

# Overwhelming Asplenic Sepsis due to Babesiosis

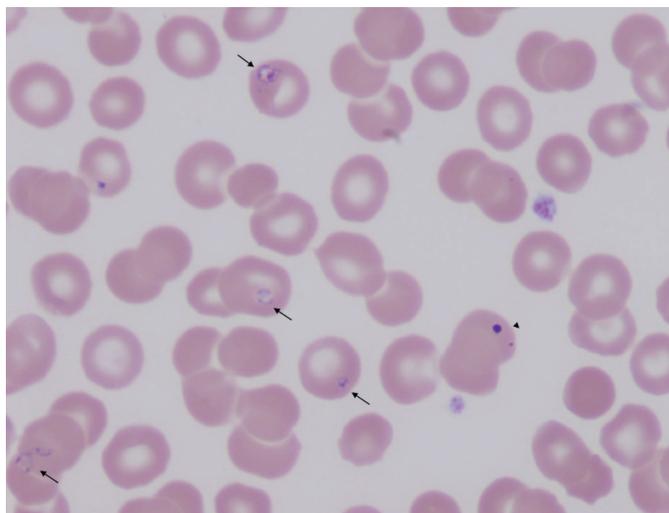
## Babesiyozis İlişkili Ağır Asplenik Sepsis

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### To the Editor,

A 70-year-old female from southern Massachusetts, USA, was admitted to the intensive care unit with septic shock and acute respiratory distress syndrome (ARDS) after 3 days of acute febrile illness. She had undergone splenectomy at the age of 5 related to trauma from a traffic accident. Laboratory studies revealed pancytopenia, acute renal insufficiency, increased lactate dehydrogenase, depressed haptoglobin, and elevated liver enzymes with indirect hyperbilirubinemia. Prothrombin time and activated partial thromboplastin time were both elevated and fibrinogen level was low, consistent with disseminated intravascular coagulation (DIC). A direct anti-globulin test was negative. A thin blood smear with oil immersion showed intraerythrocytic polymorphic ring forms (Figure 1, arrows) morphologically consistent with *Babesia* species and the presence of Howell-Jolly bodies (Figure 1, arrowhead), confirming the history of splenectomy. Real-time DNA-PCR confirmed *Babesia microti* as the offending parasite. The patient was started on treatment for babesiosis with quinine, azithromycin, and atovaquone. She also received red cell exchange transfusion due to the high level of parasitemia (14% of the erythrocytes) and completely recovered in the next few weeks.



**Figure 1.** Thin blood smear with oil immersion showed intraerythrocytic polymorphic ring forms (arrows) morphologically consistent with *Babesia* species and the presence of Howell-Jolly bodies (arrowhead), confirming the history of splenectomy.

Human babesiosis is a malaria-like tick-borne illness caused by the protozoan parasite *Babesia microti*, endemic in the Midwest and Northeast USA; it has also been reported in parts of Europe, Asia, and Australia [1]. It has also been reported after transfusion of contaminated blood products [2]. Infection is usually mild to moderate in an immunocompetent host but a severe infection requiring hospitalization can occur in patients with a history of splenectomy or immunodeficiency such as cancer, human immunodeficiency virus infection, or hemoglobinopathy and in the elderly with co-morbidities and allogeneic hematopoietic stem cell transplant recipients [1,3]. Severe babesiosis with ARDS and DIC can occur in immunocompromised or asplenic individuals, which can be fatal [4,5]. Milder illness in immunocompetent hosts manifests with malaise, fever, headache, myalgia, and nausea. Laboratory findings typically show non-immune hemolytic anemia and thrombocytopenia, but immune hemolytic anemia has also been reported. A rapid diagnosis can be made by identification of *Babesia* organisms on thin blood smears under oil immersion. The diagnosis can be confirmed by using DNA-PCR to identify the DNA of the parasite. Serology is available, but it is difficult to distinguish current from recent or past infection in a patient coming from an endemic area. The most commonly used agents for treatment of severe babesiosis include azithromycin, atovaquone, quinine, and clindamycin. Patients with severe infection with high-grade parasitemia, severe hemolysis, or compromised organ functions (pulmonary, liver, or renal impairment) may benefit from red cell exchange transfusion.

