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CASE REPORT

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Pelvic extramedullary hematopoiesis with life-threatening hemorrhage: a case report

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Abstract

Background Thalassemia is an inherited blood disorder characterized by defective hemoglobin production, leading to chronic anemia and the necessity for lifelong blood transfusions. Extramedullary hematopoiesis is a compensatory mechanism in which blood-producing tissue forms outside the bone marrow. The occurrence of extramedullary hematopoiesis as a pelvic mass is rare.

Case Presentation We report the case of a 44-year-old woman with β-thalassemia major who was found to have a pelvic fat-containing mass incidentally on imaging. A diagnostic laparotomy revealed a highly vascular mass. Complete resection was not feasible due to significant hemorrhage, necessitating postoperative embolization for hemostasis. Histopathological examination confirmed the diagnosis of extramedullary hematopoiesis. Postoperatively, the patient developed severe anemia, requiring frequent transfusions. To reduce transfusion dependency, splenic artery embolization was performed.

Conclusion This case underscores the rare presentation of extramedullary hematopoiesis as a pelvic mass. Accurate diagnosis is critical, as surgical removal can result in severe hemorrhage and worsening anemia. Splenic artery embolization provides a valuable therapeutic approach to managing transfusion burden in such cases.

Keywords Extramedullary hematopoiesis, Thalassemia major, Pelvic mass, Chronic anemia, Blood transfusion, Splenic artery embolization, Hemorrhagic shock

Background

Thalassemia major is a severe, inherited blood disorder characterized by mutations in the *HBB* gene that lead to impaired hemoglobin production. This results in chronic anemia, which is typically severe and requires regular blood transfusions to maintain adequate hemoglobin levels [1]. A significant complication of thalassemia major is the development of extramedullary hematopoiesis, typically in the spleen, liver, or lymph nodes [2]. This occurs as a compensatory mechanism due to ineffective hematopoiesis in the bone marrow. The occurrence of extramedullary hematopoiesis in the pelvis is rare [3, 4]. These ectopic hematopoietic tissues are highly vascular and present significant therapeutic challenges [5].

This case report describes the inadvertent removal of an extramedullary hematopoiesis mass in the pelvis, which can lead to life-threatening hemorrhage and severe anemia. This study also highlights the critical role of advanced interventional techniques in managing these complications.

Case presentation

A 44-year-old female with β-thalassemia major (homozygous for *HBB*:c.118C > T), diagnosed at age 30, has not undergone regular follow-up or blood transfusions since

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the initial diagnosis. She presented with persistent left hip pain following a fall, prompting further investigation.

MRI revealed an incidental pelvic mass with well-defined margins extending toward the abdominal cavity. The mass measured 13×15×10 cm and exhibited heterogeneous signal intensity, with intermediate signal intensity on T1-weighted images, low signal intensity on T2-weighted images, and areas of high signal intensity consistent with fat. The mass exerted significant pressure, displacing adjacent bowel loops without evidence of invasion into nearby structures (Fig. 1). The provisional diagnosis for this fat-containing lesion included an ovarian neoplasm or a pedunculated uterine lipoleiomyoma.

A comprehensive laboratory evaluation was performed prior to surgery (Table 1). A diagnostic laparotomy revealed a large pelvic mass with associated hemorrhage. The mass was initially suspected to be an ovarian cyst, and during the procedure, it was accessed and suctioned. However, a massive hemorrhage (estimated at 11 L) occurred during the procedure, prompting early termination and incomplete resection. The patient was transferred to the intensive care unit for management of hemorrhagic shock and remained there for 7 days. Selective embolization of the pelvic mass was performed to control the hemorrhage (Fig. 2).

Histopathological examination of the resected tissue confirmed the diagnosis of extramedullary hematopoiesis, consisting of a mixture of hematopoietic cells, megakaryocytes, and scattered mature adipocytes. Immunohistochemistry for factor VII highlighted the megakaryocytes, whereas CD20 and CD3 staining identified B and T lymphocytes, respectively.

During the hospital stay, a CT scan of the abdomen and pelvis was performed, revealing features consistent with extramedullary hematopoiesis, including splenomegaly (craniocaudal diameter of 18 cm), paravertebral soft tissue masses, and rib expansions with associated soft tissue lesions (Fig. 3).

