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PICTORIAL ESSAY

Ectopic Thyroid on Scintigraphy

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ABSTRACT

Ectopic thyroid is a rare condition that involves aberrant embryogenesis of the thyroid primordium during its descent from the foramen caecum to its final pretracheal position. Ectopic thyroid tissue can be found around its route of migration, in the mediastinum and subdiaphragmatic areas. Histologically, ectopic thyroid resembles normal thyroid parenchyma. Preoperative recognition of the presence of ectopia is crucial for the surgeon when completing a thyroidectomy. Ectopic thyroid tissue left behind during total thyroidectomy may jeopardise treatment outcome and cause monitoring conflict. This article shares our nuclear findings that will aid awareness of ectopic thyroid tissue.

中文摘要

異位甲狀腺核醫造影圖文回顧

朱任公

異位甲狀腺是臨床罕見的形式，在胚胎發育過程中，甲狀腺原基從盲孔下降到氣管前其最終位置，因下降誤導致組織異位；在遷徙路線周圍、縱膈腔及橫膈下區皆會出現異位甲狀腺。異位組織類似於正常的甲狀腺實質，而且通常沒有症狀，卻會影響甲狀腺切除術的成敗，外科醫生必須在手術前察覺異位症的存在；在全甲狀腺切除後遺留的甲狀腺異位組織，會危及治療效果，並導致術後監測困難。本文旨在分享異位甲狀腺的核醫造影表現，幫助臨床認知異位甲狀腺組織。

INTRODUCTION

During embryological development, the thyroid primordium descends caudally (while in contact with the aortic primordium) from the foramen caecum to the pretracheal location of the neck. A tubular tract (the thyroglossal duct) forms as a connection to the origin during caudal migration of the thyroid bud, and involutes between the 8th and 10th weeks of gestation. Aberration in this embryological descent results in ectopic thyroid tissue (Figure 1). Most thyroid ectopias are found along the thyroid descent pathway of the anterior neck (Wölfler area).[1] It is hypothesised that lateral aberrant thyroids found in the submandibular and lateral neck regions originate from a defective lateral thyroid anlage that cannot migrate and fuse with the median thyroid anlage.[2]

The exact incidence of ectopic thyroid remains unknown since many remain asymptomatic until later in life, and some are never diagnosed. Of 1,007,350

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newborns screened in Saudi Arabia, 62 infants were identified to have ectopic thyroid, showing an incidence of 1:16,248.[3] A pilot screening for congenital hypothyroidism in Taiwan (from January 1984 to June 1987) found a higher incidence among neonates.[4] The incidence of ectopic thyroid is about 1: 11,258 (8/90,062) newborns.[4] The substantial variation in the prevalence of ectopic thyroid among different cohorts is probably attributable to the comprehensive evaluation, the highly selected populations, and geographic variation.

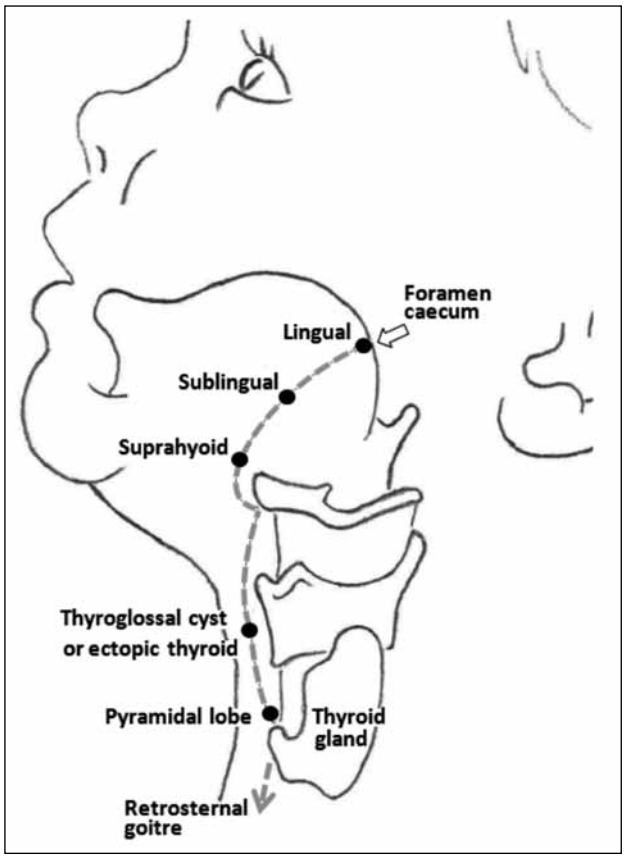

Figure 1. Descent of the thyroid primordium along the thyroglossal duct. Schematic drawing showing common sites of ectopic thyroid tissue or thyroglossal duct remnants.

