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Congenital thyroid hemiagenesis with thyroid nodules—Role of TI-RADS to prevent long term thyroid replacement therapy

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ABSTRACT

BACKGROUND: Hemi-agenesis of thyroid is a rare congenital condition with the clinical significance lying only in cases where the remnant tissue is affected by a pathology mandating removal of the gland. Henceforth, the hemi-thyroidectomy technically becomes a total thyroidectomy with a need for long term thyroid replacement therapy.

CASE SUMMARY: We present a series of three cases noted over a period of two years where preoperative imaging evaluation confirmed the developmental abnormality. Further presence of a thyroid nodule in each of these cases posed a specific clinical situation whereby characterization of nodule appeared mandatory for a rational management involving life-long thyroid replacement therapy due to an 'apparent total thyroidectomy', if the remnant gland is removed. Ultrasound TI-RADS is a new system for evidence based sub-classification of thyroid nodules enabling both the surgeon and patient to take a streamlined decision about the overall approach for management.

CONCLUSION: Prospective nodule characterization based on the thyroid image reporting and data system (TI-RADS), enables the surgeon to decide the treatment strategy sparing the patient of the cost and morbidity associated with long term thyroid replacement therapy.

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1. Introduction

Hemiagenesis of thyroid first described about a century and half ago still remains a rare and a challenging clinical situation [1]. Rare because most cases remain asymptomatic, a single lobe of thyroid being enough to fulfil the needs of human body, and challenging when this solitary lobe is affected by a pathology which mandates removal of the gland [1,2]. In the latter situation a potential hemithyroidectomy (if the person have had both lobes) gets converted into a total thyroidectomy. In many of these cases the resected thyroid specimen shows no histological sign of by malignancy, still subjecting the patient to effects and side-effects of long term thyroid replacement therapy (TRT). Thyroid image reporting and data system (TI-RADS) was recently introduced on lines similar as breast image reporting and data system (BI-RADS) for risk stratification of thyroid nodules towards malignancy [3]. We present an introspective analysis in three cases of thyroid hemi-agenesis where surgical removal with long term TRT was done irrespective of the cases being assessed as benign by TI-RADS. The aim is to evaluate the pro-and-cons of treatments done in light of TI-RADS with emphasis on the effects of long term TRT.

2. Case series

The presentation in all our patients (Table 1) was swelling in the neck with no symptoms referable to thyrotoxic state. On examination a nodule measuring about 3 cm was noted in the region of right thyroid fossa which was mobile in all planes. No fixity with the trachea was noted while free movement was seen on deglutition. No cervical adenopathy, enlargement of salivary glands or any other neck nodule were palpable. The structures of carotid sheath could be palpated free of the nodule while no trophic changes were noted on the skin overlying the front or nape of neck. Suspecting the nodule to be from the thyroid gland a radionuclide scan was done whereby the first patient had a hot nodule while no activity was picked up on the other two patients. Ultrasonography was done as the first-line cross sectional imaging and revealed absence of left lobe & isthmus in all patients, while pathology was noted in the right lobe (Table 1). TI-RADS evaluation labelled all nodules to be benign in nature (TI-RADS 2) hence the patients were counselled to undergo fine needle aspiration cytology but they refused. All three patients chose the surgical option due to the statistical risk of cancer [4] in an unevaluated thyroid nodule. CT angiography was done in the first two patients (Figs. 1 and 2) to confirm the absence of thyroid arteries but was omitted subsequently (sonographic images of third patient not shown), also the same would not have altered the management protocol planned. Thyroidectomy was done as per standardized technique, none of the patients revealed any his-

