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CLINICAL RECORD

Simultaneous occurrence of a thyroglossal duct cyst and a lingual thyroid in the absence of an orthotopic thyroid gland

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Abstract

Objective: We report an extremely rare case of the simultaneous occurrence of a thyroglossal duct cyst and a lingual thyroid in the absence of an orthotopic thyroid gland, in a seven-year-old girl from South India.

Method: Case report and a review of the English language literature on the subject.

Results: The patient presented with a mass on the tongue that had been present for three years, and an anterior neck swelling that had been present for two years. Examination revealed a midline, pinkish, firm mass present on the posterior one-third of the tongue. The neck showed a midline cystic swelling in the infrahyoid position. Radiological imaging confirmed the clinical findings, revealing the absence of her thyroid gland in the normal location. Sistrunk's procedure was performed leaving behind a lingual thyroid. At 13-month follow up, the patient was euthyroid with no recurrence.

Conclusion: To our knowledge the association of a lingual thyroid and a thyroglossal cyst has only been reported once in the literature. The presence of a lingual thyroid in the absence of a normally located thyroid gland or functioning thyroid tissue along the thyroglossal tract, confirmed by radionuclide and computed tomography imaging, may indicate the failure of the normal descent of the thyroid gland during embryonic development. This probable absence of the descent of the thyroid raises questions regarding the origin of thyroglossal duct cysts.

Key words: Thyroid, Lingual; Thyroglossal Duct Cyst; Orthotopic; Thyroid Gland

Introduction

Thyroglossal duct anomalies are the most common malformations in the midline of the neck.[1] They may sometimes harbour ectopic thyroid tissue.[2] Lingual thyroid is a developmental anomaly that is formed from a rest of thyroid tissue left behind during embryonic migration.[3] An orthotopic, functioning thyroid gland may be absent in either of the above anomalies. The simultaneous occurrence of all three anomalies is extremely rare and had been reported only once in the English language literature, by McCoul and de Vries.[2] We report the coexistence of these thyroid anomalies in a seven-year-old girl from South India.

Case report

A seven-year-old girl presented with a mass on the posterior part of the tongue that had been slowly increasing in size for three years, and an anterior neck swelling that had been present for two years. The onset had been spontaneous with no history of pain. She did not complain of dysphagia, dyspnoea or voice change. There was no history of radiation exposure or familial thyroid disease. There was no history suggestive of hyper- or hypothyroidism.

Examination of the neck revealed a 3×3 cm, smooth-surfaced, soft, cystic, round swelling present on the anterior surface of the upper part of the midline neck (Figure 1). The swelling moved upwards with protrusion of the tongue and with deglutition. On examination of the oral cavity, a hypervascular, pinkish mass with a lobulated surface was noted on the posterior one-third of the tongue at the midline (Figure 2). Systemic examination revealed no signs of hypo- or hyperthyroidism. Ultrasound of the neck confirmed the presence of a heterogeneous mass with multiple cystic and nodular areas, with no evidence of normal thyroid tissue in its orthotopic position. Fine needle aspiration cytology of the neck swelling was consistent with a thyroglossal duct cyst and there were no signs of malignancy.

A computed tomography (CT) scan with intravenous contrast revealed a 3.8×3.4 cm, rounded, soft-tissue mass lesion with central hypodense areas and peripheral enhancement, located antero-inferior to the body of the hyoid bone (Figure 3). It also revealed a 3.2×3 cm soft-tissue lesion with contrast enhancement on the posterior one-third of the tongue (Figure 4). Radionuclide scanning with iodine-131 revealed dense uptake at the base of the tongue, and mild uptake was noted in the anterior aspect of the neck, above the expected position of the thyroid gland. There was no radioactive uptake in the normal location of the thyroid gland (Figure 5). The patient's thyroid function test was within normal limits.

