Undifferentiated Embryonal Sarcoma of the Liver: An Enigma of Diagnosis

Karaciğerin İndiferansiye Embriyonal Sarkoması: Tanının Gizemi

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ABSTRACT

A 20-year-old man presented with peritonitis requiring emergency laparotomy. He was found to have a huge liver abscess on the right lobe. Open drainage was performed but the abscess was persistently unresolved upon follow up. Computed tomography (CT) revealed a solid liver lesion with raised Ca 19-9. Right hepatectomy was subjected and histology was consistent with a rare and aggressive case of embryonal sarcoma of the liver. Despite oncologic resection, he developed local recurrence and succumbed to death in 6 month postoperatively. We describe a unique aetiology of liver abscess with literature reviews of undifferentiated embryonal sarcoma of the liver.

Key Words: Abscess, liver, embryonal, sarcoma

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ÖZET

Yirmi yaşında erkek hasta acil laparotomi gerektiren peritonit ile başvurdu. Sağ lobda büyük bir karaciğer apsesine sahip olduğu tespit edildi. Açık drenaj yapıldı, ancak apse takiben sürekli olarak çözümlenemedi. Bilgisayarlı tomografi (BT), solid karaciğer lezyonu ile birlikte yüksek Ca 19-9 gösterdi. Sağ hepatektomi yapıldı ve histoloji, karaciğerin nadir ve agresif bir embriyonal sarkomu ile uyumluydu. Onkolojik rezeksiyona rağmen lokal rekürrens gelişti ve postoperatif 6 ayda ölümle sonuçlandı. Burada, karaciğer apselerinin benzersiz etiyolojisini, karaciğerdeki indiferansiye embriyonal sarkomun literatür bilgilerini tanımladık.

Anahtar Sözcükler: Apse, karaciğer, embriyonal, sarkom

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INTRODUCTION

Undifferentiated embryonal sarcoma of the liver (UESL) is an uncommon but extremely malignant neoplasm which usually involves children between 6 to 10 years old (1). Most of these liver malignancies are carcinoma with primary hepatic sarcomas seen only in 0.1 to 2% (2). UESL was firstly described by Stocker and Ishak in 1978 (3). This tumour is categorized as a solid hepatic tumour and rarely presents as a cystic liver tumour (3). The diagnosis is an enigma due to its unspecific and rare presentations. The presentations in adults mimic any other acute abdomen. In view of its predilection in children, its condition in adults poses much more challenges. Herein, we describe a unique presentation of liver abscess that was confirmed as an undifferentiated embryonal sarcoma after hepatectomy.

CASE REPORT

A 20-year-old healthy Malay man presented at a periphery healthcare facility with right hypochondrium pain which was dull aching and associated with nausea, intermittent vomiting and fever. The symptoms were progressively worsening for one week duration. He was hemodynamically stable however physical examination revealed tenderness and board-like rigidity at the upper abdomen. Blood investigation showed an elevated white cell and derangement of liver function test. Fluid resuscitation and antibiotic were initiated. He underwent emergency exploratory laparotomy and intraoperatively revealed a huge liver abscess which occupied within the right lobe of liver. An open drainage was performed and drain inserted.

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There was no improvement in his clinical status with continuous drainage of 50-100 mL/day. However, the abscess culture and sensitivity were negative. He was referred to our institution after repeated ultrasonography showed none resolution of the abscess.

He was clinically and hemodynamically stable, afebrile with drain in situ containing old blood. White blood cell and C-reactive protein were remarkably raised. Hepatitis bloods screening were negative. The tumour marker, serum CA 19-9 was elevated 1508 (normal value: 0-37 U/mL) with normal serum alpha fetoprotein. Computed tomography (CT) of the abdomen showed a liver lesion occupying right side of the liver size 19 x 12.8 x 9.7 cm with presence of area of necrosis within (Figure 1A, 1B). A provisional diagnosis of cholangiocarcinoma was made based on the correlation between CT and tumour markers. A liver biopsy was performed in view of indefinite diagnosis and non-resolving abscess. The liver biopsy suggested a possibility of plasmacytoma which necessitate an extended right hepatectomy. A huge solid-cystic liver mass with vascular invasion and necrotic lymph node was found intraoperatively.

