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Case Report

Undifferentiated embryonal sarcoma of the liver in a 9-year-old girl

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Abstract

Undifferentiated embryonal sarcoma of the liver (UESL) is a rare adult neoplasm. The primary treatments for UESL are total surgical resection and adjuvant multiagent chemotherapy. The current study presents a case of UESL in a 9-year-old female with abdominal pain and fever as her first symptoms. A cystic mass with solid components was discovered on sonography and computed tomography (CT) imaging. The patient underwent tumor removal surgery in addition to a liver lobectomy on the right side. The pathologist reported a single-focal cystic solid mass confined to the liver with no capsular invasion, and the final diagnosis based on the immunohistochemistry (IHC) results was UESL. The patient received chemotherapy, and she is doing well eight months after surgery with no signs of recurrence. In conclusion, large cystic hepatic lesions should be considered in the differential diagnosis. Despite the poor prognosis of UESL, aggressive surgical resection should be the most important factor in ensuring long-term survival.

Keywords: Undifferentiated embryonal sarcoma of the liver, Child, Case report

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Introduction

Malignant mesenchymoma, also known as undifferentiated embryonal sarcoma of the liver (UESL), is a rare and severe malignant tumor that primarily affects children aged 6-10 years old (1). Abdominal pain, palpable mass, and fever are the common clinical features of the disease (2), while cystic lesions in abdominal computed tomography (CT) or magnetic resonance imaging are radiological features (3). There is no discernible gender difference in the disease. Considering that the pathogenic characteristics of UESL are unknown, the other types of hepatic malignant tumors are frequently misdiagnosed as UESL. Postoperative pathology and immunostaining analysis are used to diagnose UESL (4). Because of its high malignancy, UESL frequently metastasizes to the lung, peritoneum, and pleura, resulting in a poor prognosis (5). The primary treatments for UESL are gross total surgical resection and adjuvant multiagent radio-chemotherapy, resulting in improved long-term survival (6). This study reports a case of UESL in a 9-year-old girl. The findings may contribute to a better understanding of child UESL and improvement in the treatment strategy of the disease.

Case Presentation

A 9-year-old girl was admitted to the hospital due to abdominal pain and fever. In the physical examination, a large mass was palpated in the right upper quadrant extending to the right lower quadrant (Figure 1).

A cystic mass with solid components was reported on sonography and CT scan, and the primary radiologic diagnosis was a hydatid cyst.

The patient underwent surgery, including mass excision and right lobectomy. Hepatoblastoma was the primary diagnosis based on surgery findings and the gross tumor shape. The mass was sent to the pathology department, and the pathologist reported a unifocal cystic solid mass, confined to the liver with no capsular invasion. The immunohistochemistry (IHC) study was performed, and the results were positive for vimentin while being negative for hepPar 1, SALLA4, CK, CD31, CD34, myoD1, and Ki67 (MIB1) (90%).

The final diagnosis was “undifferentiated fetal sarcoma of the liver” based on laboratory findings and CT scan results. A CT scan of the chest, abdominopelvic, and bone scans demonstrated no evidence of metastases. The patient received chemotherapy, including ifosfamide, etoposide, doxorubicin, cyclophosphamide, and vincristine, and she is doing well approximately 8 months after surgery with no signs of recurrence.

Discussion

Stocker and Ishak first described and characterized UESL as an uncommon hepatic mesenchymal tumor in 1978 (7). It is a highly invasive malignant tumor of the hepatic primary mesenchymal tissue with distant metastasis. There is no substantial difference in the disease incidence between men and women. Furthermore, the disease is responsible for 5%-8% of all pediatric hepatic tumors. The tumor is most commonly found in the hepatic right lobe (59%) and only rarely develops in the hepatic