The patient was discharged in stable condition after 27 days of hospitalization. Following discharge, she developed severe anemia, with hemoglobin levels as low as 4.5 g/dL, necessitating frequent blood transfusions. To reduce transfusion dependency, splenic artery embolization was performed (Fig. 4). Hemoglobin levels subsequently improved and stabilized between 8 and 10 g/dL, with extended intervals between transfusions (Fig. 5).

Discussion

This case presents a rare instance of extramedullary hematopoiesis in a middle-aged woman with β-thalassemia major. While extramedullary hematopoiesis is a well-known complication of thalassemia, its occurrence as a pelvic mass is an unusual presentation [3, 4].

Extramedullary hematopoiesis typically occurs in the liver, spleen, and paraspinal regions, reflecting their hematopoietic roles during fetal development. However, extramedullary hematopoiesis can also occur in a wide range of locations [2]. Involvement of the bones is common, with medullary

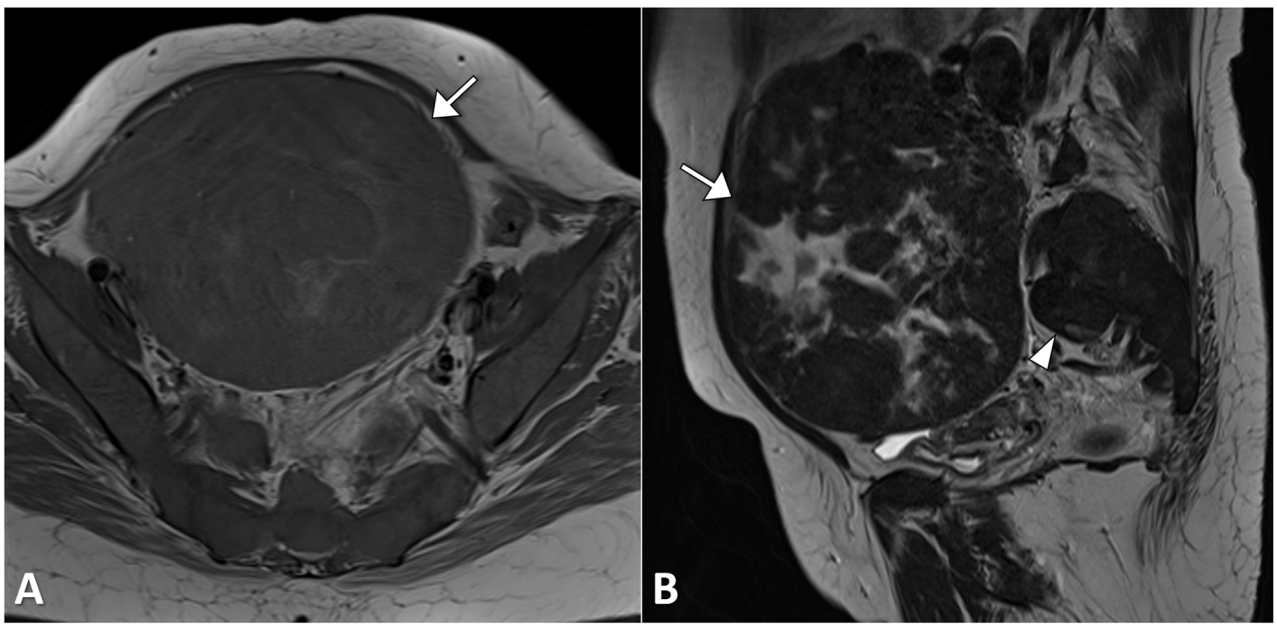

Fig. 1 MRI of the pelvis. Axial T1-weighted (**A**) and sagittal T2-weighted (**B**) MR images showing an incidental finding of a well-defined, large pelvic mass (arrow) with heterogeneous signal intensity interspersed with areas of fat. The mass has a significant effect on the displacement of adjacent bowel loops. A presacral mass of similar signal intensity is also noted (arrowhead)

