Case Report

Hypothyroidism Following Sistrunk Procedure: Thyroglossal Duct Cyst or Ectopic Thyroid?

Hassan Abdullahi Elechi
Department of Paediatrics, University of Maiduguri, Maiduguri, Nigeria

Abstract

Thyroglossal duct cyst is the most common cause of anterior midline neck swelling in children, but ectopic thyroid is an important but rare differential diagnosis. Establishing the orthotopic thyroid gland status is crucial as inadvertent removal of a lone functioning ectopic thyroid tissue in young children could lead to fluttering of growth and development if not identified early. We report the case of a 2 years 10-month-old boy who had Sistrunk's procedure for 'thyroglossal duct cyst': Diagnosed using USS, defaulted follow-up and presented subsequently with growth retardation from hypothyroidism.

Keywords: Ectopic thyroid, hypothyroidism, thyroglossal duct cyst, ultrasound scan

Introduction

The thyroid gland develops from endodermal epithelial thickening at the foramen caecum, the junction between the anterior two-third and posterior third of the tongue and descend in front of the pharyngeal gut to its destination in front of the trachea by 45–50 days of gestation. During this migration, the thyroid remains connected to the tongue through a narrow canal, the thyroglossal duct which normally disappears later. In a few individuals, the thyroglossal duct fails to obliterate either partially or along its entire length giving rise to a cystic swelling – thyroglossal duct cyst. It is a midline swelling in most cases but could be laterally displaced in a few.[1,2]

Thyroglossal duct cyst is the most common congenital neck swelling affecting <1%–7% of the population. It often manifests during childhood but can first be noticed during adulthood.[3,4] Although the cyst can lie at any point along the migratory pathway of the thyroid, 50%–80% are inferior to the hyoid bone.[5,6] Thyroglossal duct cyst may present as a painless midline neck swelling, but in many patients, a painful infected cyst may be the first presenting feature. Others may present as fistula following rupture of the abscess.[7] About 25%–65% of the wall of thyroglossal cysts examined histologically contain thyroid tissue in the form of small thyroid follicles.[8] In very few cases, the entire thyroid tissue fails to descend to its normal location giving rise to an ectopic thyroid gland which may lie in a thyroglossal duct cyst[9] or may be the sole cause of a midline anterior neck swelling.

Ectopic thyroid gland, similar to thyroglossal cyst, can be found at any point along the normal migratory pathway of the thyroid gland and could occasionally extend up to the mediastinum and other parts of the body.[10] It is the most common form of thyroid dysgenesis accounting for up to 61%.[10] It has a prevalence of about 1/100,000–300,000 persons and seen in 4000–8000 patients with thyroid disorder.[11] Lingual thyroid is the most frequently encountered form, up to 90%, of ectopic thyroid.[12] Ectopic thyroid can coexist with orthotopic thyroid gland in about 30% of cases[11] in which there is usually no anatomical or vascular connection between the two.[13] Symptoms, when present, are usually related to the location and size, to abnormal endocrine function, or due to other types of disorders that may affect the normal thyroid.[13]

Address for correspondence: Dr. Hassan Abdullahi Elechi, Department of Paediatrics, University of Maiduguri, Maiduguri, Nigeria.
E-mail: h3elechi@unimaid.edu.ng

Received: 02-10-2020 **Revised:** 24-11-2020 **Accepted:** 24-12-2020 **Available Online:** 30-07-2021

Access this article online

Quick Response Code:

Website: www.afrjpaedsurg.org

DOI: 10.4103/ajps.AJPS_147_20

How to cite this article: Elechi HA. Hypothyroidism following sistrunk procedure: Thyroglossal duct cyst or ectopic thyroid? Afr J Paediatr Surg 2021;18:231-4.

Differentiating a midline ectopic thyroid from thyroglossal duct cyst clinically may be quite unreliable.[14,15] Identifying the orthotopic thyroid gland in such a case becomes an important clinical exercise in anticipation of normal thyroid state after surgery. Ultrasound scan is safe, affordable and the most readily available modality used in evaluating a midline neck mass, and hence, identification of the orthotopic thyroid gland. In some instances, particularly in a poor resource setting like ours may be the only available tool for this purpose.

We report the case of a 2-year 10-month-old boy who had Sistrunk's procedure for ultra sound scan (USS) diagnosed 'thyroglossal duct cyst' and subsequently developed hypothyroidism.

