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**Gastric signet ring carcinoma in a patient with ataxia-telangiectasia: a case report and review of the literature**

Ataxia telangiectasia (A-T) is an autosomal recessive primary immunodeficiency disease characterized by progressive cerebellar ataxia, telangiectasia, sinopulmoner recurrent infections and cancer susceptibility. Individuals with A-T are known to be increased risk of certain malignancies including leukemia, lymphoma, breast and gastric cancer. We present an 18-year-old case of A-T with Hashimoto's thyroiditis who admitted with complaints of nausea, vomiting, anorexia and weight loss. An upper endoscopic biopsy revealed gastric signet ring cell carcinoma. According to the best of our knowledge, we report the first case of signet ring cell carcinoma in the patient with A-T. Our experience about occurrence of Hashimoto's thyroiditis and gastric signet ring cell carcinoma in the same case of A-T underlines that; the clinicians handling with A-T must be vigilant about both malignancy and auto-immune disorders.