The radionuclides that are currently used for thyroid imaging are isotopes of iodine (I-131 and I-123) and of technetium (Tc-99m). Tc-99m pertechnetate is trapped by the thyroid but not organified and is released over time as unaltered pertechnetate ion. Its short physical half-life of 6 hours and principal gamma photon of 140 keV are ideal for imaging using scintillation cameras. On a Tc-99m pertechnetate scan, the salivary glands are usually seen in addition to the thyroid (Figure 2a). I-131 and I-123 are chemically analogous to stable iodine that is preferentially handled by the thyroid (Figure 2b). I-131 has a half-life of 8 days and both beta and gamma radiations. I-123 is a good substitute for I-131 because it has a shorter half-life (13 hours), with a 159 keV gamma ray that is suitable for imaging. The radiation burden to the thyroid is far less (1%) than that of I-131. Nonetheless its use has been restricted by its high cost and complex production in a cyclotron. The resolution of detailed structures of the organ with scintiscan may not be as high as with other imaging techniques, such as computed tomography (CT) or magnetic resonance imaging. Nonetheless thyroid scans can provide information about the shape and the overall activity of the gland, and can light up ectopic thyroid foci, and is often unobtainable with other imaging techniques.

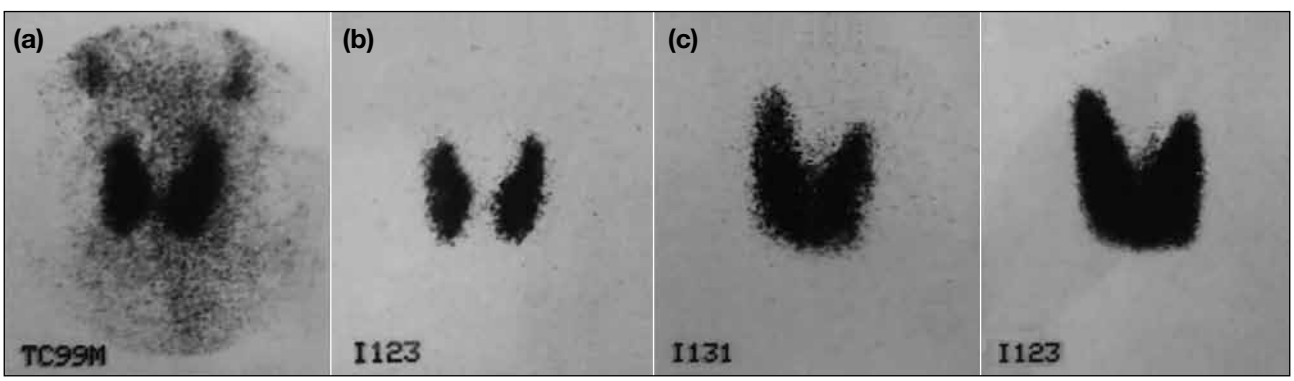

Figure 2. Normal thyroid scintiscans showing homogeneous distribution of radioactivity in both lobes. (a) On technetium-99m pertechnetate scan, the salivary glands are usually seen in addition to the thyroid. (b) Iodine-123 imaging in the same patient displays lower background and greater clarity of the picture. (c) Performance comparison of iodine-131 and iodine-123 in Graves' hyperthyroidism: there is diffuse enlargement of the gland with increased radionuclide uptake. Images obtained with both isotopes yield essentially the same information.