Case Report

A 2 years 10-month-old boy presented with a history of anterior neck swelling of 5 months duration. Swelling was noticed incidentally by the mother with minimal increase in size since first noticed and no associated pain, change in voice or difficulty in swallowing. There was no history of fever, sore throat, ear pain, cough, cold/heat intolerance, constipation or swelling in any other part of the body and no family history of anterior neck swelling. He had umbilical herniorrhaphy 8 months earlier.

He was not pale, acyanosed, afebrile, with a weight of 12 kg (Standard deviation score [SDS]-1.38) and height was not documented. There was a midline swelling on the neck just above the thyroid cartilage, which was non-tender, had no differential warmth, and moved with swallowing and protrusion of the tongue. The swelling measured 2.1 cm × 3 cm in its widest diameters. It was cystic and not attached to the overlying skin, and there was no significant cervical lymphadenopathy. Needle aspiration yielded yellowish brown aspirate of which microscopy, culture and sensitivity showed no pus cell and yielded no bacterial growth.

Ultrasound scan of the neck [Figure 1a] reported a well-defined cystic lesion measuring 2.1 cm × 1.03 cm anterior to the isthmus and above the right and left lobes of the thyroid.

Complete blood count result showed packed cell volume of 29%, total leucocyte count of 15.3 × 10^9/L and differential leucocyte count: Neutrophil – 44% and lymphocytes − 56%. Serum electrolyte, urea and creatinine were essentially within the normal limit.

He had sistrunk procedure and subsequently discharged home. He defaulted follow-up post-operatively, but reported having had recurrent body pain, jaundice and paleness of the palms and soles and was subsequently diagnosed as having sickle cell anaemia in another clinic.

Eighteen months post-operation, he represented with dried and coarse skin with associated poor growth. Physical examination revealed a dull looking child, markedly pale, jaundiced with marked periorbital fullness. He had a weight of 14 kg (SDS-1.67) and a height of 92 cm (SDS-3.10). The result of the histology section of the excised tissue (retrieved only at this point) showed thyroglossal duct cyst lined by stratified squamous epithelium with intraluminal secretions. The duct is surrounded by abundant normal thyroid tissue. Focal areas showing lymphocytic aggregates and skeletal muscle within the wall are also noted. There is no evidence of malignancy [Figure 2].

A working diagnosis of hypothyroidism was made, and investigation results are below:

Thyroid function test: Triiodothyronine (T_3) 0.4 ng/dl (0.6–2.0); thyroxine (T_4) 3.0 µg/dl (5.0–14. 0) and thyroid-stimulating hormone (TSH) 40.0 iu/ml (0.4–7.0).

A repeat USS [Figure 1b] reported empty thyroid bed, while a radiograph of the left wrist and hand [Figure 3] estimated the bone age at 2 years 8 months as against the chronological age of 5 years 6 months. Furthermore, noted on the X-ray was generalised decrease in bone density and multiple growth arrest lines in the distal metaphysis of the radius while packed cell volume was 18%.

He was commenced on L-thyroxine with subsequent normalisation of the thyroid function test. He grew by 8 cm in the first 12 months on drugs and 5–6 cm/year afterwards

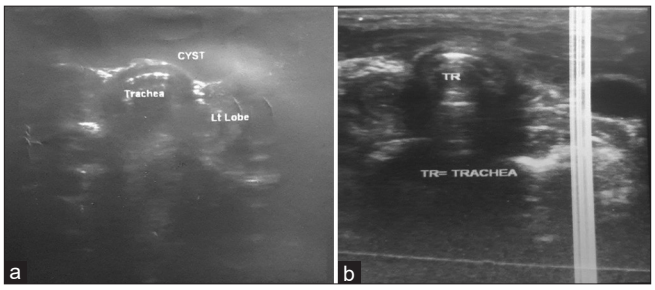

Figure 1: Transverse greyscale sonographic images. (a) Pre-surgery. (b) Post-surgery: apart from the cyst in (a), image appears similar to pre-surgery despite the contrasting reports

