Received: 28 March 2018 | Revised: 22 August 2018 | Accepted: 20 September 2018

DOI: 10.1002/ccr3.1857

CLINICAL IMAGE

WILEY Clinical Case Reports Open Access

# Splenic mass in a case of *CALR*-mutated essential thrombocythemia

**Shinsaku Imashuku**[1] | **Shinichi Kotani**[2] | **Junsuke Hinami**[3] | **Keisuke Nishimura**[4]

[1]Department of Laboratory Medicine, Uji-Tokushukai Medical Center, Uji, Kyoto, Japan

[2]Division of Hematology, Uji-Tokushukai Medical Center, Uji, Kyoto, Japan

[3]Department of Surgery, Uji-Tokushukai Medical Center, Uji, Kyoto, Japan

[4]Division of Pathology, Uji-Tokushukai Medical Center, Uji, Kyoto, Japan

**Correspondence**
Shinsaku Imashuku, Department of Laboratory Medicine, Uji-Tokushukai Medical Center, Uji, Kyoto, Japan.
Email: shinim95@mbox.kyoto-inet.or.jp

**Key Clinical Message**

We report here an intrasplenic large mass in an elderly case of essential thrombocythemia (ET)-myelofibrosis. Laparoscopic splenectomy revealed extramedullary hematopoiesis (EMH) and a type 1 *CALR* gene mutation (CALR-c.1092_1143del52) in the splenic mass. It remains to be determined if *CALR*-mutated ET has an increased tendency to develop mass-forming EMH.

**KEYWORDS**

calreticulin, essential thrombocythemia, extramedullary hematopoiesis, myelofibrosis, splenic mass

An 86-year-old Japanese female had splenomegaly with a history of elevated platelet counts (>500 000/μL) for more than 10 years. Laboratory tests showed a WBC of 30 600/μL and a platelet count of 1 031 000/μL, in association with high serum soluble IL2-receptor (1001 U/mL), low serum IgG (712 mg/dL), and low complement CH50 levels (5 U/mL). Essential thrombocythemia (ET)[1] was suspected, but *JAK2 V617F* gene mutation was negative. An abdominal CT revealed a huge mass within the spleen (Figure 1A). Bone marrow revealed myelofibrosis grade 1-2 (Figure 1B). Laparoscopic splenectomy showed that the central round tumorous part of the spleen, which measured 6.2 cm × 6.3 cm, was clearly differentiated from the non-tumorous part on the cut surface (Figure 1C). Histopathology revealed trilineage extramedullary hematopoiesis (EMH), but with particularly increased numbers of megakaryocytes with nuclear atypia in the tumorous part (Figure 1D). In the splenic mass, we found a type 1 *CALR* gene mutation (CALR-c.1092_1143del52), which is a major abnormal type of *CALR*. Eventually, the patient was diagnosed as having a splenic mass of EMH, as consequence of *CALR*-mutated ET. This case was unexpected and uncharacteristic because the spleen is not among the organs involved with mass-forming EMH.[2]

This is an open access article under the terms of the Creative Commons Attribution-NonCommercial-NoDerivs License, which permits use and distribution in any medium, provided the original work is properly cited, the use is non-commercial and no modifications or adaptations are made.
© 2018 The Authors. *Clinical Case Reports* published by John Wiley & Sons Ltd.

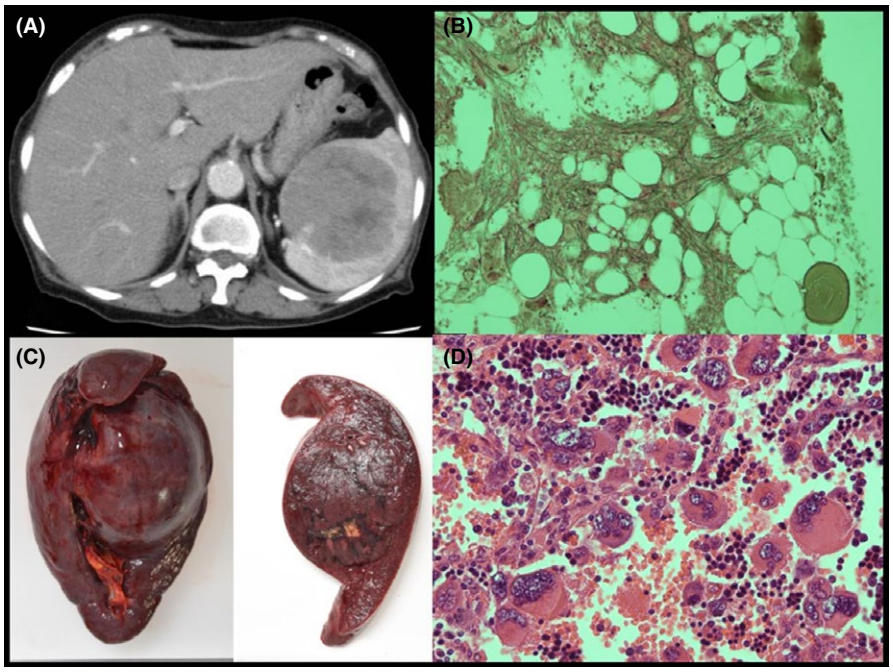


**FIGURE 1** A, Computed tomography (CT) scan of the abdomen showing the splenic mass. B, Bone marrow biopsy revealing myelofibrosis grade 1-2 (silver impregnation stain; original magnification ×400). C, Gross appearance and the cut surface of the resected spleen showing clear delineation between the tumorous and non-tumorous parts. D, Histopathology of the tumorous part of the spleen showing markedly increased numbers of megakaryocytes with nuclear atypia (HE stain; original magnification ×400)

## CONFLICT OF INTEREST

The authors have no conflict of interests to declare.

## AUTHOR CONTRIBUTION

SI and SK: treated the patient, SI: engaged in molecular diagnosis and wrote the manuscript, SK: performed bone marrow biopsy, JH: performed laparoscopic splenectomy, KN: performed hispathological diagnosis. All authors read and confirmed the final version of manuscript.

## ORCID

*Shinsaku Imashuku* http://orcid.org/0000-0001-9795-0819

## REFERENCES

1. Rumi E, Pietra D, Ferretti V, et al. JAK2 or CALR mutation status defines subtypes of essential thrombocythemia with substantially different clinical course and outcomes. *Blood*. 2014;123(10):1544-1551.
2. Policarpio-Nicolas ML, Bregman SG, Ihsan M, Atkins KA. Mass-forming extramedullary hematopoiesis diagnosed by fine-needle aspiration cytology. *Diagn Cytopathol*. 2006;34(12):807-811.

**How to cite this article:** Imashuku S, Kotani S, Hinami J, Nishimura K. Splenic mass in a case of *CALR*-mutated essential thrombocythemia. *Clin Case Rep*. 2018;6:2291–2292. https://doi.org/10.1002/ccr3.1857