Received: 17 November 2020 | Revised: 8 June 2021 | Accepted: 21 June 2021

DOI: 10.1002/ccr3.4600

CLINICAL IMAGE

Clinical Case Reports Open Access WILEY

Large abdominal purpura of neonatal retroperitoneal kaposiform hemangioendothelioma

Ryo Takemura | **Yoshiro Wada**

Rinku General Medical Center, Izumisano, Japan

Correspondence
Yoshiro Wada, Pediatrics, Rinku General Medical Center, Izumisano, 598-8577, Japan.
Email: y-wada@rgmc.izumisano.osaka.jp

Funding information
None

Abstract

Large abdominal purpuras may be caused by retroperitoneal kaposiform hemangioendothelioma with consumptive coagulopathy. Clinicians should perform serial ultrasonography studies to detect the sings of tumor until the final diagnosis is confirmed.

KEYWORDS

kaposiform hemangioendothelioma, Kasabach-Merritt phenomenon, purpura

1 | CASE DESCRIPTION

A 7-day-old female infant with large purpuras on the left abdomen (Figure 1) was referred. Abdominal ultrasonography revealed no abnormal findings (Figure 2). The platelet count was 5.6×10^4/μl, and the D-dimer was 22.4 μg/ml. However, other parameters were hemoglobin (18.9 g/dl), PT-INR (1.14), fibrinogen (151 mg/dl), and bleeding time (5.50 min). No data raised suspicions of an infection. The platelet antibody screening for alloimmune thrombocytopenia was reported negative on day 22. On the same day, the infant was diagnosed with Kasabach-Merritt phenomenon (KMP) caused by retroperitoneal kaposiform hemangioendothelioma (KHE; Figures 3 and 4). As steroid, beta-blockers, and frequent blood transfusion therapy were not effective, the infant was transferred to a tertiary referral hospital and finally cured with vincristine and radiation.[1]

Neonatal retroperitoneal KHE is rare and has a poor prognosis [2]; hence, early detection is crucial. In this case, the first presentation was limited to purpura only. If ultrasonography was repeated during differential diagnosis, retroperitoneal KHE would have been detected before the occurrence of severe bleeding due to KMP.

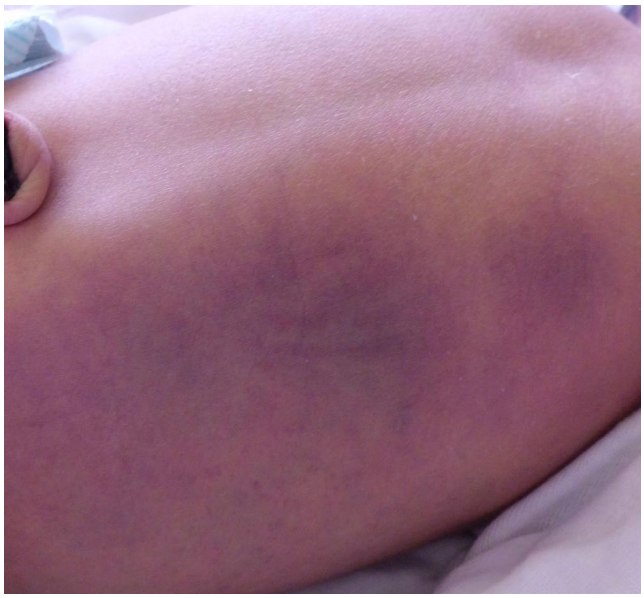

FIGURE 1 Localized large purpura over the left abdomen on day 7 of life

Sirolimus, an mTOR inhibitor, would have prevented the infant from the side effects of radiation exposure. However, it was not available in Japan at that time.

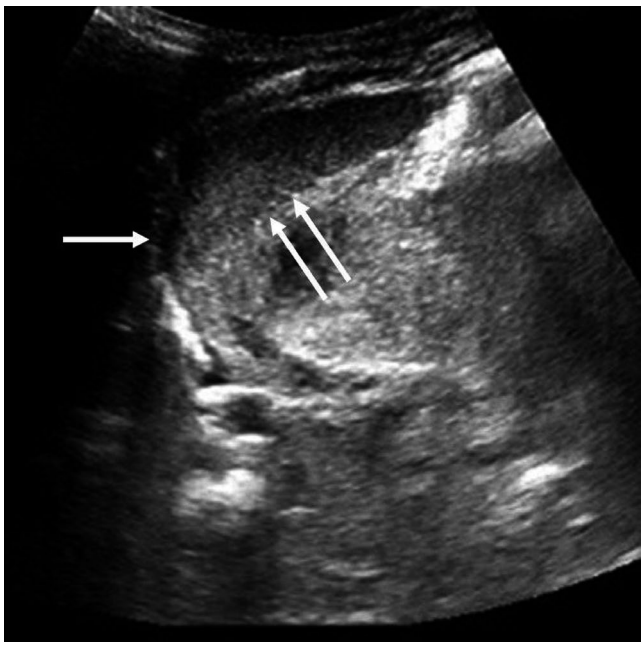

FIGURE 2 Abdominal ultrasonography. No tumor was observed in the subcutaneous area, retroperitoneum, and abdominal cavity. Diaphragm, arrow; spleen, double arrows