CLINICAL PRESENTATIONS

Lingual Thyroid

Lingual thyroid is the most common type accounting for 90% of cases, while sublingual (suprahyoid, hyoid, and infrahyoid) types are less frequently encountered. Owing to close anatomical proximity, lingual thyroid is generally the most common term used for the ectopic thyroid of the tongue base, tongue, and suprahyoid area. The lingual thyroid appears as a rounded swelling at the back of the tongue (the foramen caecum), and is commonly detected during puberty and pregnancy when increased levels of thyrotropin stimulate enlargement of the ectopic thyroid tissue. If it is big it may interfere with swallowing, speaking, and breathing. Ulceration and haemorrhage may be caused by trauma. It is best treated by full replacement with thyroxine when it should get smaller. Excision is sometimes necessary. Ectopic thyroid tissue may be the only functioning thyroid tissue in 70% to 90% of cases, particularly with lingual and cervical heterotopias.[5] Dual thyroid ectopy (Figure 3) is a rare event estimated to occur in 9% of patients with ectopic thyroid tissue. Only 43 cases were reported following a search (PubMed) of the English literature.[6] In most cases, the first lesion is lingual or sublingual and the second is subhyoid, infrahyoid, or suprahyoid.[7]

Thyroid Hemiagenesis

Thyroid hemiagenesis is caused either by abnormal descent of the thyroid rudiment or by disturbance of the lobulation process. To date, approximately 300 cases have been reported.[8] It is even rarer to have one ectopia with a hemiagenetic thyroid present simultaneously (Figure 4). For unknown reasons, the missing lobe

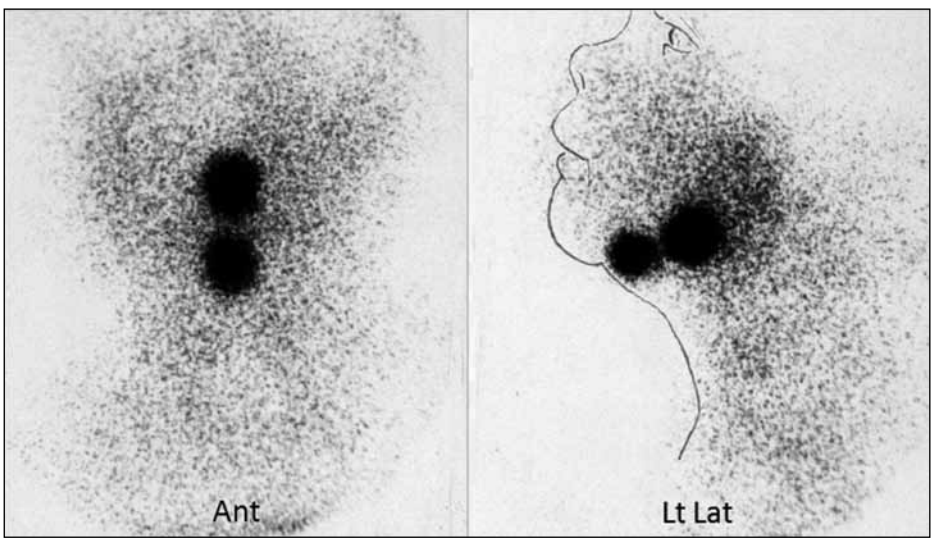

Figure 3. Dual ectopic lingual and sublingual thyroid. Technetium-99m pinhole thyroid image showing non-visualisation of the thyroid gland, and two avid foci in the midline, superior to the thyroid gland lodge. On the lateral view, the exact localisation of the foci is defined.

