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Case report

Thyroid hemiagenesis associated with Hurthle cell carcinoma: A case report

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ABSTRACT

Introduction and importance: Thyroid hemiagenesis (TH) is a rare congenital anomaly where one lobe fails to develop, especially more frequently occurs on the left lobe. The exact mechanisms for thyroid morphogenesis remain unclear. In this paper, we report a rare case of right lobe TH associated with Hurthle cell carcinoma.
Case presentation: A 59 years old woman was admitted with a neck lump increasing in size in the last 20 years. There were no symptoms of hyperthyroidism and hypothyroidism. There was a palpable, painless 5 cm mass in the middle of the neck. Initial thyroid ultrasonography (USG) revealed an enlarged left lobe, with hypoechoic lesion with cystic component and calcification (TIRADS 4). However, the right lobe was non-visualized. Fine needle aspiration biopsy result tendency was a malignancy. Hence, isthmolobectomy was conducted. Pathology result was Hurthle cell carcinoma. On the ninth month, USG revealed fibrotic tissue in the right thyroid bed and bilateral lymphadenopathy. Due to discrepancy, the patient was planned for a neck exploration surgery and a right lobe incision. Intraoperatively, the right thyroid was absent. Intraoperative USG also confirmed no right thyroid lobe.
Discussion: Thyroid hemiagenesis can be visualized by using USG due to its practicality and cost effectiveness reasons. Follow up evaluations consisted of systematic monitoring of thyroid morphology and hormonal functions should follow the diagnosis of TH. Neck exploration surgery might need to be performed to clarify any discrepancy and confirm the diagnosis.
Conclusion: TH can be recognized through supporting examination; however, discrepancy may occur.

1. Introduction

During the embryologic development, a thyroid gland passes through a process of migration, descent, lobulation, and differentiation continuously [1]. This process is organized by various hormonal and genetic factors. Thyroid hemiagenesis is rare congenital abnormality where there is an absence of one thyroid lobe while the other lobe, with or without an isthmus, normally develops [1,2]. TH is one of the mild thyroid developmental anomalies including thyroglossal duct and pyramidal lobe cysts. The precise etiology of TH remains being unknown, although familial and sporadic cases have been described [1]. The suggested mechanism is the bi-lobulation failure of the central anlage with the thyroid tissue migrating to one side of the midline [1]. It is considered as a benign abnormality with no treatment required since a single lobe is sufficient to keep euthyroidism condition of a person [2]. However, some studies reported thyroid nodules and autoimmune thyroid diseases as the most common abnormalities accompanying TH [2]. Hence, early and prompt diagnosis and follow-up is necessary.

The prevalence of TH is 0.05%–0.2% based on ultrasound examinations [3,4]. Nevertheless, the total number of cases might underestimate the real incidence of TH. This due to the fact that TH is usually incidentally found during the investigation or follow up of accompanying thyroid disease [1]. Functional and morphological thyroid disorders are commonly associated with TH [2]. Thyroid malignancies are rarely found with TH cases. Based on a literature review, thyroid malignancies were reported in only 3% of TH cases [5]. Similarly, ectopic thyroid tissues were also reported in 3% of cases [6]. It is more common in female patients and the majority includes left lobe hemiagenesis comprising more than 80% of all diagnosed cases [2,5,7].

Up to now, there is no established consensus on optimal management

Abbreviations: TH, thyroid hemiagenesis; USG, ultrasonography; FNAB, fine needle aspiration biopsy; CT, Computed Tomography.

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of TH perhaps because of its small number of cases [8]. Whether TH predisposes to the thyroid diseases requiring therapy or whether TH poses clinically insignificant role are subjects to controversy. Hence, we report a rare case of TH associated with Hurthle cell carcinoma and how we had managed it. The work has been reported in line with the SCARE 2020 criteria [9].

2. Case presentation

We report a case of a 59 years old woman who was admitted to our hospital with a chief complaint of a neck lump that has persisted in the last 20 years before admission. The initial size was 1 cm and slowly enlarged to 5 cm in 20 years. Symptoms of hyperthyroidism or hypothyroidism were denied. Moreover, there weren't any complains of dyspnea, hoarseness, and swallowing problem.

Based on the physical examination, a palpable and painless mass was located in the middle of the neck. All of her vital signs were stable. Her ion calcium, serum calcium, ft4, and TSH were normal. However, her thyroglobulin level was high (564.5 ng/mL).

