THYROID HEMIAGENESIS WITH PAPILLARY CARCINOMA: A RARE CASE REPORT

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Abstract

Thyroid hemiagenesis is a rare condition in which there is failure of development of one lobe of thyroid. The condition is diagnosed incidentally either for a pathological coexistence of malignancy or functional condition of the thyroid gland. we report a rare case of thyroid hemiagenesis co-existent with papillary carcinoma of the thyroid.

Key Words: Papillary carcinoma, Thyroid hemiagenesis.

Introduction

Thyroid hemiagenesis is a rare embryological condition in which one lobe fail to develop. It is seen predominantly in females(3:1).[1] The coexistence of thyroid hemiagenesis and thyroid carcinoma is very rare.It is caused by the developmental defect of a thyroid lobe or failure of its precursor to migrate to the normal location and was first described by Hand-field Jones in 1866.[2]

The prevalence of thyroid hemiagenesis is 0.02% to .2%.[3] Pathologic condition associated with the remaining thyroid lobe are Thyroid Adenoma,Graves' disease, Multinodular Goiter, Chronic Thyroiditis.The association of thyroid hemiagenesis with thyroid carcinoma is extremely rare. Among them papillary carcinoma is seen more than any other types. We report a case of thyroid hemiagenesis coexistant with papillary carcinoma of thyroid.

Case Report

A 22 year old female who presented in our department with anterior neck swelling for 3 years.Past history of any neck surgeries or radiation was absent. On examination, the right lobe was palpable measuring approx 3cm x 4cm. The left lobe was not palpable, thyroid function test was normal. On ultrasonography,the right lobe of thyroid was larger in size with multiple heterogenous regular outlined nodule largest one measuring 13.5mm x 10.9mm, the left lobe was not visualized. Fine needle cytology of the right lobe revealed features suspicious of papillary