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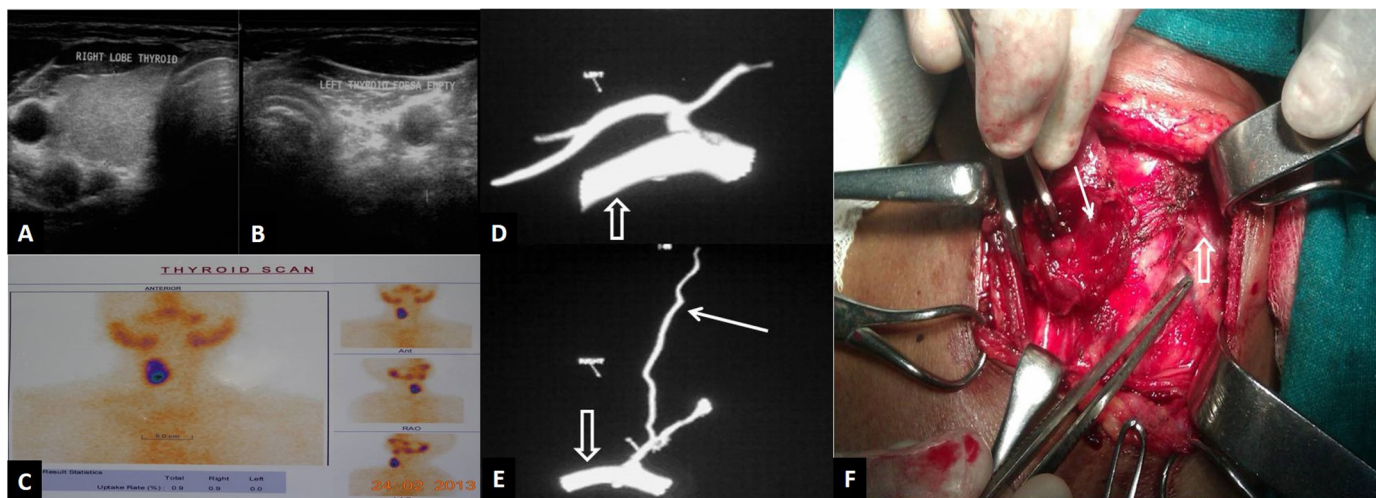


Fig. 1. (A, B) High frequency ultrasonography thyroid tissue in the right thyroid fossa with an empty left thyroid fossa. (C) Thyroid scintigraphy revealed absence of left lobe and isthmus with normal uptake on the right side. (D, E) Focused reconstruction of the first part of subclavian artery (open arrows) on CT-Angiography reveals underdeveloped left inferior thyroid artery. Note the corresponding normal inferior thyroid artery on the right side (straight arrow) (F) Operative photograph shows bulky nodular right lobe of thyroid (solid arrow). Note the left thyroid fossa is empty (open arrow).



Fig. 2. (A, B) Pre-contrast and post-contrast CT scan images showing presence of the left lobe and isthmus with a nodule in the substance (straight arrow). (C) 3-D angiographic reconstruction showing presence of left superior thyroid artery while corresponding artery on the right is not seen (solid arrow).

tological sign of malignancy on histopathology of the operative specimen. Long term thyroid replacement therapy was instituted which was tolerated well by two patients while the third patient presented with oral dryness and dizziness. This patient was given a drug holiday following which the therapy the dosage protocol was re-titrated to manage the drug intolerance.

In a small pilot study of three cases we found a 100% sensitivity and specificity of imaging features that are combined to give an ultrasonographic TI-RADS scoring. In each of the patients a blinded scoring was initially done which correlated well with the surgical impression along with excluding any observer bias. Though the sample size is small and the group consists of patients with a single entity only, it may be noted that consecutive cases were selected hence obviating any chances of selection bias. The diagnostic accuracy of the scoring system turns out to be 100% in the small sample. The present series has been tailored as per the CARE guidelines.

