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A rare primary liver tumour case report: 53-year-old man with primary hepatic leiomyosarcoma

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Case Report

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ABSTRACT

Primary hepatic leiomyosarcoma (PHL) is a very rare primary liver tumour. These tumours usually arise from intrahepatic vascular structures, gallbladder, or ligamentum teres. The pathogenesis of this disease is still unknown. We report a 53-year-old man with complaints of intermittent right upper abdominal pain for one month, decreased appetite, nausea, weight loss, and tea-coloured urination. The patient denied any complaints of defecation pattern. On physical examination, jaundice was present in both eyes and the whole-body skin, tenderness in the right hypochondriacal region, and enlarged liver with a lumpy surface. The laboratory examination demonstrated increased transaminase enzymes and bilirubin, while hepatitis B and C were negative. The patient underwent several tests for tumour markers, such as CA 19-9 50 (<37 U/mL), AFP 1.23 (<5.81 IU/mL), and CEA 0.83 (<3 ng/mL). Ultrasound demonstrated an enlarged liver, multiple liver nodules, and cholelithiasis. Meanwhile, the abdomen's computerised tomography (CT) scan shows a solid heterogeneity image with an irregular border in segments 1 and 4b and a tumoral thrombus in the inferior vena cava. The diagnosis was confirmed by biopsy and immunohistochemistry (IHC); vimentin and smooth muscle actin (SMA) results were positive, while CD 34, CD 117, and cytokeratin were negative. Our patient was diagnosed with PHL stage IVa (T4N2M0). PHL is a particularly rare tumour with a poor prognosis. The patient died after one month of diagnosis. Diagnosis of PHL is challenging. It was based on clinical features, physical examinations, laboratory examinations, and other supporting investigations.

Leiomyosarcoma hati primer (PHL) adalah tumor primer yang sangat langka. Tumor ini muncul dari struktur vaskular intrahepatic, empedu, atau ligamentum teres. Patogenesis penyakit ini masih belum diketahui. Kami melaporkan kasus pria 53 tahun dengan keluhan nyeri perut kanan atas selama satu bulan, penurunan nafsu makan serta berat badan, mual, dan warna urin seperti teh. Pasien menyangkal adanya keluhan pola buang air besarnya. Pada pemeriksaan fisik, sklera iterik ditemukan pada kedua mata, kuning di seluruh tubuh, nyeri tekan regio hypochondriaca kanan, dan pembesaran hati disertai permukaan yang tidak rata. Dari pemeriksaan laboratorium, terdapat peningkatan enzim transaminase, bilirubin, sementara viral marker hepatitis B dan C negatif. Pemeriksaan tumor marker juga dilakukan: CA 19-9 50 (<37 U/mL), AFP 1.23 (<5.81 IU/mL), CEA 0.83 (<3 ng/mL). Pemeriksaan dengan ultrasound menunjukan pembesaran hati,

nodul hati multipel, dan cholelithiasis. Sementara itu, berdasarkan pemeriksaan computerised tomography (CT) scan abdomen, terdapat gambaran heterodensity padat dengan batas tidak tegas di segmen 1 dan 4b, serta trombus tumor di vena cava inferior. Diagnosis dikonfirmasi melalui biopsi dan imunohistokimia (IHC), hasilnya didapatkan vimentin dan SMA positif, sementara CD 34, CD 117, dan cytokeratin negatif. Pasien kami didiagnosis sebagai PHL stadium IVa (T4N2M0). PHL merupakan tumor langka dan memiliki prognosis yang buruk. Pasien meninggal satu bulan pasca diagnosis. Penegakan diagnosis PHL sangat menantang perlu pertimbangan dari gejala klinis, pemeriksaan fisik, pemeriksaan lab, dan penunjang lainnya.

INTRODUCTION

Mesenchymal liver cancer is a rare case. These cancers include hemangioma, angiosarcoma, epithelioid hemangioendothelioma, undifferentiated embryonal sarcoma, fibrosarcoma, and leiomyosarcoma.[1] The primary hepatic leiomyosarcoma (PHL) prevalence is 0.2-2% of primary liver malignancies. A total of 76 newly published cases. PHL usually arises from intrahepatic vascular structures, gallbladder, or ligamentum teres. The pathogenesis of this disease is still unknown due to the rarity of its cases.[2] The aetiology of PHL is still unclear; several reports link this case to the use of contrast agents, such as Thorotrast, acquired immunodeficiency syndrome (AIDS), Epstein-Barr virus infection, and hepatitis C.[3]

CASE DESCRIPTION

In this case, a 53-year-old male patient presented with jaundice in both eyes and whole-body skin, as well as tea-coloured urine. Meanwhile, the complaints of pale stools like putty were denied. In addition, the patient also complained of upper right abdominal pain that is felt continuously, like being stabbed. These symptoms have been felt in the last two months before hospital admission.

Laboratory findings demonstrated an increase in total bilirubin 20.37 (reference 0.00-0.30 mg/dl), direct bilirubin 15.03 (normal range 0.00-0.30 ng/dL), indirect bilirubin 5.34 (normal range 0.00-0.70 mg/dl), gamma GT 441 (normal range <55 U/L), alkali phosphatase 153 (normal range 53-128 U/L), SGOT levels of 54 (normal range <31 U/I) , and SGPT 34 (normal range <31 U/I). The patient had no hepatitis B and C comorbidities. The patient underwent several examinations, including tumour marker CA 19-9 50 U/mL (normal range <37 U/mL), alpha-fetoprotein (AFP) 1.23 ng/mL (normal range <5.81 (IU/mL), carcinoembryonic antigen (CEA) 0.83 (normal range <3 ng/mL), and urinalysis bilirubin +1 (normal). CT-scan findings were irregular solid heterogeneity and irregular border in segments 1 and 4b of the liver. Post-contrast showed slight contrast enhancement narrowing bilateral intrahepatic bile duct (IHBD) and tumoural thrombus in the inferior vena cava, leading to cholangiocarcinoma appearance (Figure 1).

