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Delayed diagnosis of hypothyroidism following excision of a thyroglossal duct cyst

Jessica S. Lilley, MD and **Jefferson P. Lomenick, MD**
Department of Pediatrics, Division of Endocrinology, Vanderbilt University School of Medicine, Nashville, TN, USA

Abstract

Thyroglossal duct cysts can contain ectopic thyroid tissue, and in some cases this tissue may be the only functional thyroid gland. We present a 6-year-old girl with delayed diagnosis of iatrogenic hypothyroidism that developed following excision of a thyroglossal duct cyst.

Hypothyroidism is a common pediatric endocrine disorder in children that is usually due to Hashimoto's (autoimmune) thyroiditis in school-aged children and teenagers. The etiology in younger children is usually congenital hypothyroidism, which affects one in 4000 live births in the United States.[1] Although many patients with hypothyroidism have classic symptoms like fatigue, constipation, cold intolerance, and dry skin, the symptoms often develop insidiously, so the diagnosis is occasionally not made until months or years later when the patient is noted to have growth failure. We describe a patient with severe, long-standing iatrogenic hypothyroidism due to removal of her only functional thyroid tissue during surgery to resect a thyroglossal duct cyst (TGDC).

Case Report

A 6-year-old girl presented to her pediatrician's office with a 2-year history of growth deceleration. She had always been healthy and had had an uneventful perinatal course, including a normal state newborn screen thyroid stimulating hormone (TSH) measurement. Her height was around the 70th percentile until age 4 years, but her height velocity between her 4- and 6-year well child visits was less than 1 cm/yr, causing her height to decline to <3rd percentile. Review of systems revealed chronic constipation which had worsened recently requiring daily polyethylene glycol therapy and increased fiber intake, cold intolerance (wearing fleece pajamas in the summer), emotional changes, dry skin, and swelling around her eyes. Family history was negative for thyroid disorders or other autoimmune disease.

Correspondence: Jessica S. Lilley, MD 2200 Children's Way 11136 Doctors' Office Tower Nashville, TN 37232-9170 Phone 615-936-1874 Fax (615) 875-7633 jessica.lilley@vanderbilt.edu.

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Past medical history was significant for a midline neck mass thought to be a TGDC two years prior, which was surgically removed. The protrusion had been present since infancy and had appeared to grow along with her. No preoperative imaging or laboratory assessment was obtained. She did well with the surgery, and the mass and a portion of the hyoid bone, as indicated in the operative report, were removed. The surgical pathology report read "thyroglossal duct cyst with abundant thyroid tissue, portions of hyoid bone and cartilage."

Physical exam revealed a small, overweight child with nonpalpable thyroid gland; height was <3rd percentile (−3.12 SD) and weight at the 27th percentile. Laboratory evaluation was significant for TSH >150 μU/ml (normal 0.3-5 μU/ml) and free T4 0.25 ng/dl (normal 0.89-1.76 ng/dl); anti-thyroid antibodies were negative. Bone age was markedly delayed, interpreted as 2 years 6 months at chronologic age 6 years 0 months. Neck ultrasound showed no thyroid gland present. Levothyroxine therapy was initiated at 37.5 micrograms daily and was gradually titrated over several months to her current dose of 75 micrograms daily. Four months after starting treatment, her height had improved significantly (−2.7 SD); BMI decreased from the 94th to 66th percentile.

Discussion

Ectopic thyroid is a rare cause of hypothyroidism, occurring in approximately 1 in 200,000 individuals, though this may be a conservative estimate due to the often asymptomatic nature of this anomaly.[2] The primordial thyroid gland forms from the first and second pharyngeal pouches at the base of the tongue during the fourth week of gestation and descends over the midline of the neck to its usual position by the seventh week. The tract of its descent usually disappears by the tenth week of gestation but may persist, along with remnants of associated thyroid tissue. Thyroid that does not descend normally is termed ectopic and most commonly stays at the base of the tongue; 90% of ectopic thyroid glands are lingual. The tract may persist and expand to form a TGDC, which is the second most common location for ectopic thyroid tissue.[2-4] Many TGDCs go undetected; one autopsy study revealed a 7% prevalence of undiagnosed TGDCs in a series of adults.[5]

TGDCs may become apparent with infection, carcinoma (in 1-2% of cases[4]), or hypertrophy of associated thyroid tissue in response to hypothyroidism-mediated thyrotropin release. Although 25-65% of TGDCs are associated with rests of thyroid tissue in people with normally formed eutopic thyoid,[6] patients who present with apparent TGDC can actually have an ectopic thyroid gland (ETG) associated with the cyst, sometimes without functioning thyroid tissue anywhere else.[6,7] Two-thirds of ETGs function normally, but about a third present with hypothyroidism. It is thought that even though ETGs may produce sufficient thyroid hormone during early childhood, times of stress such as growth, puberty, illness, or surgery may demand more output than the ETG can provide and will result in rising TSH and increased production of colloid. This tissue, like eutopic thyroid tissue, will hypertrophy in response to increased TSH, making it more obvious and bothersome.

Because our patient did not have thyroid function tests pre-operatively, it is unclear whether she had a normally functioning ETG contained within a TGDC that happened to expand or if she was becoming hypothyroid and the apparent enlarging TGDC was actually TSH-

stimulated ectopic thyroid tissue. Given her history, we favor the former, but this cannot be proven. Regardless, it is clear that the TGDC contained her only functional thyroid tissue, and it was removed during surgery. Fortunately, she was older than three years when she became hypothyroid. Had she been younger than three, her cognitive function might have suffered. Whether she will regain the lost height potential during her period of hypothyroidism is uncertain at this time.

A small number of children with similar presentations as our patient have been described in the literature.[2,8-11] Additionally, a 14 year old girl was reported with multiple foci of thyroid ectopia presenting as a neck mass; she had no orthotopic thyroid and was already hypothyroid at presentation.[12] Various options for pre-operative evaluation have been suggested, including radionuclide imaging[3,5,13] and ultrasound,[13] to identify ETG when a patient is referred for evaluation of TGDC.

We propose checking TSH prior to referral to a surgeon and obtaining thyroid ultrasound to identify normal thyroid gland or ectopic thyroid tissue. If hypothyroidism is discovered, a trial of levothyroxine therapy may be able to suppress the enlarged ETG, and surgery may be avoided. However, if a patient does need surgery due to persistent enlargement, deformity, symptoms like odynophagia, or infection, we emphasize that hypothyroidism is a preventable complication that should be discussed with families prior to the procedure and screened by the primary care provider by checking a TSH after the procedure. If there is insufficient thyroid tissue left to allow euthyroidism or the only functioning thyroid tissue was removed, as in our case, this would easily be identified, as the TSH is typically >30 μU/ml within two weeks of thyroidectomy.

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