Pelger-Huët anomaly is a rare benign autosomal-dominant anomaly with incidence of about 1 in 6000 persons. Pelger-Huët cells are morphologically abnormal neutrophils that are either bilobed or completely unsegmented.1,2 It should be differentiated from acquired or pseudo-Pelger-Huët anomaly (PPHA), which has similar morphology and association with different pathological states like myelodysplasia, Acute myeloid leukemia (AML), chronic myelogenous leukemia (CML) as well as with certain infections and drugs (Valproate and Ibuprofen).3-9

An 18-year-old woman who was a case of mental retardation since early childhood was admitted with episodes of epistaxis and pancytopenia. She had a history of end stage renal disease on chronic intermittent dialysis and history of seizure on sodium valproate and nitrazepam. Since 1 month ago valproate dosage was increased from 200 mg twice a day to 500 mg twice a day. Physical examination showed pallor, and inability to speak and follow orders. There was no lymphadenopathy and organomegaly. Laboratory findings showed WBC: 3300/µL, Hb: 6 gr/dl, Platelet: 45000/µL, Creatinine: 7 mg/dL. Peripheral blood smear showed many bi-lobed neutrophils (pseudo pelger huet cells) (Figure 1).

We considered a decrease in valproate dosage and initiation of folate and B12 supplement for her. In the follow-up visits, leukocyte counts had increased to 5000/µl. Bone marrow aspiration and biopsy revealed marked hypocellular marrow with evidence of dyserythropoiesis and dysmyelopoiesis. Cytogenetic study and chromosomal breakage should be considered on bone marrow aspiration of subjects with Pelger-Huët neutrophils to rule out the diagnosis of hereditary forms of myelodysplastic syndromes and Fanconi anemia in patients with Pelger-Huët anomaly.

Conflict of Interest: None declared.

References