Table 1 Preoperative baseline laboratory results

Category	Test	Result	Unit	Reference Range
Hematology	White Blood Cell Count	4.74	$\times 10^9$/L	3.6–9.6
	Red Blood Cell Count	4.00	$\times 10^{12}$/L	3.9–5.2
	Hemoglobin	9.0	g/dL	12.0–14.5
	Hematocrit	29.2	%	33–45
	Mean Cell Volume	73.1	fL	80.0–97.0
	Mean Cell Hemoglobin	22.5	pg	27.0–33.0
	Mean Cell Hemoglobin Concentration	30.7	g/dL	30.0–37.0
	Platelets	136	$\times 10^9$/L	150.0–400.0
	Red Cell Distribution Width	25.4	fL	11.6–13.7
Coagulation	Prothrombin Time	14.00	s	10.7–13.9
	International Normalized Ratio	1.24	-	0.61–1.17
	Activated Partial Thromboplastin Time	30.30	s	25–43
	Fibrinogen	225.80	mg/dL	217.0–496.0
	Thrombin Time	14.70	s	12.9–17.3
	D-Dimer	1.26	µg/mL	0–0.5
Biochemistry	Sodium	134	mmol/L	128–148
	Potassium	4.4	mmol/L	3.5–5.1
	Chloride	99	mmol/L	99–109
	Bicarbonate	26	mmol/L	20–31
	Ferritin	1888.3	µg/L	10–291
	C-Reactive Protein	2.81	mg/L	< 5.0
Tumor Markers	Cancer Antigen–125	Negative	U/mL	Negative
	Alpha-fetoprotein	Negative	ng/mL	Negative
	Human Chorionic Gonadotropin	Negative	mIU/mL	Negative

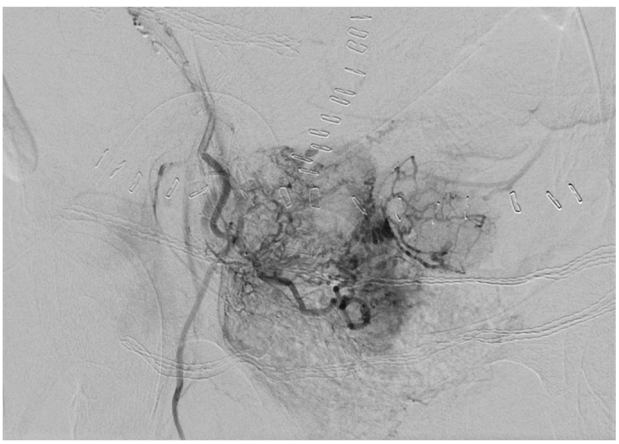

Fig. 2 Pelvic mass embolization. Selective angiography revealed a hypervascular pelvic mass with multiple feeder vessels arising from the right and left internal iliac arteries. Embolization of the pelvic mass was performed by injecting polyvinyl alcohol particles into these feeder vessels

expansion leading to cortical thinning, diploic widening, and the characteristic "hair-on-end" appearance of the skull. Manifestations near bones, such as periosteal reactions and rib or paraspinal soft tissue masses, are also common [6]. In the abdomen, it frequently presents as hepatosplenomegaly, although focal mass-like lesions in these organs can develop [3, 7]. Other unusual abdominal locations include areas around the kidneys, mesentery, and presacral region [2, 6]. In rare cases, extramedullary hematopoiesis involves the nasopharynx, paranasal sinuses, and lacrimal fossae [8].

The differential diagnosis of pelvic tumors containing fat is broad and includes several distinct pathologies. The most common ovarian neoplasm associated with fat is teratoma, which is characterized by macroscopic fat. Other tumors that may present with fat include uterine lipoleiomyomas, peritoneal loose bodies, and primary lipomatous tumors such as lipomas and liposarcomas. Extraperitoneal and extra-adrenal masses, such as myelolipomas, may also exhibit fat content. Examples of pelvic masses containing microscopic fat include immature ovarian teratomas and steroid cell ovarian neoplasms [9].