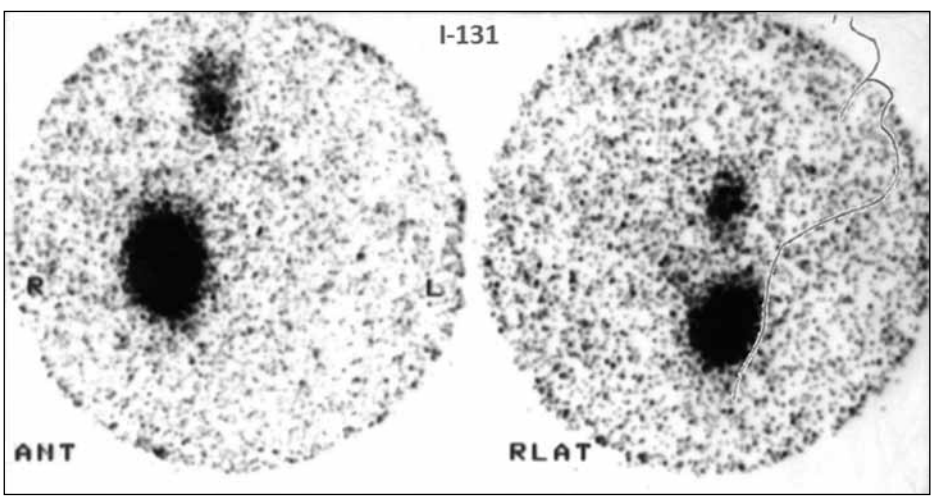

Figure 4. Hemiagenesis with lingual ectopia. Iodine-131 thyroid scan demonstrates absence of the left lobe and the isthmus of the thyroid with an ectopic focus at the tongue base. There might be a common pathogenic mechanism in these two types of thyroid dysgenesis.

is usually the left one (88%).[9] A recent large cohort case study revealed that patients with thyroid hemiagenesis were more likely to develop pathology like hyperthyroidism, hypothyroidism, multinodular goitre, chronic thyroiditis, adenocarcinoma, and papillary thyroid carcinoma.[9] The observed high incidence of morphological abnormalities in the existing lobe is presumably due to sustained thyroid-stimulating hormone overstimulation. The existing lobe is usually enlarged and capable of covering hormone requirements. During the growth or pregnancy, as well as environmental factors, increased demand for thyroid hormone may worsen a subclinical thyroid disorder.[9]

Thyroglossal Tract Remnant

The thyroglossal tract is an epithelium-lined tube that connects the thyroid anlage and the foramen caecum, representing the track of embryological descent of the thyroid primordium.[10] Once the primordium reaches its pretracheal position in the neck the tract involutes (Figure 1). Nevertheless a portion of the tract may occasionally remain patent and give rise to cystic enlargement, referred to as thyroglossal cyst (Figure 5). The distal portion of the tract differentiates into the pyramidal lobe of the thyroid (Figure 6), and the rest of the duct disappears. Approximately 7% of the population have thyroglossal tract remnants, representing the most common congenital cervical abnormality.[11] The remaining thyroid tissue may be left along the course of the duct, as it is estimated that 35% to 70% of thyroglossal cysts contain thyroid tissue.[12] Thus, the remnant may present as a cyst, a duct, a fistula, or a tumour.[13] Benign thyroglossal duct cysts usually present as nontender and generally movable masses

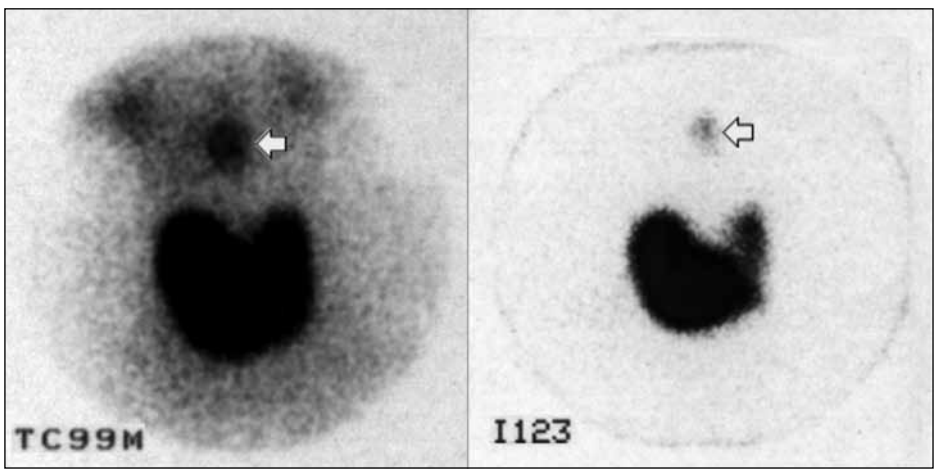

Figure 5. Thyroglossal cyst with aberrant thyroid tissue. Technetium-99m thyroid scan displays a hyperactive nodule in the right lobe as well as focal uptake (arrow) in the submental region, concordant with the palpable lesion. Activity is seen in the salivary glands on technetium-99m imaging. To obviate misinterpretation, a repeat iodine-123 scan confirmed the aberration of thyroid tissue (arrow).

