

## A one-year- old boy with thrombositosis

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### Case presentation

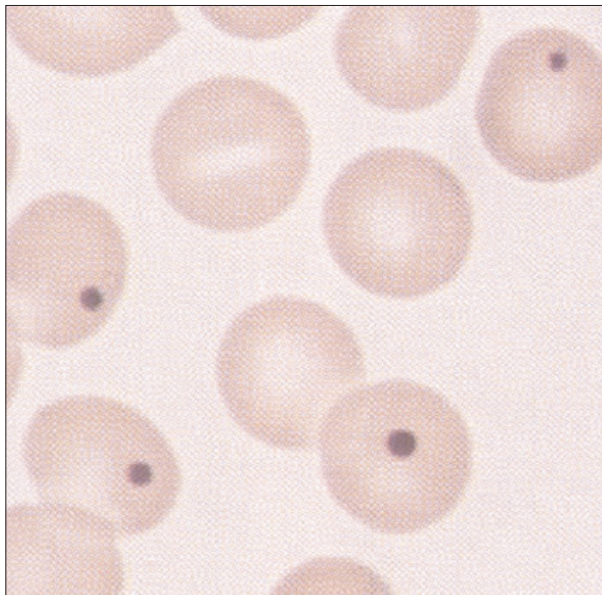
A 13 months old male patient was hospitalized with a diagnosis of reactive airway disease because of frequently recurring attacks of bronchiolitis and started to be treated. In his history, it was learned that the parents were healthy and had third degree consanguinity between each other. The patient was diagnosed as right renal atrophy prenatally and was operated postnatally when he was 9 days old because of anal atresia and uretherocele. Physical examination revealed the following: height 80 cm (50-75<sup>th</sup> percentile), weight 11.5 kg (75<sup>th</sup> percentile), blood pressure: 100/55 mmHg, heart rate:110/min, respiratory rate: 44/min, body temperature: 37.5°C. Wheezing was heard on auscultation of the lungs and the patient had respiratory distress. Intercostal and subcostal

retractions were observed. Seven fingers were present in the right hand and 6 fingers were present in both lower extremities. Examination of the other systems was normal.

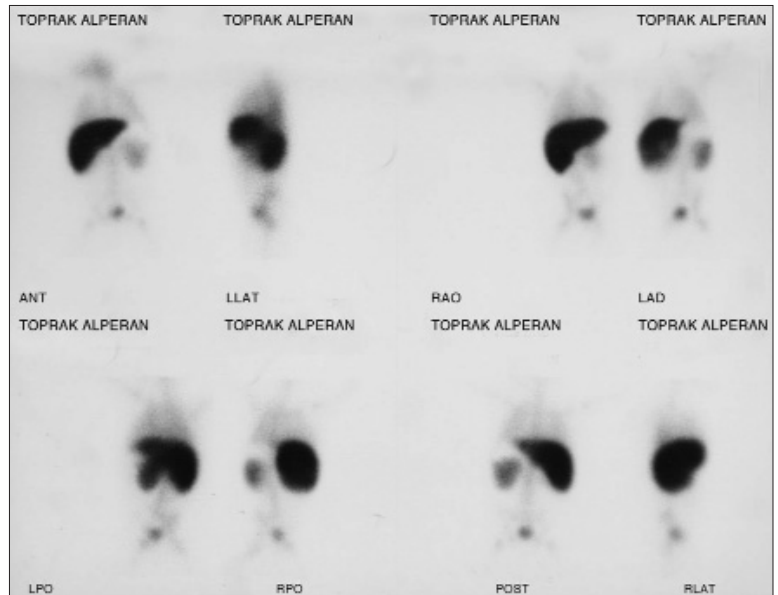
On the posteroanterior chest graphy, bilateral increased aeration, flattened costae and intensive involvement were observed.

Laboratory tests revealed the following: hemoglobin 11.6 g/dL, hematocrit 33.7%, platelet count 901000/mm<sup>3</sup>, white blood cells 23400/mm<sup>3</sup>. Peripheral blood smear revealed 90% neutrophil dominance. C-reactive protein was found to be 51 mg/L. Recurrent platelet counts were found to be 949000/mm<sup>3</sup> and 643000/mm<sup>3</sup>, respectively.

