

Case Study

Multipl Thyroid Ectopia with Normally Located Tyroid: Can It Be Hereditary Disorder?

ABSTRACT

Heterotopic thyroid tissue -also called as ectopic thyroid - is a rare developmental anomaly. Above all, multiple ectopic thyroid glands including an intrapulmonary one are extremely rare.

47 years old female has been admitted to our clinic with 12 mm nodule found in the lower lob anterobasal segment of her right lung while she was under investigation for multinodular goiter. We have learned that she had undergone thyroidectomy 31 years ago and multinodular goiter has occurred in the residual gland. Her sister had a history of operations both for ortotopic multinodular goiter and ectopic thyroid more than two foci; and her mother had multinodular goiter but has not been investigated in terms of an ectopic gland. In the operative exploration, the mass has been found out at the inferior part of middle lobe and wedge resection has been performed. Pathologic examination revealed that the nodule was a 1 cm well circumscribed thyroid tissue with a gray-yellow color. It was positive for Thyroid transcription factor -1 and Thyroglobulin in immunohistochemical staining. Interestingly, a sublingual ectopic thyroid tissue was detected by Tc99 scintigraphy at postoperative period.

Such a case sharing a similar medical history with her sibling in terms of thyroid pathology brings to mind that the condition might have a hereditary aspect.

Key words; Ectopic thyroid, Multiple, intraparenchymal, lung, hereditary

24 Introduction

25 Heterotopic thyroid gland - also known as ectopic thyroid- is a developmental anomaly which
26 can be described as the existence of thyroidal tissue beside its expected place. The condition
27 is caused by the abnormalities in migration of the thyroid cells during the intrauterine
28 development phases

29 As it is known, the thyroid tissue migrates from endodermal protrusion of the first and second
30 pharyngeal arch at 3 weeks to 7 weeks of gestation. It descends anteriorly along the midline
31 from the foramen caecum. A thyroglossal duct forms along the descending path of the thyroid
32 and under goes atrophy during the embryonic stages. During the development, abnormal
33 migration of the tissue causes the gland to be misplaced leading to ectopia

34 Case

35 47 years old female has been admitted to our clinic for the investigation of a lung mass. She
36 had a history of subtotal thyroidectomy 31 years ago for multinodular goiter (MNG); and
37 years after, she had experienced the development of MNG from the residual tissue (Figure 1).
38 It was learned that a 12 mm nodule had been detected in lower lob antero basal segment of
39 her right lung during the preoperative examinations of second thyroidectomy (Figure 2). In
40 her family; her sister had a history of operation for ectopic thyroid glands which were
41 detected in the neck and intratracheal region. Also her mother had goiter but was not
42 examined for ectopia. The preoperative thyroid function tests T3;2,60 pg/mL(1,71-3,71),T4;
43 1,13 ng/dL(0,7-1,48), TSH;1,4054 uIU/mL(0,35-4,94); while using levothyroxine. Her
44 respiratory function tests were as FEV1: 2,63 %85,9, Fvc: 3,34 Lt %93,9, FEV1/FVC: 78.80.

45 The operation was performed by thoracotomy. During the intra operative exploration; the mass
46 had been found out at the inferior part of middle lobe and wedge resection was carried out.

47 Pathologic examination revealed that the nodule was a 1 cm well circumscribed thyroid tissue

48 with a gray-yellow color. It was positive for Thyroid transcription factor -1 and thyroglobulin,
49 in immunohistochemical staining. We did not face any postoperative complications and the
50 drain was removed at the second day. A sublingual ectopic thyroid tissue was detected with
51 whole body Tc99 scintigraphy scanning at postoperative period (Figure 3).

52 Discussion

53 In the literature; multiple ectopic thyroid glands is not a commonly reported condition. In
54 English series, 45 cases with dual ectopic gland had been reported. Another study based on
55 the thyroid scintigraphy results announced the incidence of dual ectopy as 0.05 %. The most
56 common ones can be pronounced as lingual (33.73%), sublingual (27.71%) and subhyoid
57 (22.89%) (1). Also, dual ectopic thyroid tissue with a normally located thyroid gland has been
58 reported in only two cases in literature (2,3) and non of these were in the lung.

59 Our case is consonant to dual ectopic thyroid tissue with a normally located thyroid gland, as
60 we have detected a sublingual tissue in addition to lung.

61 Three or more ectopic foci are quite rare. By the reason of the fact that we do not have enough
62 information about the prior subtotal thyroidectomy operations, it is impossible for us to
63 classify the case in such a group; but also, the available data let us declare that it is a multiple
64 one.