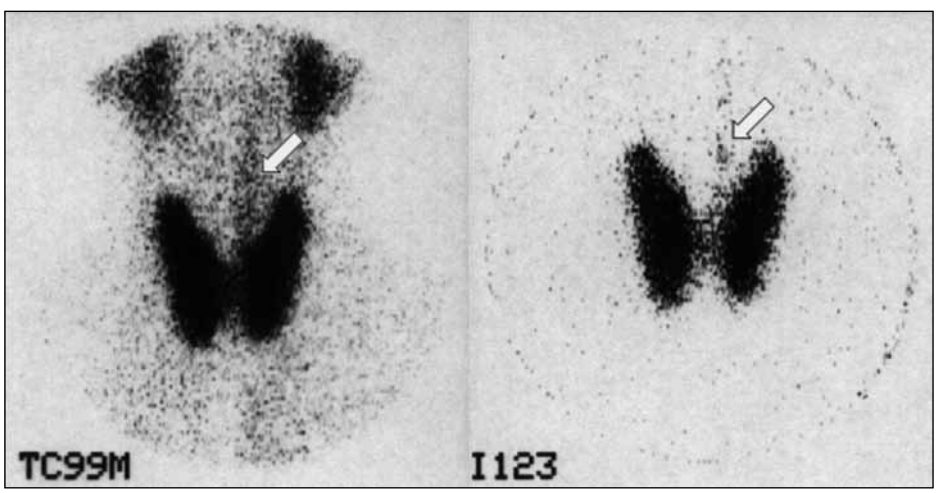

Figure 6. Pyramidal lobe. Technetium-99m versus iodine-123 thyroid imaging. A small linear streak of tracer activity is seen to extend superiorly (arrow). Radioactive saliva accumulated in the oesophagus may imitate a pyramidal lobe on technetium-99m imaging. Iodine-123 scan confirmed the presence of pyramidal lobe (arrow).

in the midline of the anterior neck. Thyroglossal duct carcinoma occurs in approximately 1% to 2% of thyroglossal cyst cases. Malignant transformation should be suspected in any thyroglossal cyst that is irregular or has undergone recent change.[13]

The pyramidal lobe (Figure 6) arising from the upper border of the isthmus is a vestigial remnant of the fetal thyroid stalk (thyroglossal tract). It has been reported to present in 41% (135/327) of patients undergoing neck CT[14] and 12% (74/604) of cases undergoing total thyroidectomy.[15] In a retrospective study of tc-99m pertechnetate images of 866 patients, the rate of pyramidal lobe visualisation was 13% in nodular goitre patients, 43% in diffuse goitre patients, and 20% in patients whose scintigraphy showed normal thyroid glands.[16] The pyramidal lobe is considered a normal variant or a component of the thyroid,[15] but of great importance to the thyroid surgeon during thyroidectomy. It can be a cause of incomplete thyroidectomy for various thyroid diseases.[15]

Intrathoracic Struma

Intrathoracic (retrosternal, mediastinal) struma refers to goitre with more than 50% of its mass located below the thoracic inlet, mostly in the anterior mediastinum. According to the origin of thyroid tissue, intrathoracic goitre is classified as primary or secondary. The primary type occurs as an aberrant descent of the thyroid anlage along with the heart's descent, accounting for only 0.2% to 1% of all intrathoracic goitres.[5] More than 90% of intrathoracic goitres are secondary types, and refer to retrosternal extension of goitrous tissue from enlarged thyroids. It is postulated that negative intrathoracic pressure, swallowing, and gravity facilitate the descent and eventual entrapment of an enlarged thyroid in the mediastinum.[17] Most (85-95%) intrathoracic thyroid masses represent a benign goitre[17] and are composed of nontoxic multinodular glands. Due to repeated episodes of hyperplasia and degeneration, the goitrous thyroid is present with cystic areas, haemorrhage, fibrosis, and fibrous deposits. Nuclear imaging may demonstrate functioning thyroid in the mediastinum, whereas lesions with limited functional activity, as shown in intrathoracic goitres, do not show significant uptake on scan (Figure 7). CT can reveal the nature and extent of the lesion and specifically identify the anatomic continuity with the cervical thyroid.