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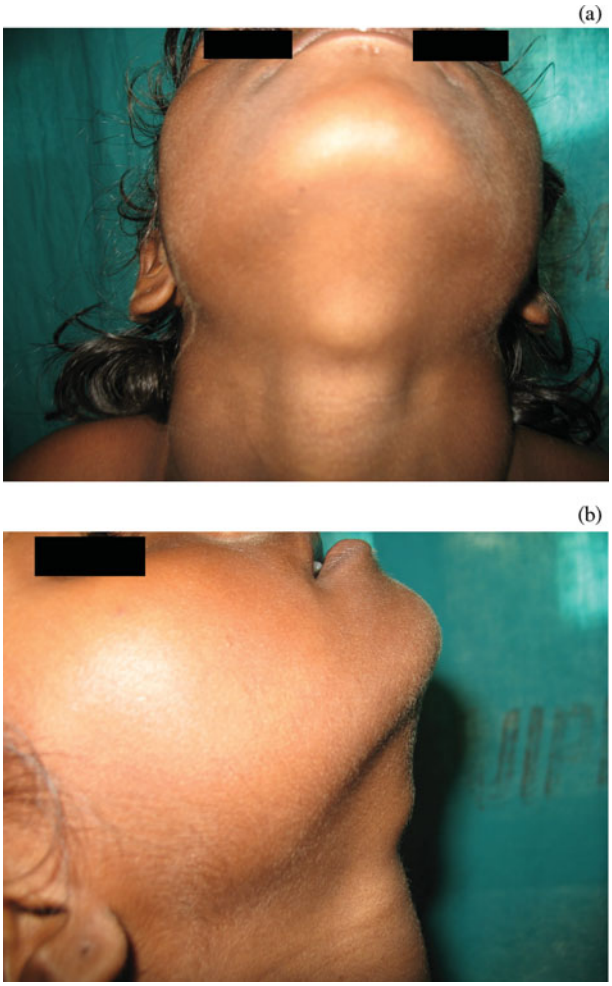

FIG. 1

Clinical photographs of a 3 × 3 cm, smooth-surfaced, soft, cystic, round swelling present in the anterior aspect of the upper part of the midline neck.

Sistrunk's procedure was planned for cosmetic reasons and because of the patient's anxiety, with preservation of the lingual thyroid as the patient was asymptomatic and its excision would have resulted in lifelong dependence on exogenous levothyroxine.

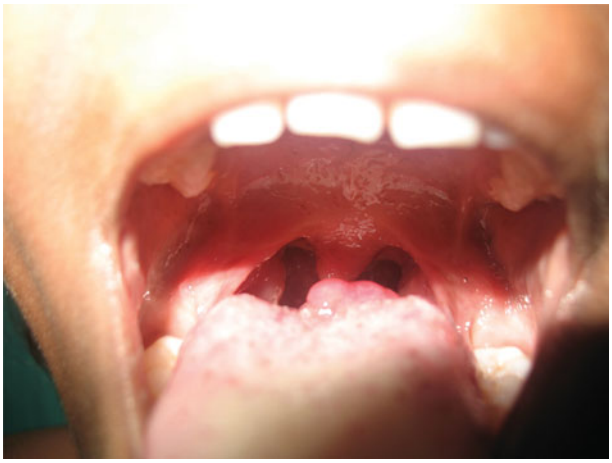

FIG. 2

Intra-oral examination showing a hypervascular, pinkish mass with a lobulated surface on the posterior one-third of the tongue at the midline.