Thyroid ultrasonography (USG) depicted an enlarged left lobe accompanied by hypoechoic lesion with cystic component and calcification (TIRADS 4) (Fig. 1). There were ovoid lymph nodes visualized in left level 1A and bilateral 1B neck region with less than 1 cm size. However, the right thyroid was difficult to evaluate. Color Doppler was done confirming the lesion's vascularization. Fine needle aspiration biopsy (FNAB) result was inconclusive, but raised suspicion towards malignancy. Hence, the patient was initially diagnosed with suspected malignant left nontoxic goiter.

Thereafter, she undergone isthmolobectomy procedure. Collar incision was performed to start the procedure, followed by cranial and caudal flap. Left thyroid lobe was removed after necessary artery and vein ligations followed by recurrent laryngeal nerve and parathyroid preservations (Fig. 2). The intraoperative frozen section evaluation revealed Hurthle cell neoplasm. Hence, we did not explore the contralateral lobe since there is not yet any confirmation of carcinoma diagnosis. Postoperative anatomical pathology result confirmed the Hurthle cell carcinoma (Fig. 3). Therefore, A completion surgery was scheduled to conduct.

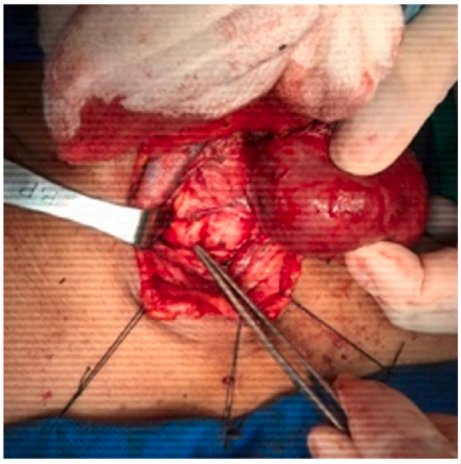

Fig. 2. Left thyroid lobe and its isthmus were identified and removed in the first surgery.

There was no residual lesion detected until the fourth month of follow up with 1 × 100 mg thyrax consumption per day. After 9 months, USG evaluation showed fibrotic tissue in the right thyroid bed and bilateral lymphadenopathy. There were lymph nodes enlargements with ovoid shape, hilus center, and a 1.5 cm largest diameter on the bilateral level IB neck area. Thereafter, the patient undergone neck exploration surgery. An excision of the right lobe was planned due to the malignant pathology result. The same collar incision as the previous surgery was performed before making cranial and caudal flaps. Strap muscle was opened laterally from the midline due to some difficulties. Nevertheless, the right thyroid lobe was absent intraoperatively (Fig. 4). Moreover, intraoperative USG confirmed no thyroid tissue was found in the right lobe. Hence, the incision was closed leaving one drain to finish the neck exploration surgery. Due to the lymph nodes appearance in the preoperative USG (Ovoid shape and hilus center) and no lymph node