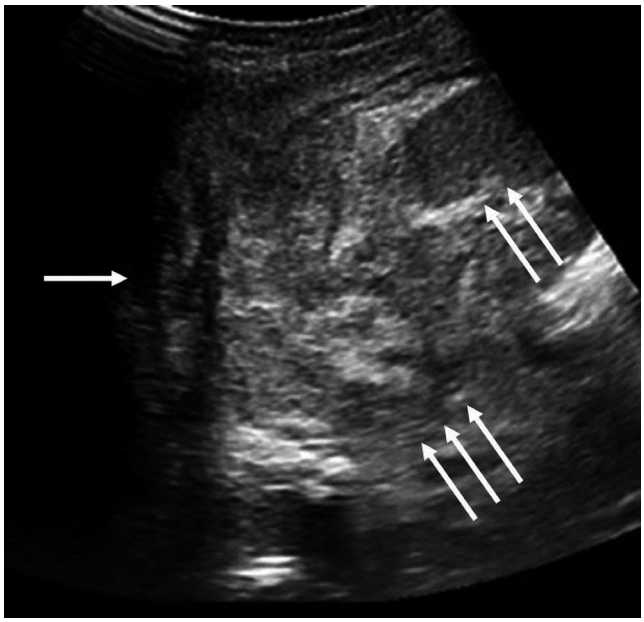

FIGURE 3 Abdominal ultrasonography of the kaposiform hemangioendothelioma. A heterogeneous, irregular-shaped tumor (triple arrows) was observed between the diaphragm (arrow) and spleen (double arrows)

ACKNOWLEDGMENTS

The authors thank the medical team at the tertiary referral hospital, Osaka Women's and Children's Hospital, for taking care of the infant. The authors and editors of Reference [1] are specially thanked for allowing the submission of this short report to the journal. Published with written consent of the patient.

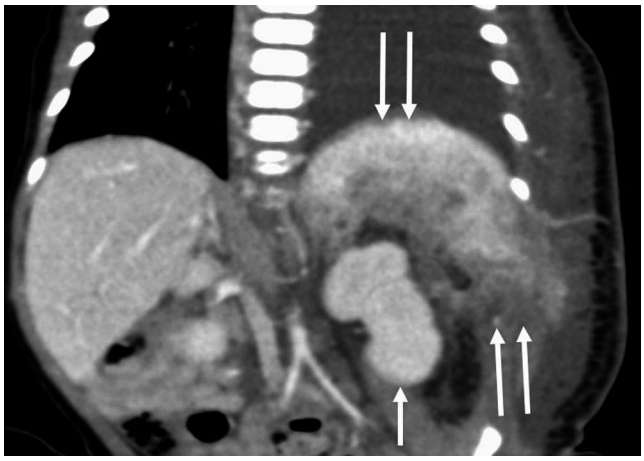

FIGURE 4 Contrast-enhanced computed tomography image of kaposiform hemangioendothelioma. A large tumor to the left of the retroperitoneum (double arrows) displacing the kidney (single arrow) downward. Ipsilateral side subcutaneous thickening and hemothorax were present

CONFLICTS OF INTEREST

The authors have no conflicts of interest to declare.

AUTHOR CONTRIBUTIONS

Both authors made substantial contributions to the preparation of this manuscript and approved the final version for submission. RT drafted the initial version of the manuscript. WY performed the literature search and revised the manuscript for critically important intellectual content.

INFORMED CONSENT

Written informed consent has been obtained from the parent for the publication of this manuscript as a Clinical Image.

DATA AVAILABILITY STATEMENT

Data sharing not applicable to this article as no datasets were generated or analyzed during the current study.

REFERENCES

1. Kodama T, Soh S, Tazuke Y, et al. A case of vascular tumor associated with Kasabach-Merritt syndrome in the neonatal period successfully treated with radiation therapy. *Osaka Women's and Children's Hospital Journal*. 2014;30:87-92. (In Japanese).
2. Chinello M, Carlo DD, Olivieri F, et al. Successful management of kaposiform hemangioendothelioma with long-term sirolimus treatment. *Mediterr J Hematol Infect Dis*. 2018;10:e2018043.

How to cite this article: Takemura R, Wada Y. Large abdominal purpura of neonatal retroperitoneal kaposiform hemangioendothelioma. *Clin Case Rep*. 2021;9:e04600. https://doi.org/10.1002/ccr3.4600