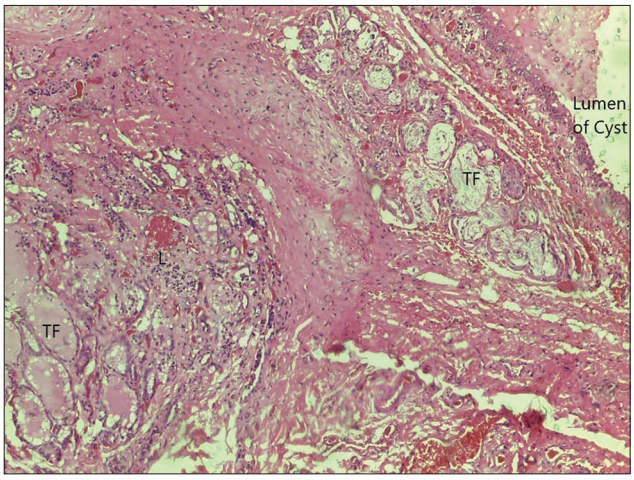

Figure 2: Histology of the excised tissue showing normal thyroid folicles (TF), lymphocyte aggregates 'L', and lumen of the cyst lined by stratified squamous epithelium

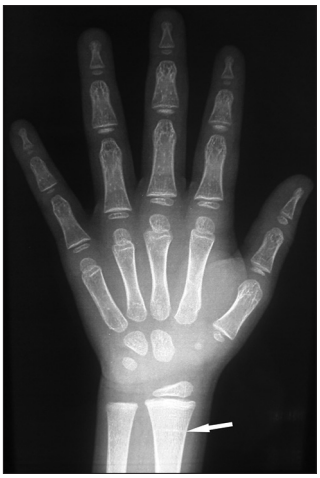

Figure 3: Radiograph of the left hand showing bone age of 2 years 8 months as against chronological age of 5 years 6 months. Also noted are multiple growth arrest lines (white arrow)

and packed cell volume rose to pre-surgery level 27%–29% and height as at last follow-up visit was 115 cm (SDS-2.34).

Discussion

Thyroglossal cyst is the most frequent diagnosis in the evaluation of a child with midline cystic cervical swelling. Other differential diagnoses include submental adenitis, dermoid or sebaceous cysts, lipoma and bronchiogenic cyst. Misdiagnosing any of these differential diagnoses for another and subsequent excision may have little or no long-term clinical consequences for the patient. A rare but extremely clinically important differential diagnosis is ectopic thyroid. This is because of the immense and life-long clinical implication inadvertent removal of a lone functioning ectopic thyroid poses to the child. Accurately identifying the status of the orthotopic thyroid gland is the key to avoiding such occurrence.

The pre-operative USS done in the index case reported identifying the orthotopic thyroid with its various lobes, while the post-operative USS following symptomatic hypothyroidism reported empty thyroid bed. The two scanogram [Figure 1] - pre-operative (a) and post-operative (b) apart from the cyst in the former, appear quite similar despite contrasting reports. While the sensitivity of USS in identifying orthotopic thyroid could be 100%, the same cannot be said of its specificity. Several false-positive identifications of orthotopic thyroid gland using USS have led to excision of ectopic thyroids which were the only functioning thyroid tissue in the body.[16-18] Holland *et al*.[16] reported a similar case where a pre-operative USS demonstrated orthotropic thyroid and hence excision of the presumed 'thyroglossal duct cyst' but post-operative radioisotope thyroid scan showed empty thyroid bed. In situations where the orthotopic thyroid gland is completely absent, the infrahyoid muscles occupying the thyroid bed are known to resemble thyroid tissue on a transverse USS and could easily be reported as such.[16]

The index case was 2 years 10 months of age at diagnosis. This is within the age range of 1–4 years of most reported cases of ectopic thyroid tissue presenting as a new neck mass in children.[14,15,19-22] Others have reported similar cases in the adolescent age.[16,23] This bimodal presentation (early childhood and adolescence) is thought to be due to the inability of the poorly developed ectopic thyroid to meet the increasing demand for thyroid hormone associated with rapid growth and development during these stages. This increase demand in the phase of underdeveloped ectopic thyroid leads to increase TSH drive and hence the enlargement.[15] This implies that ectopic thyroid could be a significant cause of anterior midline neck swelling presenting at early childhood or adolescence. We thus suggest pre-operative thyroid function test in conjunction with USS in the evaluation of anterior midline mass presenting in early childhood and adolescence. It is believed that the few with enlarged lone functioning ectopic thyroid tissue will have abnormal thyroid function test and thus will indicate the need for a closer attention and scrutiny during USS.

