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## Hodgkin lymphoma in a patient with IFAP syndrome

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**Introduction:** The IFAP syndrome is a rare X-linked recessive inheritance disorder with defect of the gene MBTPS2 defined by the triad of follicular ichthyosis, atrichia, and photophobia. Up to 40 cases were described till now, but association of this syndrome with Hodgkin lymphoma has not been reported so far.

**Case Presentation:** A 3.5 years old girl, known case of IFAP syndrome confirmed by Next Gene Sequencing method that initially presented with congenital baldness, kyphosis and photophobia was examined in this study. She had syndactyly and polydactyly, ichthyosis, bilateral inguinal hernia, failure to thrive, developmental delay, psychomotor retardation, seizure, congenital hypothyroidism and gastroesophageal reflux disease. Metabolic study was normal. The chromosomal study revealed a karyotype 46, XX. Next Gene Sequencing showed defect in MBTPS2 gene. The patient developed fever 1.5 month ago and then, cervical and axillary lymphadenopathy but no organomegaly. She showed no response to antibiotic therapy.

**Lab Data:** Data revealed WBC: 9800/mm<sup>3</sup>, Hb: 8.2 mg/dL, PLT: 950000/mm<sup>3</sup>. ESR: 137, CRP: 36. Her C3, C4, ANA, DSDNA and all immunoglobulin levels were normal. So bone marrow aspiration and biopsy revealed normocellular bone marrow. Cervical lymph node biopsy showed Hodgkin lymphoma with mixed cellularity. Immunohistochemistry showed positive CD15, CD20, CD3, CD30, CD5 and PAX5, and Ki67: 80% high large cells.

**Conclusions:** This report implies that the IFAP syndrome may be prone to developing Hodgkin lymphoma because, Hodgkin lymphoma is not common in children younger than 6 years. So, the IFAP syndrome may be a risk factor for developing Hodgkin lymphoma.

### Biography

Mahdi Shahriari is currently working as an Associate Professor in Department of Paediatrics in Shiraz University of Medical Sciences, Iran since 1994. He has published many articles in reputed national & international journals. His area of expertise includes Medicine, Oncology, Hematology, Pediatric Hematology, Pediatric Oncology, Hemophilia, Thalassemia & Cord Blood Stem Cell Transplantation.

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