The diagnosis of extramedullary hematopoiesis poses significant challenges when it occurs in atypical locations, as it can easily mimic benign and malignant neoplasms. Therefore, a high index of suspicion is essential for an accurate diagnosis. In this context, ^{99m}Tc-sulfur colloid scintigraphy, a noninvasive imaging modality

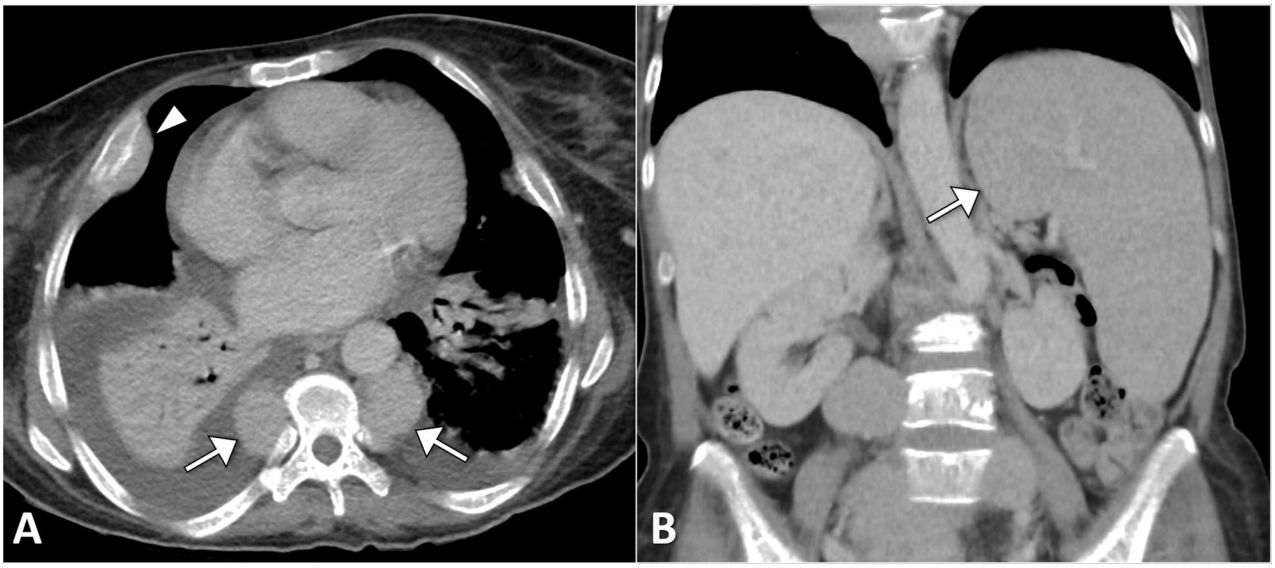

Fig. 3 CT scans of the abdomen and pelvis. Axial (**A**) and coronal (**B**) images of a postoperative contrast-enhanced CT scan showing characteristic features of extramedullary hematopoiesis, including paravertebral soft tissue masses (arrows in A), rib expansions with associated soft tissue lesions (arrowheads), and splenomegaly (arrow in B)

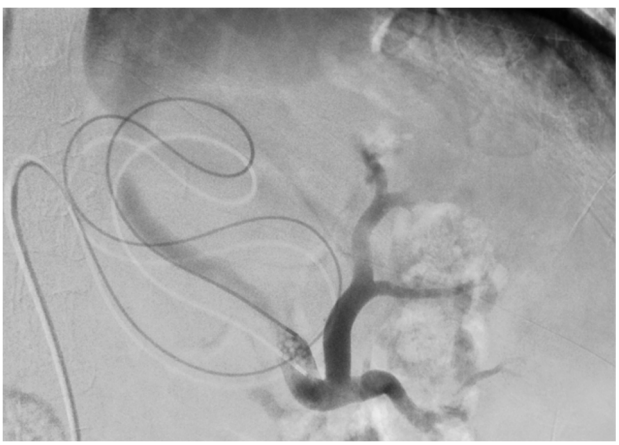

Fig. 4 Splenic artery embolization. Selective angiography and embolization of polyvinyl alcohol particles into the splenic artery to reduce splenic perfusion

targeting the reticuloendothelial system, has been widely used to evaluate bone marrow activity and diagnose extramedullary hematopoiesis [10]. FDG PET/CT is also useful in patients with known malignancies, as extramedullary hematopoiesis typically shows low metabolic activity, distinguishing it from more active tumors [11].

