well-demarcated tumor arising from right lobe of liver with encasement of mesocolon was noted at laparotomy.

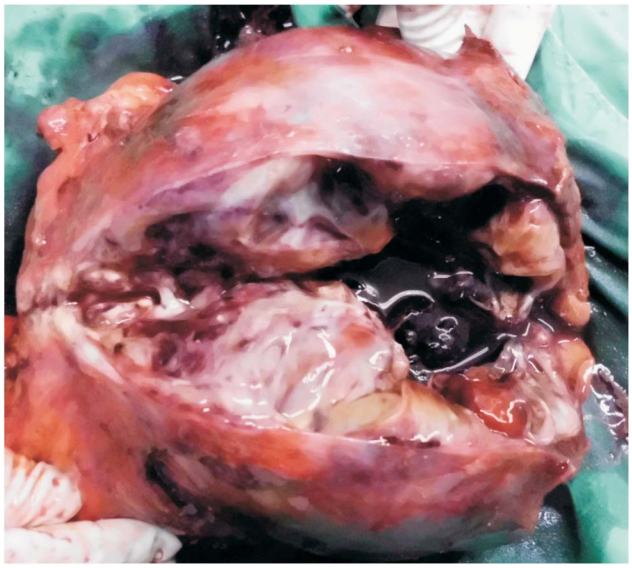

Fig. 4. The gross specimen showed multilobulated cyst with areas of hemorrhage and necrosis.

and cholecystectomy was performed. The tumor measured 14.5 × 14 × 12 cm in size and the cut surface appeared gelatinous, variegated, and yellowish brown in color, with areas of hemorrhage and necrosis (Fig. 4). The surgical margins were free of tumor. Microscopically, the tumor showed a biphasic pattern and was composed of dilated glands lined by eosinophilic simple cuboidal or columnar epithelium in a background of spindle cell sarcoma with focal myxoid liposarcomatous or fibrosarcomatous pattern. The spindle cells showed marked nuclear atypia, cellular pleomorphism, and brisk mitotic activities and contained some intracytoplasmic PAS-positive hyaline globules. Due to the lack of immunopositivity of TLE-1, synovial sarcoma was deemed to be unlikely and gastrointestinal stromal tumor was also excluded because of the absence of immunopositivity of CD117 and DOG1. According to the histomorphology and immunopositivity of AAT and lysozyme, the diagnosis of undifferentiated sarcoma of liver was made. The postoperative course was uneventful except for wound infection which improved with wet dressing. During the follow-up, recurrence of liver tumor at S7 was noted after

5 months and he received wedge resection followed by adjuvant chemotherapy in combination with Doxorubicin, Dacarbazine, Ifosfamide, which was given in 4 courses. However, the liver tumor still recurred and progressed with retroperitoneal infiltration rendering it unresectable condition. Hepatic coma developed thereafter and he expired about 14 months after first operation.

Discussion

Undifferentiated embryonal sarcoma (UES) of the liver is a rare tumor that commonly occurs in pediatric patients and accounts for less than 1% of primary liver tumor in the adults. It mostly arises from the right lobe of liver but UES has also been reported in the left or bilateral lobes. The most common metastasis site is the lung but distant metastasis at the time of diagnosis is rare. The etiology still remains unclear and patients always show non-specific symptoms including palpable epigastric or RUQ mass, dull epigastric pain, and body weight loss.[6] Routine laboratory tests are often within normal limits, as are hepatitis markers, tumor markers, and liver functions, even with huge tumor burden. With regard to the radiographic features of UES, abdominal CT scan and MRI tend to disclose a predominantly cystic appearance whereas in ultrasonography the appearance is predominantly solid.[9] However, the lesion may occasionally appear cystic on ultrasonography and can be mistaken for a hydatid cyst.[10] For detection of vascular invasion, angiography is useful for evaluating the resectability of the tumor. The prognosis of UES has been poor until recently and no standard treatment has been established. Positive surgical margins and spontaneous or iatrogenic rupture of the tumor were thought to be associated with early recurrence and poor outcome. Recurrence is relatively common in the first 2 years and the repeated resection should be performed in combination with chemotherapy if feasible. Bisogno and colleagues have reported long-term survival with routine use of ifosfamide and/or doxorubicin-based neoadjuvant and adjuvant chemotherapy, and imply that the tumor is potentially curable.[4] The other combination regimens similar to that used for rhabdomyosarcoma, including cisplatin, tetrahydro-pyranyl-adriamycin, cyclophosphamide, were also shown to be effective in treating UES with tumor rupture. Furthermore, adjuvant transcatheter arterial chemoembolization may provide another option to improve survival time. Even though early recurrence in our patient may have been due to lack of aggressive chemotherapy after the first operation, no clear correlation between disease-free survival and the choice or schedule of chemotherapy has been demonstrated. Complete resection with multiple approaches, including neo-adjuvant or adjuvant chemotherapy, remains the treatment of choice. In a literature review, some tumors that appeared unresectable at presentation did regress enough to allow excision with clear margin when receiving neo-adjuvant chemotherapy.[4] Therefore, approaches of this type have been advocated by some experts, but no standard chemotherapeutic protocol is recommended due to lack of support from large clinical trials. Although UES is commonly chemosensitive, some such tumors remain unresectable and significant side effects to chemotherapy have also been reported.[7] Another postulated option is liver transplantation, but no reports in adults are available and outcomes in other histological subtypes have been disappointing.[8]

Conclusion

When evaluating a liver tumor, the differential diagnosis should include UES regardless of the patient's age. The treatment for UES is challenging due to its rarity and the difficulty of achieving complete resection at presentation. In some cases, the cooperation of surgeons from different fields and multidisciplinary treatment may be necessary to improve the prognosis.

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病例報告

肝臟未分化胚胎肉瘤合併結腸繫膜侵犯擬似結腸腫瘤

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肝臟未分化胚胎肉瘤是一個少見且由間葉細胞起源之高度惡性腫瘤。此腫瘤最常發生在6-10 歲之幼兒時期，但也有零星成人個案曾被報告。此一名稱最早是由 Stocker 與 Ishak 在 1978 年描述並認定為臨床病理學上之獨特分類。在小兒肝臟腫瘤中，未分化胚胎肉瘤是第三位常見之惡性腫瘤約佔有 13%。病患常以非特異性症狀如發燒、腹痛，及可觸摸之右上腹腫瘤來表現，而其實驗室檢查通常為正常且無可信賴之腫瘤標記可供參考。影像學檢查通常可發現巨大，異質性，及低血管性之病灶。此腫瘤最早被認為較具侵襲性且預後差，但近幾年長期存活已被發現於接受完整切除合併術前或術後化學治療之病人族群身上。此病例報告為一 18 歲年輕成人患有肝臟未分化胚胎肉瘤合併結腸繫膜侵犯之外科完整切除治療經驗。

關鍵詞　未分化肉瘤、肝臟腫瘤、結腸繫膜侵犯。