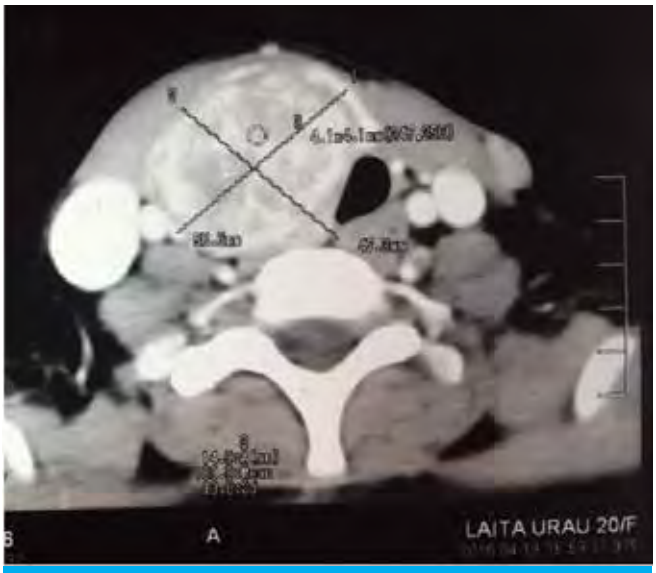

Fig I CT of thyroid with enlarged right lobe and absence left lobe

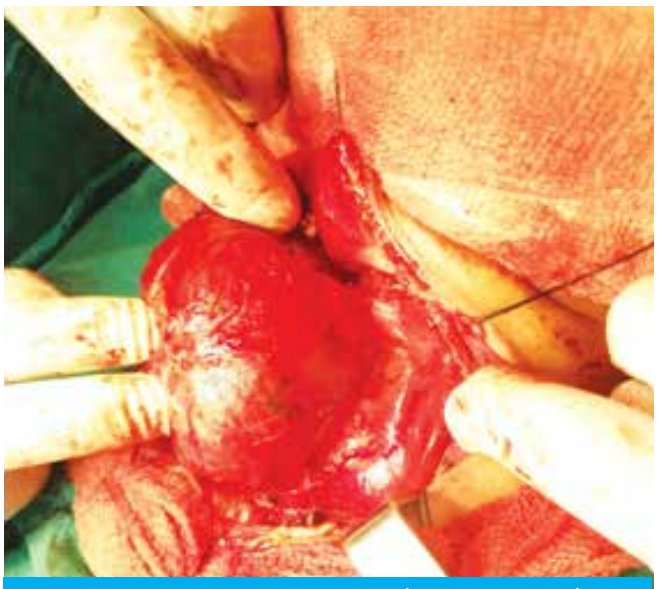

Fig II : Operative picture showing complete absence of left lobe and mass in the right lobe.

carcinoma. CECT neck showed irregularly outlined highly enhancing mass involving the right lobe and isthmus of the thyroid with intrathyroidal extension and few cervical lymphadenopathy at level 1B and level II.The left lobe of the gland was not visualized separately.Total thyroidectomy was performed. The final histopathology was papillary carcinoma of thyroid.

Discussion

Thyroid hemiagenesis is a rare congenital anomaly. Only about 350 cases has been reported in the literature[4]. Hemiagenesis commonly involves the left lobe , followed by isthmus and very rarely the right lobe[5]. Most of them presents with concomitant thyroid pathology like Simple Goiter, Toxic and Nontoxic Nodules, Hashimoto Thyroiditis, Subacute Thyroiditis, Papillary Carcinoma, Follicular Carcinoma and rarely Medullary Carcinoma[1,6].The thyroid gland develops in the floor of the pharynx at the level of foramen caecum. It descends in the neck and attains the final position just in front of the trachea by seventh week. Failure of this descends leads to ectopic thyroid which can present any where from the floor of mouth to the neck[7].The cause of thyroid hemiagnesis is not very clear. It may result from failure of cells to migrate laterally in the neck. It is also unclear that the lobulation process is due to genetic or environmental factors. The hypothesis that a genetic mechanism may underlie thyroid hemiagenesis is based on both identification of familial cases of thyroid hemiagenesis[3-6] and demonstration of thyroid hemiagenesis in first degree relatives of patients affected by thyroid dysgenesis or ectopy. Several genes have been found to control thyroid descent and development.Three thyroid transcription factors, TTF1, TTF2, and Pax-8 are reported in thyroid developmental defects but these genes have not been investigated in thyroid hemiagenesis[8].

The total number of thyroid agenesis case reported are not the true incidence of thyroid hemiagenesis as most of thyroid lobes are normal or discovered incidentally during the investigation of either thyroid diseases or routine ultrasound. Among the common disease in the remaining lobe are multinodular goiter, hyperthyroidism, adenoma, hypothyroidism, adenocarcinoma, chronic thyroiditis, and subacute thyroiditis. Only few cases of thyroid hemiagenesis and thyroid carcinoma have been reported. Out of them papillary carcinoma is the most common. Medullary carcinoma has been reported only once.

The prevalence of thyroid hemiagenesis is between 0.05% to 0.2%. It commonly affects the left lobes in 80% of cases and absence of isthmus is found in 50% cases. It is common in females than males(3:1), hemiagenesis is usually diagnosed by imaging like ultrasonigraphy, CT, MRI, or thyroid scintigraphy. The treatment of disorder like hyperthyroidism is based on the same criteria for normal thyroid gland. The condition that mimic thyroid hemiagenesis are hyperfunctioning nodule with marked suppression of all other thyroid tissue, inflammation, neoplasm or infiltrative diseases like amyloidosis.In those cases, radiological investigations like USG,CT or MRI will confirm the findings of thyroid scans. The cases of thyroid hemiagenesis who are asymptomatic should have monitoring of their thyrotropin levels, if elevated treatment with thyroid hormone and regular follow-up for any development of thyroid neoplasms.

Conclusion

Thyroid hemiagenesis is a rare abnormality of thyroid, they usually are asymptomatic and are detected by routine radiological investigation. Occurrence of papillary thyroid carcinoma is an extremely rare finding.

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