65 Ectopic thyroid gland is generally observed throughout the radiological monitoring and CT
66 scans. Radiologic examinations for the diagnosis include CT, magnetic resonance imaging,
67 Tc-99m, I-131, and I-123. It has been reported that ectopic tissue might be seen in the
68 mediastinum, lungs, and heart; manifesting usually with dry cough, dyspnea, and hemoptysis.
69 Less commonly, patients may present with dysphagia or the superior vena cava syndrome.
70 Intra thoracic thyroid may also be revealed incidentally on chest radiography (4–6) or during
71 an autopsy (7). Ectopic intrapulmonary thyroid can mimic pulmonary metastasis such our

72 case (8) Malignant transformation is relatively uncommon, reported in about 15% of patients
73 (9).

74 It is usually asymptomatic; but can cause symptoms due to its location, size, and the presence
75 of malignancy. Indeed, our case had no symptoms; ectopic gland was detected incidentally by
76 computerised tomography (CT) as she was examined before her second thyroidectomy.

77 In addition, it is reported that orthotopic thyroid coexisted in all cases of intra pulmonary
78 thyroid [6,10]. Interestingly; in our case, another ectopic gland was determined at tongue
79 basement by whole body technetium scintigraphy at eighth postoperative month.

80 On the other hand, her sister had a history operations both for ortotopic multinodular goiter
81 and ectopic thyroid. Moreover, we have learned that she had undergone servical trachea
82 resection for intratracheal ectopic thyroid. The family history of more than three foci made us
83 think that the condition might have a familial aspect.

84 Also, a recently identified familial occurrence of congenital hypothyroidism (CH) associated
85 with thyroid dysgenesis suggests the involvement of hereditary factors (11).

86 Evaluation of asymptomatic first-degree relatives of children with CH has revealed a high
87 frequency of different thyroid developmental abnormalities (11).This suggests that the thyroid
88 phenotype may be variable despite a common genetic alteration.

89 A study on an animal model has shown that Sonic hedgehog (Members of the hedge hog
90 family) are soluble ligands for Patched (Ptc) receptors and act as key regulators of
91 embryogenesis. Sonic hedgehog (Shh) is of particular interest because it has been found to be
92 essential to foregut development-deficient mice display distinct features of thyroid dysgenesis
93 and causes hemiagenesis and ectopic development of the thyroid in mouse (12).

Also by the genetic researches on these cases; it has been detected that 19 genes which genes involved in embryonic development and in the Wnt pathway were exclusively associated with thyroid ectopy. Our case might have such a genetic disorder (13).

There is no consensus on the optimal therapeutic strategy for ectopic thyroid tissue because of the rarity of this clinical entity. Surgical treatment is planned according to the age, size, local symptoms, functional thyroid status and complications and can be performed considering the presence of symptoms or malignancy. Depending on the position of the tissue; sternotomy or thoracotomy can be performed for surgical treatment. Complete surgical resection may be preferred if there is a suspicion of malignant transformation. Otherwise, the follow up is suggested in terms of mass enlargement or complications in asymptomatic cases with euthyroid status (14).

Also, thoracoscopic treatment might be considered (15). As in our case, surgery can be performed in order to determine the presence of any malignancy; and/or to diagnose and treat the ectopic thyroid.

Considering the fact that the existence of multiple ectopic thyroid gland in both siblings; we may suggest that a genetic predisposition might have a role in the development of this condition. It can be beneficial to scan the whole body in all the cases and scrutinize their relatives.