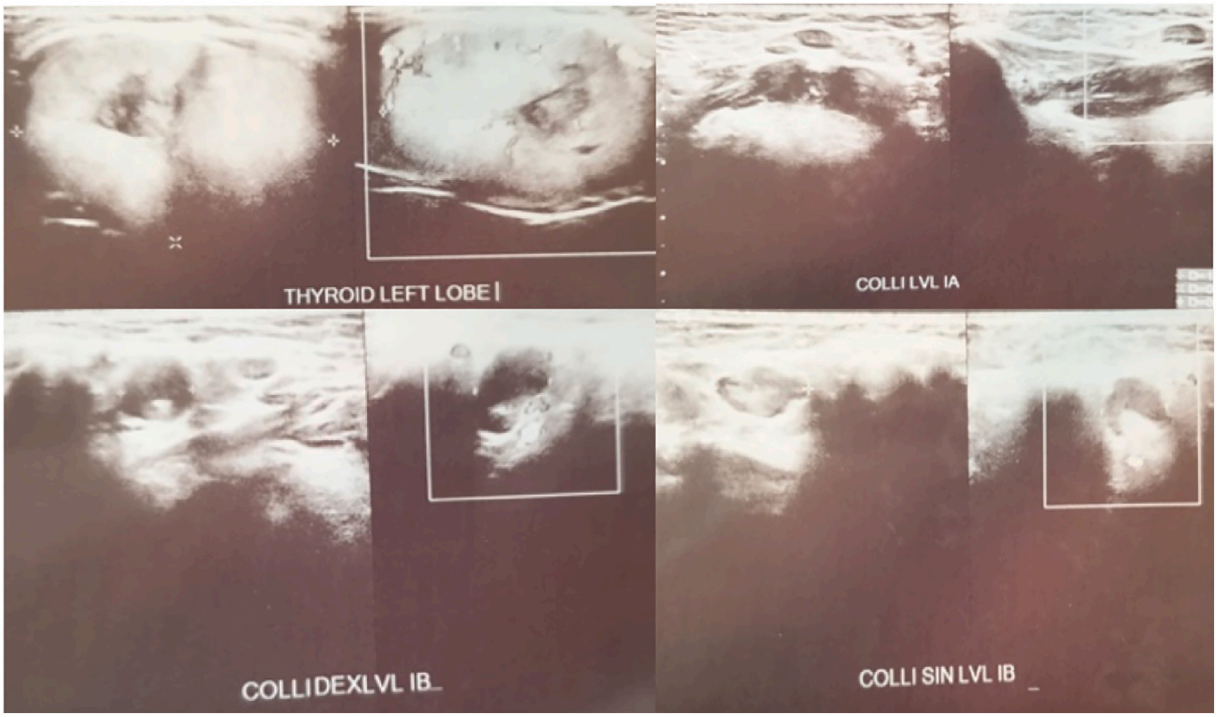

Fig. 1. Initial thyroid ultrasonography showing hypoechoic lesion with cystic component and calcification on enlarged left lobe thyroid (TIRADS 4).

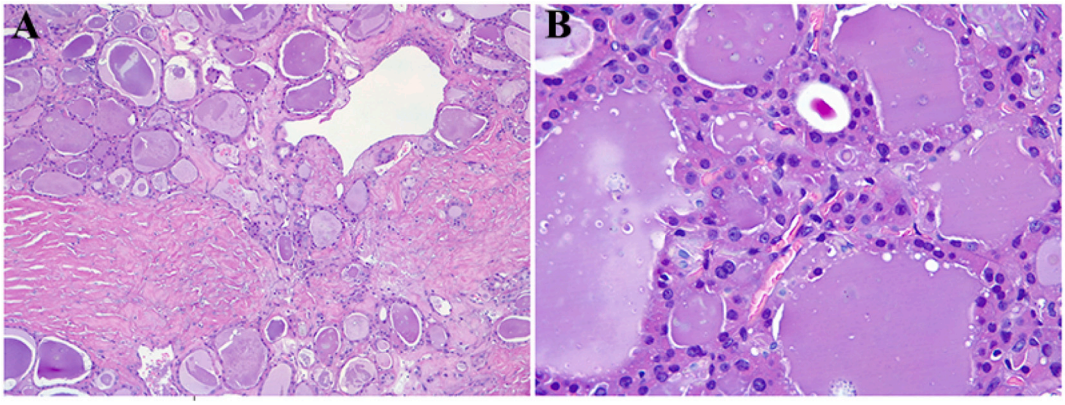

Fig. 3. Postoperative histopathology result of the first surgery. A. Tumor cells arranged in follicular structures invaded the fibrous capsule. B. Tumor cells showed mild to moderate nuclei pleomorphism, macronucleoli and eosinophilic, granular cytoplasms. These morphologic features are compatible with Hurthle cell carcinoma.

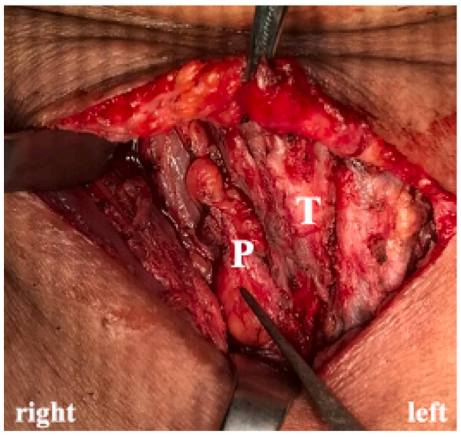

Fig. 4. Parathyroid gland was found instead of right thyroid lobe intraoperatively in the neck exploration procedure. P: parathyroid gland, T: trachea.

identified intraoperatively, we did not perform any lymph node dissection. In the follow up sessions, there were not any signs and symptoms on this patient. The patient's treatment was continued with routine suppression dose of euthyrax therapy.

3. Discussion

Thyroid hemiagenesis is a rare congenital anomaly with varying prevalence from 0.05% to 0.2% [3,4]. Based on the previous literatures, TH is more often in women than men and more frequently on the left thyroid lobe [10]. On the other hand, our case of hemiagenesis was identified on the right thyroid lobe of a woman.