The swelling was noticed incidentally which is in keeping with normal clinical pattern of ectopic midline cervical thyroid in which the usual presenting complaint is painless neck swelling. This is also true for uninfected thyroglossal cyst. In many cases of thyroglossal duct cyst, pain may be the first symptom pointing to an ongoing inflammation or discharging fistula in case of rupture.[7] The swelling was located just above the thyroid cartilage which is along the course of thyroid migration, but above the normal location of orthotopic thyroid gland and hence could not have been anterior to a normally located isthmus as suggested by the pre-operative USS. Although ectopic thyroid could be located at any point along the migration path and beyond, it is lingual in 90% of the time.[12] This location is more in keeping with thyroglossal cyst which is more frequently located below the hyoid.[5,6] Similar to the findings of Sugiyama *et al*.,[24] needle aspirate yielded a yellowish-brown fluid which neither yielded pus cells on microscopy nor bacterial growth on culture.

The weight-for-age Z-score at both initial presentation and diagnosis of hypothyroidism was within the normal range. Although the pre-operative height was not recorded, poor linear growth among other symptoms was the presenting complaint at the diagnosis of hypothyroidism, and the recorded height was 3.1 SDS. This, however, improved following commencement of L-thyroxine with accelerated growth rate in the 1st year and current height is at −2.34 SDS. It is a common knowledge that acquired hypothyroidism in a growing child will lead to deceleration in linear growth and weight gain. It is not clear weather hypothyroidism for a short duration of 18 months could account for such a marked lag in height, but the concomitant chronic anaemia from sickle cell disease could have contributed. In addition, the enlargement of the ectopic thyroid at presentation could suggest suboptimal functioning gland as reported by Hanmayyagari *et al*.,[25] although pre-operative thyroid function test was not done. This assumption may be supported by the delayed bone age

at the diagnosis of hypothyroidism which was approximately 3 years behind the chronological age with multiple growth arrest line in the distal radius suggesting that impaired linear growth may predate the surgery.

The histology result showed features consistent with thyroglossal duct cyst surrounded by normal thyroid tissue. Lilley *et al*.[21] also reported similar histology finding. Other reports have documented thyroglossal duct cyst occurring in an ectopic thyroid[7] and orthotopic thyroid.[26] This finding further highlights the difficulty in differentiating between thyroglossal duct cyst and ectopic thyroid based on the location and clinical appearance. Unfortunately, the index patient defaulted follow-up and the histology report was not retrieved until 18 months later and hence the delay in commencement of L-thyroxine.

Features of sickle cell anaemia became obvious post-operatively eventually leading to confirmatory diagnosis. This may be explained by the important role of thyroxine in erythropoiesis, and the effect of its deficiency is likely to be more marked in sickle cell anaemia due to the increase red blood cell turnover rate. This is further confirmed by the return of steady state packed cell volume to pre-operative levels following commencement of L-thyroxine.

Conclusion

Ectopic thyroid gland can mimic or lie within a thyroglossal duct cyst more frequently in early childhood and adolescent. In many such instances, it may be the only functioning thyroid tissue in the body. Accurately identifying the orthotopic thyroid gland is an important step in excluding a lone functioning ectopic thyroid tissue. Ultrasound scan, which is the most commonly used tool for this purpose, may yield false-positive report in those with no orthotopic thyroid gland due to the anatomical and operator factors. Pre-operative basic thyroid function test in combination with USS could help pick up these false-positive reports and may be quite reassuring for both the doctor and patient.