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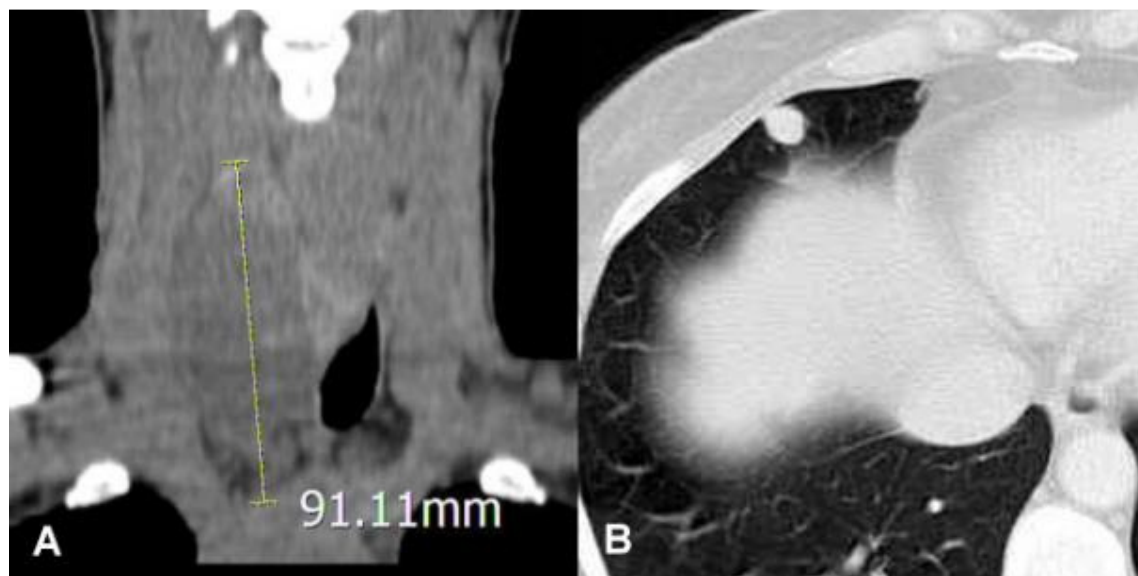
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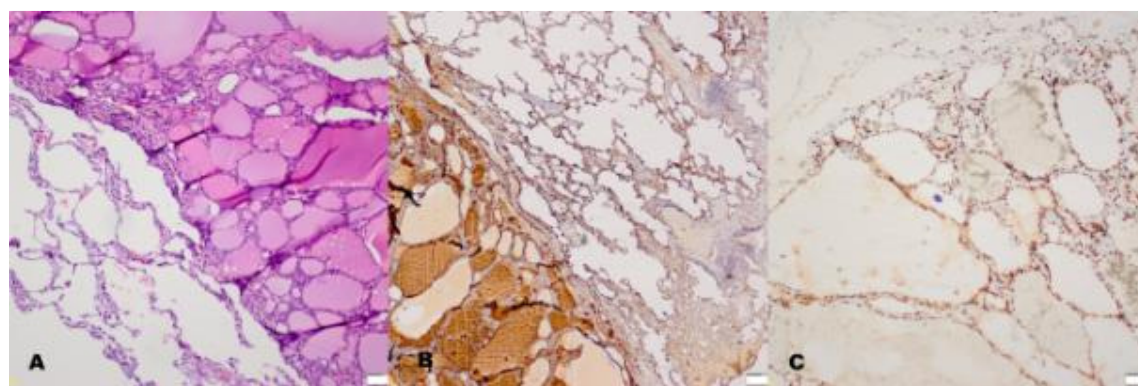
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- 150 Figure 1; A; Pulmonary nodule seen on thorax computed tomography B; Giant multinodular
151 goiter caused tracheal deviation on neck computed tomography
- 152 Figure 2; A; Lesion consisting of well-circumscribed thyroid follicles in the pulmonary
153 parenchyma (HE, x100) B; Thyroglobulin positivity of the lesion (Thyroglobulin, X100), C;
154 TTF-1 positivity of the lesion (TTF-1, x100)
- 155 Figure 3; A sublingual ectopic thyroid tissue detected by Tc99 scintigraphy
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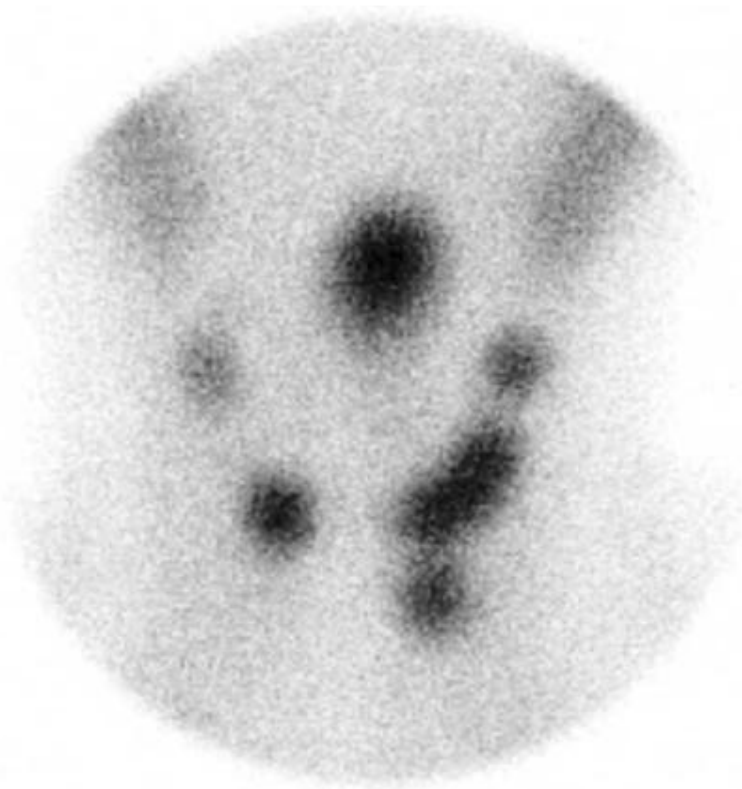
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158 figure 1



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160 figure 2



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162 figure 3