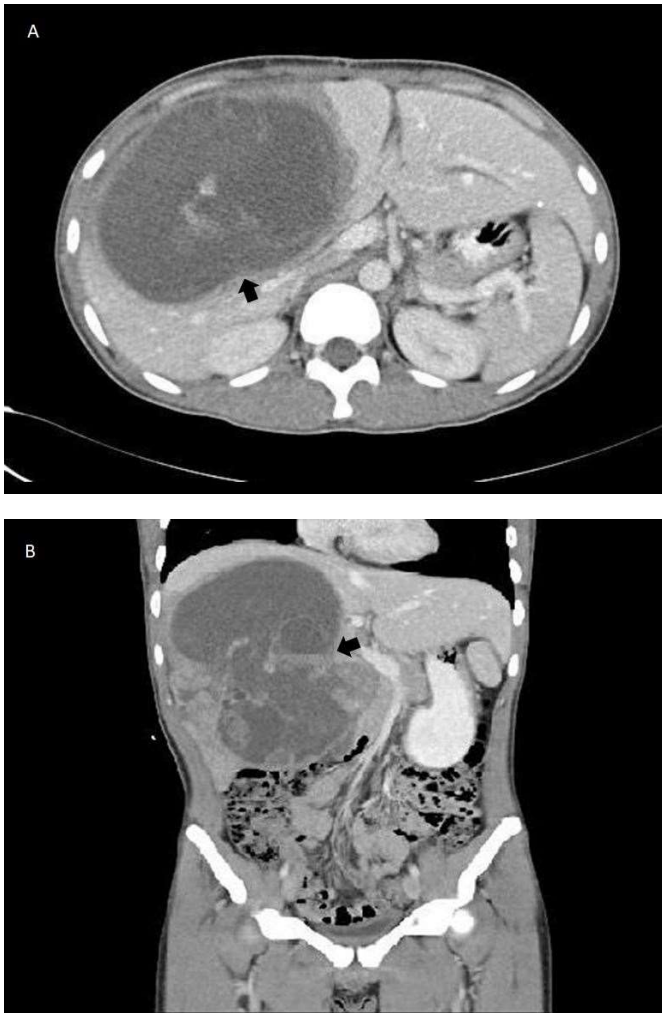

Figure 1: (A) Computed tomography scan (axial view) of the liver lesion shows a well encapsulated semi-solid mass (black arrow). (B) Computed tomography scan (coronal view) shows a suspected liver abscess (black arrow).

Macroscopic examination showed a large, well circumscribed tumor measuring 18 x 18 x 7 cm. There was presence of central necrosis and hemorrhagic solid whitish material at its periphery (Figure 2).

Figure 2: Macroscopic view of the liver lesion shows a solid with cystic components with variable degree of necrosis and haemorrhage.

Sheets of malignant infiltrations were seen within liver parenchyma which was separated from the surrounding tissue by a dense sclerotic band. There was also presence of irregular hyperchromatic and vesicular nuclei. Microscopic examination also showed eosinophilic cytoplasm and intra-cytoplasmic eosinophilic globules. These cellular components are positive for PAS and diastase. (Figure 3A). The malignant cells were positive for CD56, CD10 and desmin (Figure 3B).