Acknowledgement

I wish to acknowledge the staff of Paediatric Surgery, Histopathology and Radiology units for assisting with the materials necessary for preparing the manuscript.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Sadler TW. Langman's Medical Embryology. 9th ed. Philadelphia: Lippincott, Williams and Wilkins; 2004.
2. Blackburn ST, editor. Pituitary, adrenal and thyroid function. In: Maternal, Fetal, & Neonatal Physiology: A Clinical Perspective. 4th ed. Maryland Heights: Elsevier Saunders; 2013.
3. Delwar AH, Mazumder JA, Rashid MS, Mustafa MG, Hossain AB. Incudence and outcome of thyroglossal cyst. Int J Otorhinolaryngol Head Neck Surg 2020;6:1-4.
4. Kurt A, Ortug C, Aydar Y, Ortug G. An incidence study on thyroglossal duct cysts in adults. Saudi Med J 2007;28:593-7.
5. Santiago W, Rybak LP, Bass RM. Thyroglossal duct cyst of the tongue. J Otolaryngol 1985;14:261-4.
6. Patigaroo SA, Dar NH, Jallu AS, Ahmad R. Thyroglossal duct cysts: A clinicosurgical experience. Indian J Otolaryngol Head Neck Surg 2017;69:102-7.
7. Thompson LD, Herrera HB, Lau SK. A clinicopathologic series of 685 thyroglossal duct remnant cysts. Head Neck Pathol 2016;10:465-74.
8. Adelchi C, Mara P, Melissa L, De Stefano A, Cesare M. Ectopic thyroid tissue in the head and neck: A case series. BMC Res Notes 2014;7:790.
9. Shin AY, Lee SH, Jung WS, Ko SH, Ahn YB. Ectopic thyroid nodule in thyroglossal duct. Korean J Intern Med 2011;26:218-9.
10. Noussios G, Anagnostis P, Goulis DG, Lappas D, Natsis K. Ectopic thyroid tissue: Anatomical, clinical, and surgical implications of a rare entity. Eur J Endocrinol 2011;165:375-82.
11. Erich N, Mussak BS, Kacker A. Surgical and medical management of midline ectopic thyroid. Otolaryngol Head Neck Surg 2007;136:870-2.
12. Larochelle D, Arcand P, Belzile M, Gagnon NB. Ectopic thyroid tissue – A review of the literature. J Otolaryngol 1979;8:523-30.
13. Santangelo G, Pellino G, De Falco N, Colella G, D'Amato S, Maglione MG, *et al*. Prevalence, diagnosis and management of ectopic thyroid glands. Int J Surg 2016;28 Suppl 1:S1-6.
14. Lewis MI, Holleran WM. Ectopic thyroid gland in children. Am J Surg 1968;115:688-90.
15. Conklin WT, Davis RM, Dabb RW, Reilly CM. Hypothyroidism following removal of a "thyroglossal duct cyst". Plast Reconstr Surg 1981;68:930-2.
16. Holland AJ, Sparnon AL, LeQuesne GW. Thyroglossal duct cyst or ectopic thyroid gland? J Paediatr Child Health 1997;33:346-8.
17. Takashima S, Nomura N, Tanaka H, Itoh Y, Miki K, Harada T. Congenital hypothyroidism: Assessment with ultrasound. AJNR Am J Neuroradiol 1995;16:1117-23.
18. Muir A, Daneman D, Daneman A, Ehrlich R. Thyroid scanning, ultrasound, and serum thyroglobulin in determining the origin of congenital hypothyroidism. Am J Dis Child 1988;142:214-6.
19. Di Benedetto V. Ectopic thyroid gland in the submandibular region simulating a thyroglossal duct cyst: A case report. J Pediatr Surg 1997;32:1745-6.
20. Damiano A, Glickman AB, Rubin JS, Cohen AF. Ectopic thyroid tissue presenting as a midline neck mass. Int J Pediatr Otorhinolaryngol 1996;34:141-8.
21. Lilley JS, Lomenick JP. Delayed diagnosis of hypothyroidism following excision of a thyroglossal duct cyst. J Pediatr 2013;162:427-8.
22. Carlson S, Campbell L, Janitz E. Ectopic thyroid tissue presenting as a new neck mass in a pediatric patient. Radiol Case Rep 2019;14:55-7.
23. Popli MB, Chaudhary A, Imam N. Midline ectopic thyroid mimicking as thyroglossal duct cyst. J Diagn Med Sonogr 2015;31:190-3.
24. Sugiyama S, Iwai T, Hirota M, Mitsudo K. Ectopic thyroid cyst of the lateral neck. Ear Nose Throat J [Serial on the Internet]. 2020 Jun [cited 2020 Sep 30]: [about 3 p.]. Available from: https://journals.sagepub.com/doi/full/10.1177/0145561320937080.
25. Hanmayyagari B, Guntaka M, Ravilala VK. Ectopic thyroid in presumed thyroglossal duct cyst. Indian Pediatr 2013;50:420-1.
26. Huang LD, Gao SQ, Dai RJ, Chen DD, He B, Shi HQ, *et al*. Intra-thyroid thyroglossal duct cyst: A case report and review of literature. Int J Clin Exp Pathol 2015;8:7229-33.