The thyroid diseases found accompanying TH cases is varied with age [2]. Hence this leads to probability that there is a consequence caused by longer exposure of hemiagenetic thyroid gland to overstimulation of TSH in older patients [11]. Therefore, follow up evaluations consisted of systematic monitoring of thyroid morphology and hormonal functions should follow the diagnosis of TH.

There were high varieties of clinical and biochemical presentations of TH. Thyroid dysfunction was reported in 38-47% of cases [2,5]. Hence, a thorough follow up evaluation is warrant for each patient. Ruchala et al. also suggested that patients suspected with TH need to be carefully followed up since the thyroid pathologies are likely to develop [2].

Hemiagenesis might be a consequence of a descent impairment or lobulation defect [12]. If there is no compensatory growth of the existing lobe in its normal anatomic location, the main cause of mechanism is the lobulation impairment instead of descent abnormality [13].

The absence of a thyroid lobe usually results in no clinical symptoms but frequently coincidently diagnosed through evaluations of the other thyroid abnormalities [12]. In some healthy people, thyroid gland can be asymmetric, with the majority of the right lobe being larger than the left lobe [14,15]. Concurrent thyroid disease is often found in TH. Coexistent thyroid diseases accompanying TH includes Grave's disease, thyroiditis, adenoma, nodular goiter, and carcinomas (including primary or metastatic cancer) [13]. In our case, Hurthle cell carcinoma was the coexisting thyroid disease.

Diagnosis of TH depends on suspicion when physical examination or thyroid imaging shows no apparent thyroid tissue on one side. USG with 7.5–12 MHz frequency is one of widely available modality to examine thyroid [16]. This is commonly used because USG is easily performed, cost effective, and without any radiation exposure [17]. Thyroid USG can visualize lobe absence and presence of an isthmus. Its utility and popularity has grown due to its practicality. Thyroid scan also usually used by observing tracer accumulation in just one side of thyroid even after thyrotropin usage [13]. However, primary or secondary malignancies, amyloidosis, and unilateral inflammations can mimic TH existence [18]. Hence, thyroid scans need to be confirmed by another modality to evaluate the exact thyroid morphology. Unfortunately, we only visualize TH in this case by using USG due to its practicality and cost effectiveness reasons. Besides, there are other modalities to visualize TH such as Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) [8]. However, both modalities are expensive and time consuming with comparable results with thyroid USG [8].

In the first isthmolobectomy surgery, we did not explore the contralateral lobe unless the carcinoma diagnosis is confirmed through the frozen section result. Nevertheless, it would be more suitable if contralateral exploration was conducted since in the first surgery to confirm the thyroid hemiagenesis. In the ninth month of evaluation, thyroid USG evaluation revealed fibrotic tissue in the right thyroid bed and bilateral lymphadenopathy, although there was no recurrence complains. This is in line with a study by Azmat et al., which stated a key finding of thyroid cancer accompanying TH suspicion is a metastatic cervical node.6 However, the association between TH and thyroid cancer is uncommon [6]. Moreover, the possibility of an ectopic thyroid tissue is also need to consider. USG is known to be operator dependent [8]. Therefore, discrepancy may occur as found in this study. Despite TH is considered as a benign entity, being unaware of its existence might cause incorrect assumptions during treatment decision-making [12]. Due to this discrepancy, we decided to perform neck exploration surgery to clarify whether there are unresolved abnormalities. TH was finally confirmed when there is no thyroid lobe intraoperatively. Instead, we

only found parathyroid gland in that plane.

4. Conclusions

Thorough history taking and physical examination might not sufficient in establishing the diagnosis of TH accompanying thyroid diseases. Thyroid hemiagenesis recognition can be supported by an imaging modality, especially ultrasonography. However, discrepancy may occur. Hence, an exploration might need to be performed to clarify the discrepancy and confirm the diagnosis.

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Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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This type of study does not require any ethical approval at our institution.

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CRediT authorship contribution statement

Diani Kartini: Concept and design of case report, data collection, drafting, revision, approval of final manuscript
Sonar Soni Panigoro: Concept and design, data collection, revision, approval of final manuscript
Maria Fransisca Ham: Data collection, histopathology interpretation, revision, approval of final manuscript
Agnes Stephanie Harahap: Data collection, histopathology interpretation, revision, approval of final manuscript
Sani Hadiyan Rasyid: Data collection, revision, approval of final manuscript
Kevin Varian Marcevianto: Data collection, revision, approval of final manuscript.

Declaration of competing interest

The authors declare that there is no conflict of interest in this case report.