This case confirms that a severe form of babesiosis can occur in patients who have undergone splenectomy. A high index of suspicion and a timely review of blood smears in asplenic patients presenting with febrile illness and hemolytic anemia from endemic areas can aid in rapid diagnosis and prompt treatment, which can be lifesaving. This case illustrates that an early diagnosis and aggressive treatment can be lifesaving even with a fulminant and severe infection with babesiosis. The key is to quickly decrease the parasitic burden for a good clinical outcome.

**Keywords:** Babesiosis, Splenectomy, Sepsis

**Anahtar Sözcükler:** Babesiyozis, Splenektomi, Sepsis

**Conflict of Interest:** The author of this paper has no conflicts of interest, including specific financial interests, relationships, and/or affiliations relevant to the subject matter or materials included.

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# Isolated Mediastinal Myeloid Sarcoma after *NPM1*-Positive Pediatric Acute Myeloid Leukemia

*NPM1*-Pozitif Pediatrik Akut Myeloid Lösemi Sonrası İzole Mediastinal Myeloid Sarkom

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To the Editor,

Myeloid sarcoma (MS) is a rare extramedullary mass that consists of immature myeloid cells. The most common locations are the soft tissue, bone, periosteum, orbit, and lymph nodes [1,2]. Mediastinal involvement is very rare and most commonly reported with concurrent bone marrow involvement [3]. Herein we present a previously treated nucleophosmin (*NPM1*)-positive acute myeloid leukemia (AML) patient who later presented with isolated mediastinal MS.

A 9-year-old female patient presented with fatigue and weakness. Physical examination revealed no pathological findings. Blood tests demonstrated hemoglobin of 12.2 g/dL, hyperleukocytosis (100,500/ $\mu$ L), and thrombocytopenia (43,000/ $\mu$ L) with 88% blasts in the peripheral blood smear. Bone marrow aspirate revealed 90% blasts with M1 subtype. Treatment was started according to the AML-BFM 2012 protocol. Conventional cytogenetic analysis failed due to lack of spontaneous mitosis and fluorescent in situ (FISH) analysis for t(8;21), inv(16), t(15;17), and t(9;22) from bone marrow samples revealed negative results. Molecular genetic analysis in the peripheral blood showed *NPM1* positivity and *FLT3-ITD* negativity. Morphologic and molecular remission was obtained at the end of the first induction block. She presented with back pain and fever seven months after cessation of maintenance treatment. Computed tomography (CT) of the thorax showed a solid mass

of 84x75x41 mm in the anterior mediastinum (Figure 1). Bone marrow examination was normal; however, peripheral blood showed *NPM1* positivity. Conventional cytogenetic analysis from the bone marrow was within normal limits, while *NPM1* could not be studied from bone marrow. Her previous CT scans that were performed for investigation of invasive pulmonary aspergillosis were all normal. Fine-needle aspiration biopsy of the mass was performed; histopathological examination revealed myeloblasts that were positive for myeloperoxidase, CD15, and CD33. Microscopic examination of the imprint of the biopsy also revealed myeloblasts of M1 subtype (Wright stain). Major reduction in tumor mass (7 mm residual tumor) and *NPM1* negativity were achieved after one block of FLAG (fludarabine, cytarabine, filgrastim) and two blocks of FLAG-mitoxantrone. The patient underwent successful bone marrow transplantation from a matched unrelated donor and has been in remission for one year.

MS of the mediastinum is very rare; most of the cases have been reported as initial presentation with concurrent bone marrow involvement [3,4,5]. MS as a relapse has been more frequently reported in post-transplant patients compared to those treated without allogeneic hematopoietic stem cell transplantation [6,7]. Our patient is unique as she presented with isolated mediastinal MS after chemotherapy treatment. Another important point about our patient is that the *NPM1* positivity was detected at