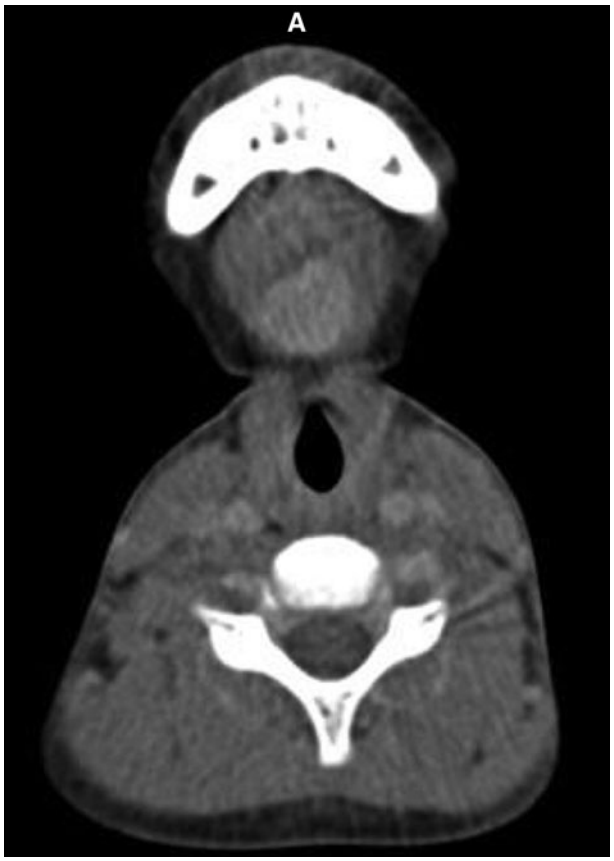

FIG. 3

Axial computed tomography scan with intravenous contrast showing a 3.8 × 3.4 cm, rounded soft-tissue mass lesion with central hypodense areas and peripheral enhancement, located antero-inferior to the body of the hyoid bone. A = anterior

Under general anaesthesia, Sistrunk's procedure was undertaken with a transverse cervical incision at the level of the swelling. The subplatysmal flap was raised and the cyst was dissected free of the infrahyoid musculature. Superiorly, the cyst was attached to the body of the hyoid bone, which was resected along with the cyst in an en-bloc fashion. Feeding vessels were identified and ligated. The patient's post-operative course was uneventful.

Histopathological examination of the specimen revealed chronic inflammation and is consistent with a thyroglossal duct cyst. At 13-month follow up with a clinical examination, a thyroid function test and ultrasonography of the neck, the patient was euthyroid with no evidence of recurrence.

Discussion

In 1891, His described the embryological basis of thyroglossal remnants in the formation of a thyroglossal duct cyst. At four weeks' embryological development, the thyroid gland originates as a median epithelial outpouching in the region of the foramen caecum, at the junction of the hypobranchial eminence and the tuberculum impar from the floor of the primitive pharynx.[1] This outpouching descends in the midline anterior neck in front of the hyoid bone to reach its final position as a bilobed diverticulum, anterior to the second and third tracheal rings during the seventh week of gestation. During its migration, the thyroid remains

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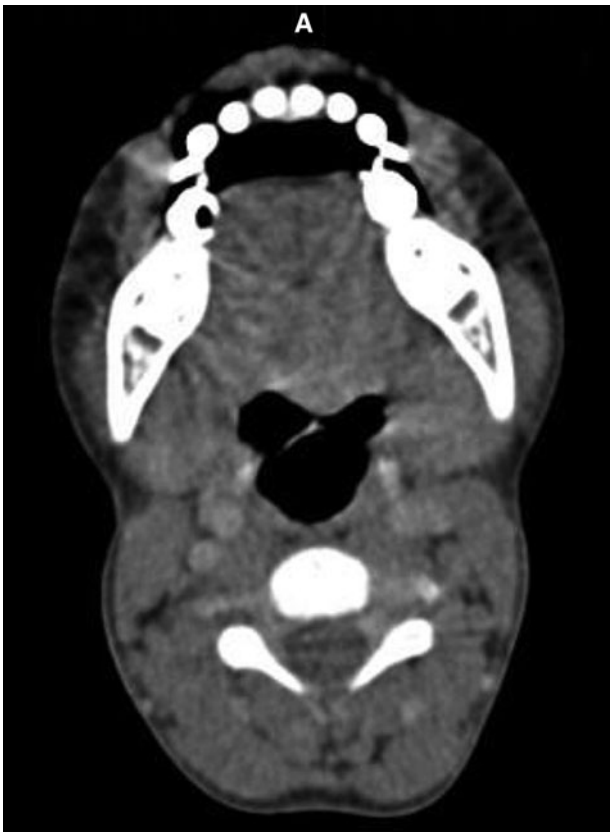

FIG. 4
Axial computed tomography scan showing a 3.2 × 3 cm soft-tissue lesion with contrast enhancement on the posterior one-third of the tongue. A = anterior

connected to the tongue by the thyroglossal duct, which atrophies by the 10th week of gestation. Ectopic thyroid rests can occur anywhere along the tract of the thyroglossal duct and have been found in 10 per cent of autopsies.[2]