Management strategies for extramedullary hematopoiesis include several approaches [12]. Conservative management with frequent blood transfusions can help suppress extramedullary tissue growth [13]. Pharmacologic therapy with hydroxyurea has been shown to reduce the size of extramedullary hematopoietic masses by inhibiting abnormal blood cell production [14]. Radiation therapy is effective because of the radiosensitivity of hematopoietic tissue [15]. Surgical intervention is generally reserved for surgical decompression or when other approaches have failed.

The present case underscores the limitations of surgical resection in the management of extramedullary hematopoiesis. The procedure was complicated by massive hemorrhage and incomplete resection. As an alternative hematopoietic site, extramedullary hematopoiesis can exacerbate anemia and may require frequent blood transfusions if removed. In such cases, splenic artery embolization represents a potential therapeutic approach. By reducing splenic blood flow, splenic artery embolization induces ischemia and a reduction in the spleen volume, which minimizes red blood cell destruction and helps reduce the transfusion burden [16].

Conclusion

In conclusion, this case highlights a rare presentation of extramedullary hematopoiesis as a pelvic mass in a patient with β-thalassemia major, underscoring the importance of considering this condition in atypical locations. It also emphasizes the diagnostic and therapeutic challenges associated with extramedullary hematopoiesis, particularly the risk of life-threatening hemorrhage during attempts at surgical resection. The functional role

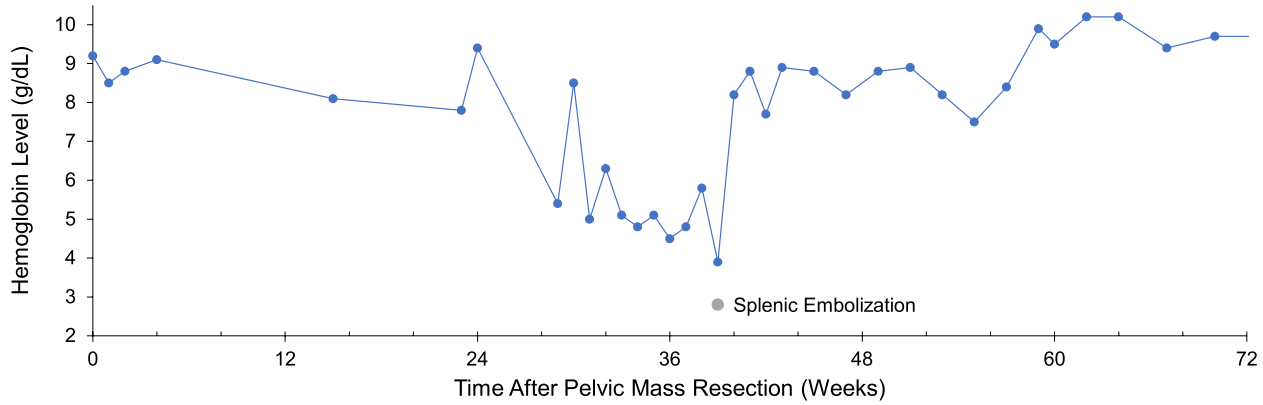

Fig. 5 Hemoglobin levels following pelvic mass resection. Hemoglobin levels over time following incomplete mass resection. The patient experienced a significant decrease in hemoglobin to 4.5 g/dL. Following splenic artery embolization, hemoglobin levels improved and stabilized between 8 and 10 g/dL

of extramedullary hematopoiesis as an alternative site of blood production is crucial, as its inadvertent removal can exacerbate anemia and increase the need for blood transfusions. Splenic artery embolization provides an effective therapeutic option to reduce the transfusion burden in such cases.

Authors' contributions

Z.H. and A.H. conceptualized and designed the study. Z.H. collected the clinical data and imaging studies. A.H. and S.K. drafted the initial manuscript. M.A. provided expertise in emergency medicine and contributed to the clinical interpretation of the findings. A.H., N.M., and M.A. reviewed and revised the manuscript for important intellectual content. All authors reviewed and approved the final version of the manuscript.

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Data availability

No datasets were generated or analysed during the current study.

Declarations

Ethics approval and consent to participate

This case report was conducted in accordance with ethical standards. Ethical approval was waived by the Research Committee of the Government Hospitals because of the nature of the case report. Informed consent was obtained from the patient for the publication of this case report, including the use of anonymized clinical data and images.

Competing interests

The authors declare no competing interests.

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