During hospitalisation, an analgesic (metamizole) was administered to control

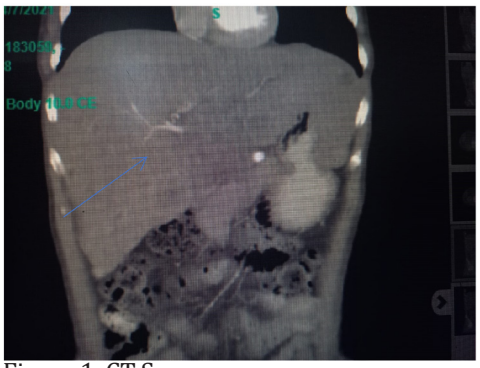

Figure 1. CT-Scan
Blue sign demonstrated solid heterogeneity irregular in segment 4b

pain, and a hepatoprotector (curcuma) was administered as a strong anti-oxidant and anti-inflammatory for the liver.

The patient was diagnosed with PHL. The reason is that the IHC examination revealed positive results in smooth muscle actin (SMA) and vimentin. Then, CD 34, CD 117, and cytokeratin were not detected (Figure 2). It is a resection contraindication because nodules are multiple and spread into the surrounding tissue. Thereby, chemotherapy is recommended. However, the patient rejected chemotherapy. In the end, the patient died after a month of diagnosis.

DISCUSSION

The prevalence of PHL is rare, with approximately only 0.2-2% of primary liver malignancies.[2] The patient has impaired hepatic transport of bilirubin, as a result, presented jaundice of both eyes and the whole-body skin, as well as tea-coloured urine. Laboratory findings revealed increased total bilirubin, direct and indirect bilirubin, gamma GT, alkaline phosphatase, CA 19-9, normal CEA, and AFP tumour markers. In some cases, the CEA is normal, and CA 19-9 increases. Although no literature demonstrated a relationship between the two.[4,5] The absence of specific tumour markers also makes diagnosing this disease difficult.[5]

CT-scan examination revealed irregular solid heterogeneity and border in segments 1 and 4b of the liver. Furthermore, post-contrast showed slight contrast enhancement narrowing the bilateral IHBD and tumoral thrombus in the inferior vena cava. PHL's CT-scan finding is usually heterogeneous and sometimes accompanied by imaging resembling a liver cyst or liver abscess.3 These findings are consistent with a previous case report.[5]

The primary diagnosis is established by eliminating other major organ causes, such as the genitourinary, retroperitoneal, and gastrointestinal tract.[4] In this case, no malignancy was found elsewhere. The current study diagnosis is based on the results of an abdominal CT scan which showed a problem with the liver, and the results of a Thorax X-ray and Bone Survey showed no abnormalities.

Several measures for managing PHL have been reported, such as liver resection surgery in the form of lobectomy, hemihepatectomy, and trisegmentectomy. Laparoscopic right hepatectomy and transcatheter arterial chemoembolisation (TACE) are reported to be

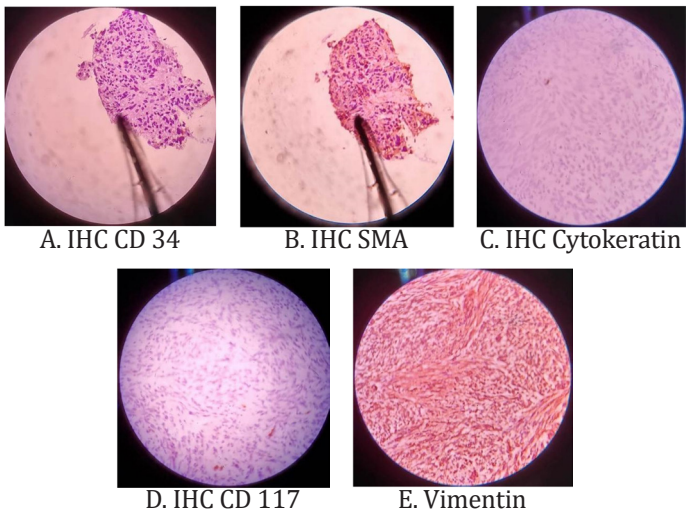

Figure 2. Immunohistochemistry image: A: CD 34; B: SMA; C: Cytokeratin; D: CD 117; E: Vimentin
IHC show negative CD 34, positive SMA, negative cytokeratin, negative CD 117, positive vimentin

successful in the early stages, although more studies are needed.[6–9] Some of these measures also consider residual liver function, tumour margins, and vascularity. Liver transplantation, chemotherapy, and radiotherapy benefits for PHL remain unclear.[8] Long-term survival is possible after complete tumour resection in a preselected population with early-stage disease.[9]

In addition, other management, such as chemotherapy, can be considered. The European Society for Medical Oncology (ESMO) and the National Comprehensive Cancer Network (NCCN) recommend anthracycline or combination with ifosfamide for sarcoma-type cancer.[10] Several chemotherapy regimens have been reported, such as (folinic acid, fluorouracil, irinotecan, bevacizumab), (mitoxantrone, cisplatin, and fluorouracil),and (ifosfamide and mesna).[11-13] Adjuvant chemotherapy of doxorubicin and ifosfamide has shown slow progress and prolonged life after complete resection.[3]

CONCLUSION

PHL is a particularly rare tumour with a poor prognosis. The patient was lost to follow-up and died after a month of diagnosis. It is important to know that liver malignancy is not only grown by usually aetiology but also from other tissue, such as smooth muscle cells.

CONFLICT OF INTEREST

None declare.

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