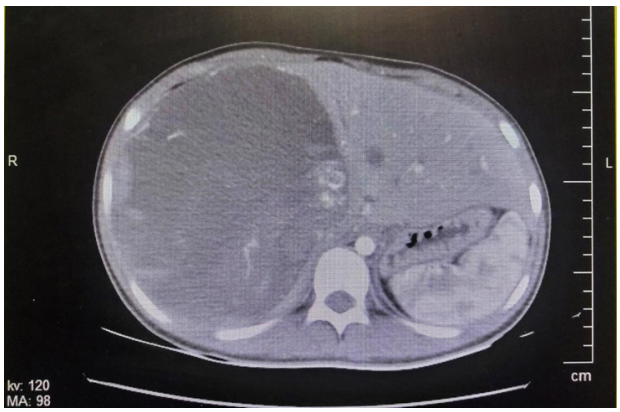

Figure 1. Large mass in the right lobe of the liver

left lobe (22%) or the bilateral lobe (20 %). The UESL has a diameter of 10-25 cm with a single clearly defined boundary (8). In contrast to hemorrhage, necrosis, and cystic degeneration, which are normal, clinical signs such as abdominal bulk, discomfort, fever, and jaundice are rare.

In our case, the radiologic diagnosis was a hydatic cyst. Based on the surgical findings and gross tumor form, hepatoblastoma was the first diagnosis, but the IHC study represented hepatoblastoma. The pathologist diagnosed the mass as a unifocal cystic solid mass restricted to the liver with no capsular invasion. The IHC results revealed that vimentin was positive, while hepPar 1, SALLA4, CK, CD31, CD34, myoD1, and Ki67 (MIB1) were negative.

In most cases, a definitive diagnosis of UESL cannot be made before surgery; instead, postoperative pathological investigation and IHC data are used to make the diagnosis. UESL should be distinguished from hepatoblastoma, embryonal rhabdomyosarcoma, hepatic mesenchymal hamartoma, and hepatic echinococcosis. Primary hepatic parenchymal cells making up hepatoblastoma have minor cellular heteromorphism, limited karyokinesis, enlarged blood sinus, and immunohistochemically confirmed positive expression of AFP, vimentin, and endosomal membrane protein. In contrast, embryonal rhabdomyosarcoma primarily affects infants under the age of six, and the tumor is primarily composed of striated muscle maternal cells in various stages and primary mesenchymal cells. Unlocalized metastases in the lung and adrenal glands were detected in a child with UESL according to Lee et al (5). The size and degree of the differentiation of the tumor were not found to be associated with the prognosis of UESL, while invasion, diffusion, and metastasis were related parameters (9). Because of advancements in therapy, survival rates have significantly increased in recent years, and long-term survival cases have been reported in this regard. The total resection of the tumor followed by a combination of postoperative therapeutic procedures (e.g., chemotherapy, radiation, and interventional therapy) can considerably increase survival rates (10). According to Yu et al, regardless of whether the tumor is ruptured, total resection is the best method for improving survival (11).

In the present case, the patient received chemotherapy with ifosfamide, etoposide, doxorubicin, cyclophosphamide, and vincristine, and she is now doing well around 8 months following surgery with no signs of recurrence.

Conclusion

As a result, it has been proposed that UESL should not be classified as hepatocellular carcinoma. Total resection combined with preoperative or postoperative chemotherapy is now thought to be the most effective way to improve survival rates. To prevent invasive tumor growth, the precise timing of the entire surgical resection is critical, and liver transplantation is the most effective treatment alternative for patients whose tumors cannot be surgically removed.

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Authors' Contribution

Conceptualization: Kiavash Fekri.
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Methodology: Shima Rahmati.
Project administration: Kiavash Fekri.
Resources: Shima Rahmati.
Software: Shima Rahmati.
Supervision: Kiavash Fekri.
Validation: Shima Rahmati.
Visualization: Kiavash Fekri.
Writing – original draft: Kiavash Fekri.
Writing – review & editing: Shima Rahmati.

Conflict of Interests

The authors declare that they have no competing interests.

Ethical Approval

This study protocol was approved by Shahrekord University of Medical Sciences (Approval No. IR.SKUMS.REC.1401.057).

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