TRAGICALLY YOUNG : THE 26-YEAR-OLD KADAZAN FEMALE WHO LOST HER BATTLE WITH HEPATITIS-NEGATIVE HEPATOCELLULAR CARCINOMA AND BETA-THALASSEMIA MAJOR.

Munirah Abdul Malek[1], Thanesh Kumar[1], Jonathan Seak Chen Ken[1], Dayang Corieza Febriany[2]
Department of Hepatobiliary Surgery, Queen Elizabeth I Hospital, Kota Kinabalu, Sabah, Malaysia[1]
Department of Radiology, Universiti Malaysia Sabah, Sabah, Malaysia[2]
NMRR ID : 22-01008-ZYC

AIM

To highlight the importance of considering beta-thalassemia as a potential risk factor for hepatocelllular carcinoma despote being hepatitis negative.

TOP 5 TAKE-AWAYS

1. *Beta-thalassemia major patients need iron chelation therapy due to iron overload.*
2. *Since 2006, Malaysia has offered free chelation therapy to improve medical care.*
3. *Poor iron chelation adherence remains a challenge, and iron overload causes cancer.*
4. *Abdominal ultrasound is the best HCC screening tool.*
5. *Beta-thalassemia may be linked to HCC.*

CASE PRESENTATION

- A 26-year-old woman presented with abdominal pain and jaundice for 2 weeks.
- She had beta-thalassemia major since 6 months old and received 2 weekly blood transfusions.
- Her MRI T2 showed severe liver iron concentration despite her regular iron chelation therapy, indicating poor compliance. Clinically she had hepatosplenomegaly.
- Blood test showed anemia and obstructive jaundice, with elevated AFP level (3725.1 U/mL). Hepatitis B & C tests were negative, and she did not consume alcohol.
- Hepatobiliary USG detected multiple liver lesions. CECT TAP(figure 1) showed extensive liver and hilar lesions compatible with malignancy and lung metastasis.
- Percutaneous liver biopsy confirmed HCC. She died 1 week later due to disease progression.

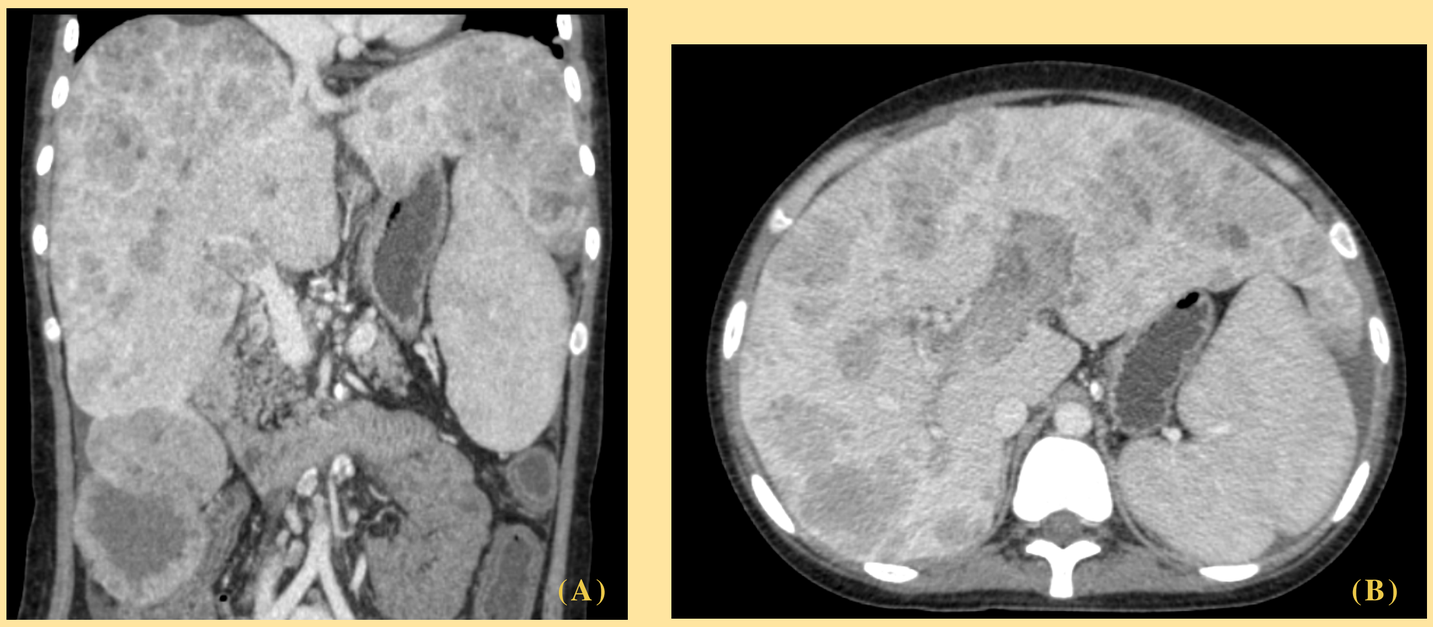

Figure 1. CECT Abdomen at the level of the liver in (A) coronal and (B) axial projections showing multiple liver and hilar lesions.

DISCUSSION

Despite limited local data, the incidence of HCC in Greece and Italy is 2.3 and 1% respectively. Malaysia's highest rate of beta-thalassemia major is in Sabah. Regular blood transfusions necessitate iron chelation therapy because they increase iron overload. In 2006, the Malaysian government provided free chelation therapy, which increased survival rates. Chelation compliance is difficult. High iron levels promote reactive oxygen species formation, which impairs protein synthesis and disrupts DNA, causing mutations leading to HCC. Abdominal USG is the best screening tool.

CONCLUSION

Physicians should be aware of the potential link between thalassemia and HCC to promt early evaluation of patients with beta-thalassemia who present with elevated AFP level and baseline ultrasound imaging, allowing for prompt intervention in the event of HCC development.

REFERENCES

1. Papadopoulos N, Koskinas J. Hepatocellular carcinoma in transfusion dependent thalassemia patients: A review from a clinical perspective. *Hepatoma Research*. 2021;7:7-75. doi:10.20517/2394-5079.2021.118
2. Mohd Ibrahim, H. ED., (2019). Malaysia Thalassemia Registry Report 2018. 1st ed. Medical Development Division, Ministry of Health, Malaysia.
3. Moukhadder HM, Halawi R, Maria, Cappellini D, Taher AT. Hepatocellular Carcinoma as an Emerging Morbidity in the Thalassemia Syndromes: A Comprehensive Review. *Cancer Journal*. doi:10.1002/cncr.30462
4. Mohamed R, Abdul Rahman AH, Masra F, Abdul Latiff Z. Barriers to adherence to iron chelation therapy among adolescent with transfusion dependent thalassemia. *Frontiers in Pediatrics*. 2022;10. doi:10.3389/fped.2022.951947
5. De Sanctis V, Soliman AT, Daar S, et al. A Concise Review on the Frequency, Major Risk Factors and Surveillance of Hepatocellular Carcinoma (HCC) in β-Thalassemias: Past, Present and Future Perspectives and the ICET-A Experience. *Mediterranean Journal of Hematology and Infectious Disease*. 2020;12:2020006. doi:10.4084/MJHID.2020.006