Struma Ovarii

Struma ovarii is a rare form of ovary teratoma composed of thyroid tissue in more than 50% of the overall mass.[18] Fewer than 500 cases have been reported.[19] The thyroid component is derived from an ovarian germ cell layer (endoderm),[20] but not an ectopic thyroid on the basis of embryology. Extrathyroidal strumas are not limited to the ovary. Cases of struma salpingii,[21] struma uteri,[22] and struma testis[23] have been reported. Histologically, struma ovarii resemble normal thyroid parenchyma and often accumulate radioiodine (Figure 8). A positive

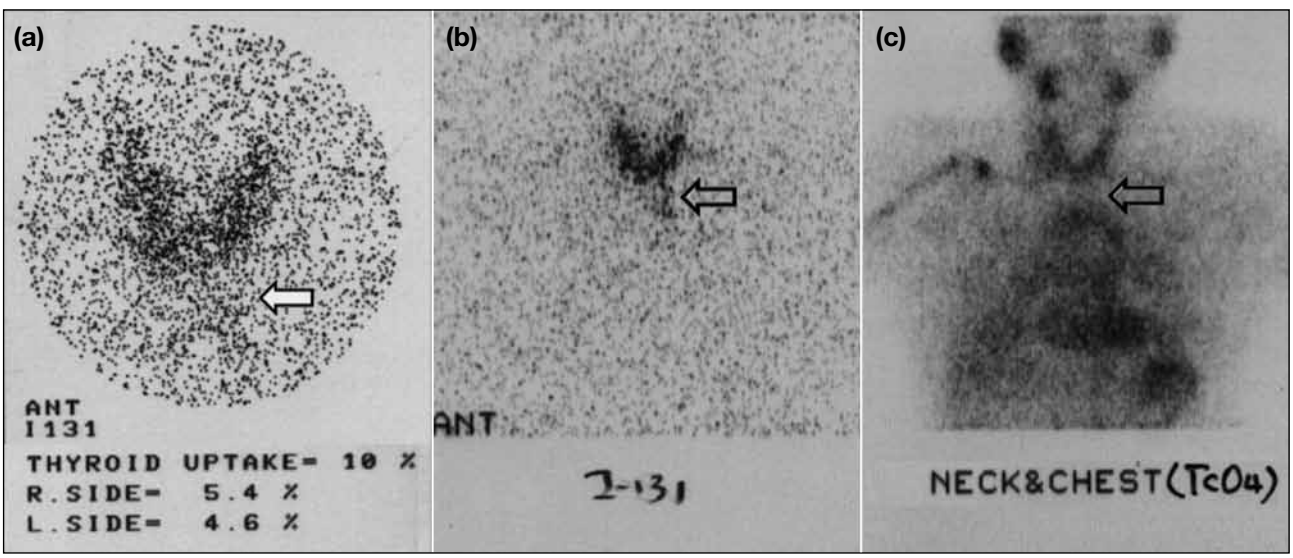

Figure 7. Substernal goitre. (a) Iodine-131 thyroid uptake was 10% (normal range: 15-40%) at 24 hours. The heterogeneous uptake of radioactivity throughout the thyroid and some faint activity extending inferiorly from the left lobe (arrow) are noted. The goitrous lesion (arrows) is more appreciable on (b) iodine-131 chest image (middle) compared with (c) technetium-99m pertechnetate image (right).