New investigations were ordered, since thrombocytosis persisted in the patient whose bronchiolitis was improved (Picture 1,2).



Picture 1-Peripheral blood smear



Picture 2-Scintigraphy

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## Diagnosis-Asplenia

Howell-Jolly bodies were found in the erythrocytes on peripheral blood smear (Picture 1). On abdominal ultrasonography which was ordered to confirm the presence of the spleen, the spleen could not be observed in its location. The spleen could not be found in the selective spleen scintigraphy performed by 6mCi TC -99m pertechnetate (Picture 2).

## Discussion

Thrombocytosis is observed in children related mainly to 2 causes; reactive thrombocytosis due to various factors and essential thrombocytosis (1). Essential thrombocytosis is observed very rarely in children and secondary causes should be considered primarily in presence of thrombocytosis. Asplenia, hyposplenism, acute blood loss, infections, iron deficiency anemia, chronic inflammatory diseases and malignancies (for example. Hodgkin lymphoma, neuroblastoma) are the most commonly observed causes of thrombocytosis (2). History and physical examination are important in the differential diagnosis.

The spleen is the largest lymphoid organ in the body and has four important functions including microfiltration, immunity, hematopoiesis and hemostasis. Normally, 1/3 of the thrombocytes are deposited in the spleen. One of the most important functions of the spleen is filtration of the erythrocytes. In presence of substance in the erythrocyte, this part of the erythrocyte is pulled off and the erythrocyte which has not lost its function is given back to the circulation. If the erythrocyte can not perform its function after cleaning procedure, it is cleaned completely from the circulation. When the spleen can not perform its function, erythrocytes can not be filtered and circulating erythrocytes have residuals which are called Howell-Jolly bodies (3,4).

Asplenia is congenital or aquired absence of the spleen. Congenital splenic anomalies can be associated with other organ anomalies especially cardiac anomalies or can be present alone (Ivemark syndrome) (5). Solitary asplenia is generally recognized after a severe, fulminant and fatal infection (6). In the absence of the spleen, the risk of fulminant infection with bacteriae with capsules including *Streptococcus pneumoniae*, *Hemophilus influenzae* and *Neisseria meningitidis* increases. Sometimes, inclusion bodies inside the erythrocytes can be the only proof of asplenia. Asplenia should also be considered in causeless thrombocytosis. In the absence of the spleen, neutrophilia, leucocytosis and mild reticulocytosis are also observed. In diagnosis, history and peripheral blood smear are very simple, but significant methods.

In this case presentation, we desired to emphasize that history and peripheral blood smear are simple, but significant elements.

## References

1. Demirören K, Çalışkan Ü, Keser M. Çocukluk çağında esansiyel trombotoz: olgu bildirisi. *Türk Ped Arş* 2002; 37: 160-3.
2. Çomak E, Örmeci AR, Kırbıyık S, Eren E, Çetin H. Alt solunum yolu enfeksiyonu olan çocuklarda sekonder trombotoz. *Türkiye Klinikleri J Pediatr* 2005; 14: 62-5.
3. Ware RE. Autoimmune hemolytic anemia. In: Nathan DG, Orkin SH, Ginsburg D, Look AT, et al (eds). *Hematology of infancy and childhood*. 7th ed. Philadelphia: Saunders, 2009; 617.
4. Ahmed SA, Zengeya S, Kini U, Pollard AJ. Familial isolated congenital asplenia: case report and literature review. *Eur J Pediatr* 2010; 169: 315-8.
5. Noack F, Sayk F, Ressel A, Berg C, Gembruch U, Reusche E. Ivemark syndrome with agenesis of the corpus callosum: a case report with a review of the literature. *Prenat Diagn* 2002; 22: 1011-5.
6. Gilbert B, Menetrey C, Belin V, Brosset P, de Lumley L, Fisher A. Familial isolated congenital asplenia: a rare, frequently hereditary dominant condition, often detected too late as a cause of overwhelming pneumococcal sepsis. Report of a new case and review of 31 others. *Eur J Pediatr* 2002; 161: 368-72.