3. Discussion

Ultrasonography based TI-RADS incorporates certain specific imaging features which show a significant association with malignancy like presence of solid component within a cyst, markedly hypoechoic solid lesion, micro-lobulations or irregular margins, micro-calcifications, and lesion being taller-than-wide in shape. Addition of each feature increases the confidence of diagnosis and

accuracy of the lesion classification [3,5]. In a study by Sánchez JF, all lesions showing features of benignity (TI-RADS 2 and 3) had a score of zero. Only 2.2% of TI-RADS 3 patients proved to be malignant. Patients with TI-RADS 4 (a,b or c) lesions had scores varying from one to four with malignancy rates between 9.5%–85%. Hundred percent malignancy rate was seen in TI-RADS 5 (score of five or more points) patients [6]. The life time risk of malignancy in patients up to TI-RADS 3 remains negligible and akin to that in normal population [5,6]. This would mean that with a score of zero in all our patients (TI-RADS 2) there would not have been any added risk of malignancy and the surgery done was purely for cosmetic reasons. In such circumstances it may be more practical to give the patient an option between the surgery with long term TRT versus watchful expectancy with periodic reclassification by TI-RADS, similar to the protocol followed in BI-RADS [7]. The suggestion is subsequent to the fact that most patients with long term TRT complain of one of the side effects of the same like [8]. The third patient in our series complained of dizziness and oral dryness which was managed by re-titration of the dose. This kind of a protocol is specific to the situation of thyroid hemi-agenesis with goitrous remnant tissue and should not be extrapolated to other clinical situations. Partial agenesis of thyroid gland, in various studies has been labelled to be as common as approximately 1 in 2000 in certain surgical and autopsy series [9]. Most of such patients are euthyroid at presentation and similar to what we encountered, agenesis of left lobe is four times

Table 1
Clinical, investigative and operative details of the patients included in the series.

Sl. No.	Age (Years)	Sex	Chief Complaint	Thyroid Function status	Status of thyroid (on sonography)	Sonographic evaluation of nodule	Proposed TI-RADS Score (Retrospective)	C:T Scan Features	Management done	Histological Result (On surgery)	Follow up
Patient 1	40	Female	longstanding gradually progressive neck swelling	Euthyroid	Right lobe normal dimensions; left lobe and isthmus absent	3.5 cm size solitary nodule; Solid with multiple hypoechoic areas	2	Absent left lobe with absent inferior thyroid artery	Removal of the remnant thyroid lobe/tissue	Adenomatous goiter with degenerative changes	Unremarkable at 6 months
Patient 2	33	Female	longstanding non-progressive neck swelling	Euthyroid	Right lobe normal dimensions; left lobe and isthmus absent	4 cm size solitary nodule; Multi-septate cystic	2	Absent left lobe with absent superior/inferior thyroid artery	Removal of the remnant thyroid lobe/tissue	Adenomatous goiter with degenerative changes	Unremarkable at 4 months
Patient 3	41	Female	longstanding initially gradually then non-progressive neck swelling	Euthyroid	Right lobe normal dimensions; left lobe and isthmus absent	3.7 cm size largest nodule with few small well defined hypoechoic areas in the gland; Main nodule being solid with multiple hypoechoic areas	2	Not done	Removal of the remnant thyroid lobe/tissue	Adenomatous goiter with degenerative changes	Immediate post-operative only

as common as that of right lobe. Isthmus is involved in about half of all cases. Women are thrice as commonly affected, notably all our patients were ladies, this may propose a gender association, the basis of which has not however been proven till date [10]. Due to the above fact there may be a case for cosmetic intervention of the patient consciously chooses to opt for the same accepting the effects of TRT. The patients in present series presented with multinodular goiter which remains the second most common cause for presentation after hyperthyroidism. Thyroid scintigraphy remains the modality for confirmation of diagnosis but it may be noted that mimics of hemi agenesis like autonomous functioning nodule with suppressed normal thyroid tissue, infiltrative disease such as amyloidosis, primary and secondary neoplasm and unilateral inflammation, on this modality may give false positive results [11]. Absence of inferior and/or superior thyroid arteries with absence of thyroid tissue seems to be the most reliable tool for confirmation of the diagnosis. In the first case we noted an absence of inferior thyroid artery of the left side while in second case both the arteries were absent [11,12]. CT scan however remains an expensive investigation with risk of radiation and contrast injection, when compared to high resolution sonography. We however omitted CT angiography in the third case as the diagnosis was established by color Doppler sonography and the surgeon wanted to go forward with the surgery anyways.