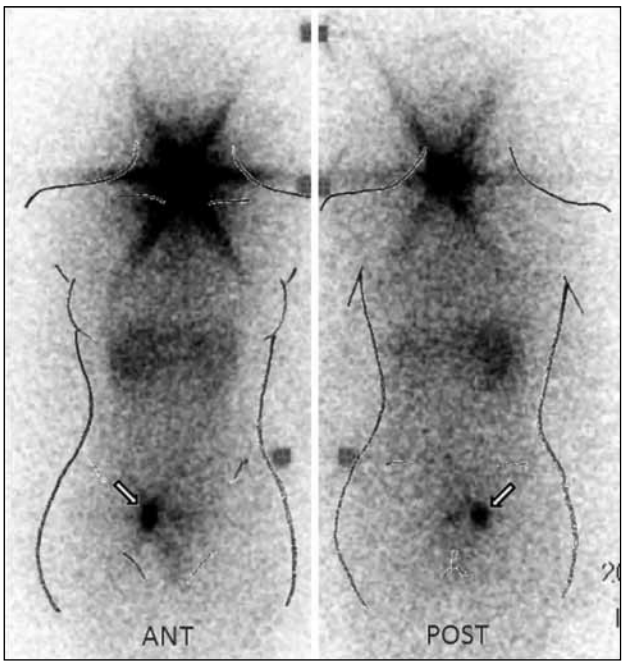

Figure 8. Struma ovarii. A woman with thyroidectomised papillary thyroid cancer 7 days after administration of a therapeutic dose of iodine-131 for remnant ablation. Aside from avid uptake in the thyroid bed, there is an intense lesion (arrows) in the right pelvis. The pelvic lesion was subsequently proven to be a struma ovarii.

finding on radionuclide imaging allows a definite diagnosis.[22]

Others

Other locations outside the embryological migration path of the thyroid include the gallbladder, pancreas, porta hepatis, small bowel, and mesentery. The presence of thyroid follicles in these subdiaphragmatic organs can be hypothetically explained as a metaplastic or heteroplastic phenomenon, since they all share a common embryological origin from the foregut endoderm.[24]

MALIGNANCY POTENTIAL

The ectopic thyroid exhibits the same histological, physiological, and pharmacological features as cervical thyroid.[25] Any disease that affects the thyroid gland may also involve the aberrant thyroid-like adenoma, hyperplasia, inflammation, and malignancy.[5] The rate of malignant transformation in ectopic thyroid is no greater than that in the orthotopic thyroid, occurring in less than 1% of cases.[26] The incidence of carcinoma arising in the thyroglossal duct cyst is approximately 1% to 2% of cases.[27] As mentioned above, the thyroidal component of struma ovarii is derived from primitive germ cells of the ovary,[20] and malignant transformation rates of teratomatous thyroid tissue range from 5% to 37%.[28] Struma ovarii that contain thyroid-type carcinoma must be distinguished from rare cases of papillary or follicular thyroid carcinoma metastatic to the ovary.

Recent evidence from retrospective cohort analyses suggests that substernal goitres have a much higher cancer rate, ranging between 6.8%[29] and 17%.[30] The high cancer prevalence of intrathoracic goitre in these studies can be attributed to the age-specific incidence rate, concordant with the peak-age incidence of thyroid cancer in the 40s or 50s in women and 60s or 70s in men. Most patients with substernal goitre are reported to be operated at a later age and to have had goitre for a long time.[29] Thyroid cancer is incredibly common in the elderly people: up to 6% of autopsy series in the United States and >20% in Japan also harbour microscopic foci of thyroid carcinoma.[31] All patients clinically suspected to have substernal goitre should undergo neck and chest CT scan, both to confirm the presence of the disease and devise a treatment plan.

Ultrasound-guided fine-needle aspiration biopsy may, with high sensitivity, enhance preoperative diagnosis. Magnetic resonance imaging is particularly useful in lingual thyroid when there is difficulty in differentiating thyroid tissue from tongue muscle.[32] Nuclear imaging provides a visual display of functioning thyroid on the basis of high uptake of the radiotracer in thyroid tissue. The absence of tracer uptake in the normal position usually means thyroid aberration or agenesis. The state of activity in the orthotopic thyroid and ectopic thyroid tissue is an important clue to diagnosis and management.

Although asymptomatic and euthyroid patients do not require any treatment, they should be followed up and evaluated for mass enlargement and any complications.[5,25] Surgery seems to be the most appropriate treatment for those who show clinical signs of upper airway obstruction or when the lesion shows signs of infection or complications.[25] Since the ectopic thyroid may be affected by diseases that affect the thyroid gland, complete thyroid removal (total thyroidectomy) is usually recommended if surgical resection is considered. Embryological remnants and ectopic tissue left behind in total thyroidectomy may jeopardise completion of the surgery and also cause monitoring conflict.[15] With

respect to struma ovarii, a surgical approach is indicated to prevent malignant alteration of this monodermic and highly specific teratoma.

CONCLUSION

The ectopic thyroid exhibits the same histological features as cervical thyroid. Isotopic scanning is capable of lighting up ectopic tissue in nearly all cases. Preoperative recognition of the presence of ectopic thyroid is crucial for a complete thyroidectomy.[15]

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