It is thought that maternal antithyroid antibodies may play a role in preventing the thyroid from progressing along its normal developmental course of descent.[3] Although thyroglossal duct cysts are congenital in origin, they are rarely identified in the neonatal or infantile period but are frequently encountered in children. Sixty per cent of these lesions are diagnosed before the age of 20 years. They commonly present before five years of age in the suprahyoid location, whereas presentation in the infrahyoid position is common among adults.[1] Infection of the cyst is usually associated with upper respiratory catarrh. Recurrent infections and suspicion of malignancy are indications for surgery. Pre-operative evaluation includes nuclear and radiological imaging to ascertain the normal position of the thyroid gland.[2]

Simple excision of the cyst is reported to result in a 50 per cent recurrence rate. Sistrunk in 1920 advocated excision of the cyst along with dissection of the body of the hyoid bone and the base of the tongue to reduce the recurrence rate to less than 10 per cent. This procedure has since become the standard treatment for thyroglossal duct cysts.[1,2]

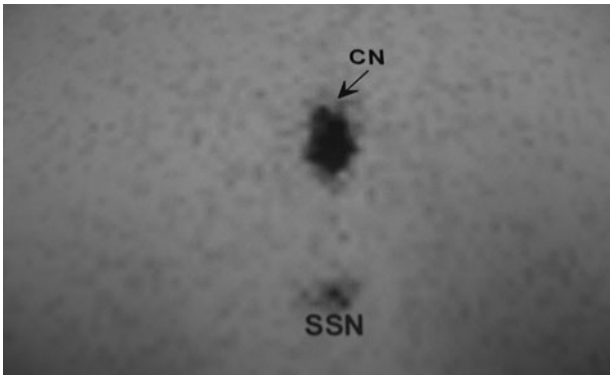

FIG. 5
Iodine-131 scan (anterior view) showing dense uptake at the base of the tongue and mild uptake in the anterior aspect of the neck, above the expected position of the thyroid gland. There was no radioactive uptake in the normal location of the thyroid gland. CN = CHIN; SSN = Suprasternal Notch

- **We describe the rare occurrence of a concurrent lingual thyroid and a thyroglossal duct cyst without an orthotopic thyroid gland**
- **These findings suggest a failure of the descent of the thyroid gland during embryological development**
- **This case raises questions regarding the origin of thyroglossal duct cysts**

Lingual thyroid accounts for 90 per cent of cases of thyroid ectopia and was first reported by Hickman in 1869.[4,5] It is hormonally responsive, hence it may present during puberty and pregnancy with acute enlargement of the swelling. This ectopic tissue is prone to functional insufficiency and may only be noticed after compensatory enlargement. As in most patients this is the only functioning thyroid tissue, the removal of the thyroid ectopia frequently results in severe hypothyroidism.[6] As many as 70 to 100 per cent of patients with symptomatic lingual thyroids do not have normally located thyroids. Diagnosis is usually made by clinical examination, although CT or nuclear imaging may provide additional information. Hypothyroidism may be seen in up to one-third of patients.[3] The use of pertechnetate and radioactive iodide-131 to determine the size, distribution and activity of the thyroid tissue is well established. Ectopic thyroid tissue can be identified with a CT scan; however, it is not used as a diagnostic investigation because of the cost and as similar results can be obtained by ultrasound and radionuclide scanning.[6]

Treatment consists mainly of surgical excision or hormonal suppression. Ablation therapy has been discouraged because of reported concerns about delayed clinical response and potential airway obstruction. Reservations have been expressed about the radioactive dose required to ablate lingual thyroid, because of concerns that most ectopic thyroid tissue is hypoactive and the ablative dose may be too toxic. However, contrary to the aforementioned concerns, there have been several reports detailing no associated airway compromise with iodine-131 treatment of symptomatic compressive goitres. This has prompted consideration of iodine-131 ablation treatment for patients with symptomatic lingual thyroid.[3] Techniques of autotransplantation of the thyroid tissue to the lateral pharyngeal wall, involving a vascular pedicle following excision of the lingual thyroid, have been described.[7,8]

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There has only been one similar case reported previously in the available English language literature. Treatment of the condition depends on the symptoms caused by the neck and tongue masses, as well as the functional status of the thyroid. In our case, Sistrunk's procedure was performed to alleviate the concerns of the patient and for cosmetic purposes. The lingual thyroid tissue was left behind because of the absence of symptoms and to preserve her euthyroid status.

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Dr J Madana takes responsibility for the integrity of the content of the paper
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