Goiter in a case of thyroid hemiagenesis may pose a difficult and rare clinical situation to manage. The importance of ultrasound TI-RADS from the patient perspective lies in the fact that being a useful tool to decide the treatment protocol in such patient it may spare most of the long term effects of thyroid replacement therapy. However, all our patients chose to undergo thyroidectomy after being counselled on the basis of the said protocol, and hence were prospectively convinced in favor of the long term replacement therapy. Though consisting of a small sample, it may be noted that the observations made in this series may bear importance on deciding the management in future cases in our as well as at other institutions as this situation is quite rare. Larger series conducted over longer durations would be required to come up with more confident guidelines having a definite statistical basis.

Conflict of interest

None.

Funding

None.

Ethical approval

This is a clinical data compilation taken from routine clinical work. The work was done as per established evidence based protocol for treatment of this disease entity.

Consent

Consent for treatment was taken.

Disclosures

Patient informed consent – obtained: IRB approval – clinical protocol as per evidence based guidelines acceptable in institute.

Author contribution

All authors have contributed significantly and equally.

Guarantor

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References

- [1] S.K. Bhartiya, A. Verma, S.P. Basu, V.K. Shukla, Congenital thyroid hemiagenesis with multinodular goiter, *Acta Radiol. Short Rep.* 3 (9) (2014) 1–4.
- [2] W. Shabana, F. Delange, M. Freson, J. DeSchepper, Prevalence of thyroid hemigenesis: ultrasound screening in normal children, *Eur. J. Pediatr.* 159 (2000) 496–498.
- [3] J.Y. Kwak, K.H. Han, J.H. Yoon, H.J. Moon, E.J. Son, S.H. Park, H.K. Jung, J.S. Choi, B.M. Kim, E.K. Kim, Thyroid imaging reporting and data system for US features of nodules: a step in establishing better stratification of cancer risk, *Radiology* 260 (3) (2011) 892–899.
- [4] S.C. Kamran, et al., Thyroid nodule size and prediction of cancer, *J. Clin. Endocrinol. Metab.* 98 (2013) 564–570.
- [5] J. Fernández Sánchez, TI-RADS classification of thyroid nodules based on a score modified according to ultrasound criteria for malignancy, *Rev. Argent. Radiol.* 78 (3) (2014) 138–148.
- [6] G.H. Tan, H. Gharib, Thyroid incidentalomas: management approaches to nonpalpable nodules discovered incidentally on thyroid imaging, *Ann. Intern. Med.* 126 (1997) 226–231.
- [7] L. Hegedus, Clinical practice. The thyroid nodule, *N. Engl. J. Med.* 351 (2004) 1764–1771.
- [8] J.A. Franklyn, Thyroid disease and its treatment: short- and long-term consequences, *J. R. Coll. Physicians Lond.* 33 (6) (1999) 564–567.
- [9] C.R. Mchenery, P.G. Walfish, I.B. Rosen, A.M. Lawrence, E. Paloyan, Congenital thyroid hemi-agenesis, *Am. Surg.* 61 (1995) 634–638.
- [10] S. Peña, H. Robertson, R.R. Walvekar, Thyroid hemiagenesis: report of a case and review of literature, *Indian J. Otolaryngol. Head Neck Surg.* 63 (2) (2011) 198–200.
- [11] S.D. Sarkar, Benign thyroid disease: what is the role of nuclear medicine? *Semin. Nucl. Med.* 36 (3) (2006) 185–193.
- [12] P. Gupta, A.S. Bhalla, S. Thulkar, A. Kumar, B.K. Mohanti, A. Thakar, A. Sharma, Variations in superior thyroid artery: a selective angiographic study, *Indian J. Radiol. Imaging* 24 (1) (2014) 66–71.

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