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References

[1] V. Sanctis, A. Soliman, S. Di Maio, H. Elsedfy, N. Soliman, R. Elalaily, Thyroid hemiagenesis from childhood to adulthood: review of literature and personal experience, Pediatr. Endocrinol. Rev. 15 (13) (2016 Mar) 499–506.
[2] M. Ruchala, E. Szczepanek, W. Szaflarski, J. Moczko, A. Czarnywojtek, L. Pietz, M. Nowicki, M. Niedziela, M. Zabel, J. Köhrle, J. Sowinski, Increased risk of thyroid pathology in patients with thyroid hemiagenesis: results of a large cohort case-control study, Eur. J. Endocrinol. 162 (1) (2010 Jan) 153–160.
[3] W. Shabana, F. Delange, M. Freson, et al., Prevalence of thyroid hemiagenesis: ultrasound screening in normal children, Eur. J. Pediatr. 159 (2000) 456–458.
[4] R. Maiorana, A. Carta, G. Floriddia, et al., Thyroid hemiagenesis: prevalence in normal children and effect on thyroid function, J. Clin. Endocrinol. Metab. 88 (2003) 1534–1536.
[5] Y.H. Wu, R.O. Wein, B. Carter, et al., Thyroid hemiagenesis: a case series and review of the literature, Am. J. Otolaryngol. 33 (2012) 299–302.
[6] U. Azmat, J.E. Phay, T. Teknos, J. Bekeny, F. Nabhan, A case of a rare association between right thyroid hemiagenesis, lingual thyroid, and papillary thyroid cancer, US Endocrinol. 13 (1) (2017) 1–2.
[7] A. Aslaner, M. Aydin, A. Ozdere, Multinodular goiter with thyroid hemiagenesis: a case report and review of the literature, Acta Chir. Belg. 105 (2005) 528–530.
[8] P.S. Jha, V. Rote-Kaginalkar, P. Titare, M.B. Jadhav, Hemiagenesis of thyroid with dual thyroid ectopia: a rare case report, Indian J. Radiol. Imaging 28 (1) (2018) 14–17, https://doi.org/10.4103/ijri.IJRI_292_17.
[9] for the SCARE Group, R.A. Agha, T. Franchi, C. Sohrabi, G. Mathew, The SCARE 2020 guideline: updating consensus Surgical CAse REport (SCARE) guidelines, Int. J. Surg. 84 (2020) 226–230.
[10] J.C. Melnick, P.E. Stemkowski, Thyroid hemiagenesis (hockey stick sign): a review of the world literature and a report of four cases, J. Clin. Endocrinol. Metab. 52 (2) (1981) 247–251.
[11] M. Korpal-Szczyrska, W. Kosiak, D. Swieton, Prevalence of thyroid hemiagenesis in an asymptomatic schoolchildren population, Thyroid 1 (18) (2008 Jul) 637–639.
[12] B. Kim, J. Lee, M. Jung, E. Lee, S. Ock, S. Kwon, et al., A case of thyroid hemiagenesis associated with graves' disease and follicular neoplasm, J. Med. Cases 1 (6) (2015 Jan) 385–387.
[13] N. Karabay, A. Comlekci, M.S. Canda, F. Bayraktar, B. Degirmenci, Thyroid hemiagenesis with multinodular goiter: a case report and review of the literature, Endocr. J. 50 (4) (2003 Aug) 409–413.
[14] M.F. Sheridan, A.D. Bruns, L.P. Burgess, Hemiagenesis of the thyroid gland, Otolaryngol. Head Neck Surg. 112 (1995) 621–623.
[15] J.G. Piera, J. Garriga, R. Calabuig, D. Bargallo, Thyroid hemiagenesis, Am. J. Surg. 151 (1986) 419–421.
[16] K.T. Wong, A.T. Ahuja, Ultrasound of thyroid cancer, Cancer Imaging 5 (1) (2005) 157–166, https://doi.org/10.1102/1470-7330.2005.0110. Published 2005 Dec 9.
[17] M. Blum, in: K.R. Feingold, B. Anawalt, A. Boyce (Eds.), Ultrasonography of the Thyroid. [Updated 2020 Apr 11], Endotext [Internet], MDText.com, Inc, South Dartmouth (MA), 2000.
[18] R. Prakash, R.V. Narayanan, S.K. Chakravarty, Differentiation between suppressed thyroid tissue and thyroid hemiagenesis with tc-99 m pertechnetate radio- nuclide angiography, Clin. Nucl. Med. 15 (1990) 605–609.