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LETTER TO THE EDITOR
Uncommon Presentation of Craniopharyngioma With Anemia in an Adolescent

To the Editor: Craniopharyngioma most commonly presents with symptoms related to increased intracranial pressure, such as headache and vomiting [1,2]. Less common presenting symptoms are secondary to the invasion of the neighboring structures, which include the pituitary, the hypothalamus, the optic pathways, and the third ventricle. Visual and endocrine abnormalities are usually present at initial evaluation although only a minority of patients seeks medical attention because of symptoms related to these abnormalities [1,3,4]. Anemia is a common, nonspecific finding in the ambulatory pediatric practice but constitutes an unusual initial presentation for craniopharyngioma. We present the case of a 13-year-old female who was referred for the evaluation of mild normocytic anemia and whose further investigation led to the diagnosis of panhypopituitarism secondary to craniopharyngioma.

The patient's academic performance had deteriorated over the last 6 months, which instigated the investigation for anemia. She also reported fatigue, constipation, and weight gain of 5 pounds in the last 6 months despite a poor appetite. She denied headache or visual symptoms. Her menarche had occurred a year prior to her presentation and her menstrual periods were regular but lasted only 2 days and consisted of spotting. The birth and past medical history were unremarkable. She was not taking any medications. The family history was significant for hypothyroidism in a 29-year-old sister and was otherwise negative. Aside from delayed puberty (Tanner stage II for breasts and Tanner stage I for pubic and axillary hair), the physical examination was essentially unremarkable, with no focal neurologic findings.

White blood cell count was 4,800/mm^3 with a differential of 29% neutrophils, 58% lymphocytes, 8% monocytes, and 5% eosinophils. Hemoglobin was 10.9 g/dl and the red blood cell count was 3.71 million/mm^3. The platelet count and the absolute reticulocyte count were within normal range. Red blood cells were normocytic and the other red cell indices were normal. Sedimentation rate, serum aminotransferases, bilirubin, blood urea nitrogen, and creatinine were within normal range.

Our patient's symptoms (weight gain despite loss of appetite, fatigue, worsening school performance, amenorrhea and constipation) suggested the presence of hypothyroidism.

Serum levels of free thyroxine and total thyroxin were low (0.5 ng/dl and 3.8 μg/dl, respectively) in the face of a normal thyroid stimulating hormone (TSH) level (1.94 μ IU/ml). The anti-thyroglobulin and antimicrosomal antibodies were negative. The inappropriate TSH response to low thyroxine levels, coupled with other abnormal findings (delayed puberty and lymphocyte predominance in the white blood cell differential) further led to a clinical suspicion of hypopituitarism in our patient. Serum levels of A.M. cortisol, insulin like growth factor-1 (IGF-1), IGF binding protein-3, follicle stimulating hormone, and luteinizing hormone levels were also low, establishing a diagnosis of hypopituitarism. Serum prolactin level was within normal range. The radiological bone age revealed mild delay in skeletal maturity. Urine and serum osmolality were not suggestive of diapedes insipidus.

A magnetic resonance imaging of the brain revealed a sellar/suprasellar lesion, 2.5 cm in diameter, demonstrating hyperintense signal in T1-weighted images, most likely representing a craniopharyngioma. The sella turcica was widened and a normal pituitary and infundibulum were not seen. A subsequent computerized tomographic scan showed a rim of calcification within the lesion. Automated perimetry using Humphrey field analyzer revealed bitemporal hemianopia.

The patient underwent bifrontal craniotomy and total resection of the tumor. The histologic examination of the tumor revealed dense fibrous tissue and cystic spaces lined by squamous epithelial cells with peripheral palisading mimicking adamantinoma of the jaw, characteristic of craniopharyngioma. Post-operatively the patient developed diabetes insipidus and monocular diplopia. She was started on oral thyroxine, hydrocortisone, and desmopressin supplementation and the hemoglobin rose from 10.9 to 12.2 g/dl 3 months after surgery.

Although an uncommon presenting symptom for craniopharyngioma, anemia may be commonly discovered

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in the initial evaluation of cases of craniopharyngioma presenting with other symptoms. Anemia was detected in 22% of patients with a pituitary tumor and 41% of patients with craniopharyngioma [5]. The anemia encountered in these tumors is a manifestation of the endocrine deficiency, which includes hypothyroidism, hypocortisolism and androgen deficiency in males [6,7]. The resolution of the anemia with hormonal supplementation alone observed in our case supports its endocrine basis. Clinicians evaluating isolated hypoproliferative normocytic anemia (i.e., low or normal reticulocyte count) should exclude acute infection, renal disease, hepatic disease, transient erythroblastopenia of childhood and hypothyroidism as a potential cause of anemia [7,8]. Accompanying absolute or relative lymphocytosis and eosinophilia usually suggests hypocortisolism and should instigate a search for hypopituitarism [6].

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