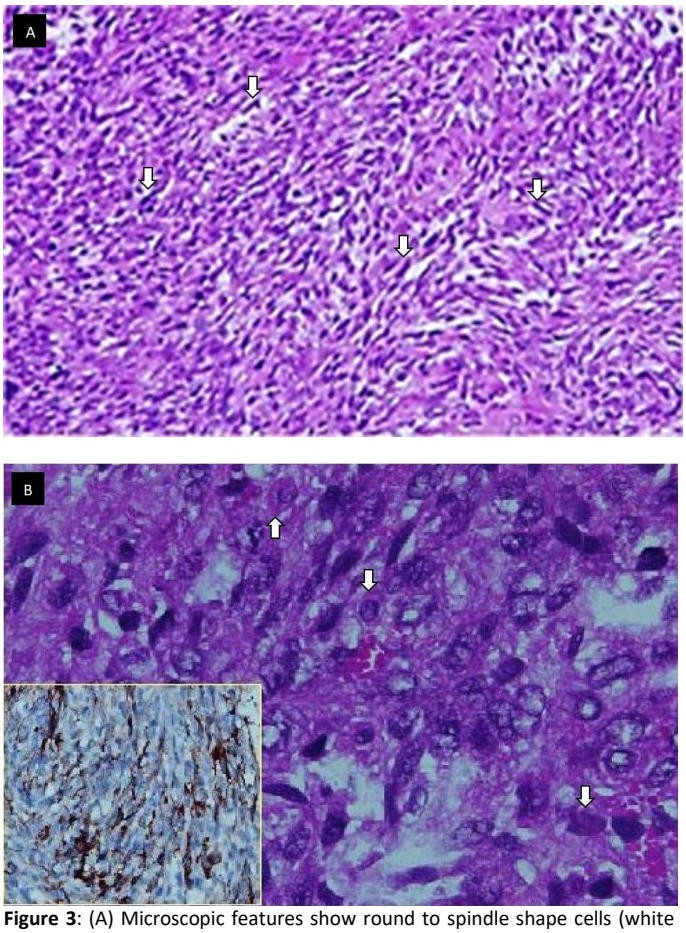

Figure 3: (A) Microscopic features show round to spindle shape cells (white arrow) with hyperchromatic to vesicular nuclei, moderate eosinophilic cytoplasm with indistinct borders. (Hematoxylin & Eosin x20). (B) Presence of intracytoplasmic eosinophilic globules which are positive for PAS (white arrow) (large box) and desmin (small box).

DISCUSSION

Liver tumours present in variable ways either asymptomatic or in advanced stage. UESL is a rare but highly malignant liver neoplasm. It tends to be a solid lesion rather than cystic. The diagnosis remains a challenge due to its non-specific symptoms with no specific and sensitive biomarkers. This condition was first described by Stocker et al (3). His research confirmed the aggressive nature of the disease with death occurring less than one year in all of his 30 subjects. However, these figures have improved drastically throughout the years. The improved technology and surgical technique have made resection feasible and safe. Curative resection has now been a realistic aim in this disease. Nevertheless, an article by Walker et al revealed only 37.5% of patients were disease free for 37 months and 2 patients for 5 years following complete surgical resection (4).

Although most presentations in pediatric cases are asymptomatic, UESL presentation in adults varies. Abdominal pain remains the most common complaint followed by a palpable mass and intermittent fever. UESL typically appears as a well-circumscribed solid mass upon computed tomography. The mass could be with or without cystic component. Though being a primary liver tumour, it rarely shows any arterial enhancement which is not characteristic. A differential of liver abscess and metastases should be suspected in such cases. The macroscopic appearance of an UESL may present as a single and large solid mass. It typically has no cystic component which is attributed by necrosis and hemorrhagic during tumour development. A presence of a capsule is not a constant feature. In microscopic examination, we could see a spindle-shaped cellular composition with eosinophilic globules. It typically stains for PAS and other immune-histochemical such as desmin, vimentin, alpha-1 antitrypsin and alpha-1 antichymotrypsin (5).

Resection remains the mainstay of treatment in these patients. An aim of a R0 resection should be the priority but it was unfortunate for our patient as he presented with a ruptured tumour mimicking an abscess. The role of adjuvant therapy is crucial in the postoperative management. Multiple articles have proven the successful role of chemotherapy in both pre and postoperative period. The recommended regime includes cisplatin, adriamaycin, and cyclophosphamide (6,7). An article by Lenze et al. has successfully compare the outcome between surgery alone and addition of adjuvant therapy which shows an improve median of 29 months survival rate in 68 patients (8). As local recurrence and metastases are common especially to the pleura, peritoneum and lung, it is highly recommended for adjuvant therapy given to improve the survival. Due to the absence of any tumour marker related to this pathology, few studies suggested a second look laparotomy for subclinical recurrences (5). However, a more conclusive report is yet to be seen. Till then, once any recurrence confirmed, a further resection should be considered after a thorough work up.

CONCLUSION

The diagnosis of UESL can pose a challenge to surgeons due to its rarity and similar presentations with other common pathology. Nevertheless, this diagnosis should be considered among the other differentials.

Conflict of interest

No conflict of interest was declared by the authors.

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