

CT and MRI findings of developmental abnormalities and ectopia varieties of the thyroid gland

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ABSTRACT

Ectopic thyroid tissue may be observed anywhere from the tongue base to the lower neck. It is rarely seen in the mediastinum and abdominal cavity. Computed tomography and magnetic resonance imaging are very sensitive for detection and localization of ectopic thyroid tissue. This pictorial essay presents the radiological characteristics of developmental abnormalities and ectopia varieties of the thyroid gland.

Key words: • thyroid gland • thyroid dysgenesis • magnetic resonance imaging • diagnostic imaging

Ectopia of the thyroid gland may not be detected until adulthood, and then is often detected as incidental findings in asymptomatic patients. However, as an increasing number of patients are undergoing magnetic resonance imaging (MRI), ultrasonography (US), and computed tomography (CT) examinations, these anomalies are being seen more frequently. Congenital anomalies and normal variants of the thyroid gland include ectopic and/or accessory thyroid tissue, variation in thyroid extension, and agenesis or hemigenesis of the thyroid gland. Here, the features of these entities on MRI, CT, and US images are presented along with clues for differentiating them from acquired diseases in adults.

Several options exist for imaging the thyroid gland: US, MRI, CT, and nuclear medicine scintigraphy. US is a readily accessible, inexpensive, and radiation-free imaging method commonly used to evaluate the thyroid gland. High-frequency (10–12 MHz) US is the least invasive of the imaging modalities. Some US features may help distinguish between benign and malignant nodules. CT provides cross-sectional information about the architecture of the thyroid gland and dissemination of malignancy. This technique can be used to scan the entire neck, from the nasopharynx to the mediastinum. The main disadvantage of CT is that it requires the administration of an iodinated contrast agent to differentiate the lymph node. In addition, imaging with iodine-based nuclear medicine agents cannot be performed until six weeks after CT, because of the uptake of the iodinated contrast agent by the thyroid gland. Conventional neck MRI is not commonly used in daily practice because susceptibility artifacts can occur on fat suppression sequences in the anterior aspect of the neck. However, the use of eight-channel surface coils for high-resolution MRI can allow adequate evaluation of the thyroid gland and entire neck. Scintigraphic imaging using iodine-131 (I-131) and/or technetium-99 (Tc-99) provides highly sensitive images in patients with ectopic thyroid tissue (1), and scintigraphic imaging modalities offer the possibility of whole-body evaluations. However, no practical value of thyroid scintigraphy exists in the management of euthyroid patients with diffuse or multi-nodular goiter or patients with hyperthyroidism (2).

Thyroid gland embryology and mechanism of ectopia

During normal embryonic development, the thyroid gland develops from endodermal cells that originate from the third branchial pouch in the floor of the primitive pharynx, at the base of the tongue (foramen cecum). This is known as the main anlage, and the thyroid develops from this point. During its maturation, the gland migrates downwards from the base of the tongue at the thyroid diverticulum, leaving the posterior aspect, the thyroglossal duct, passing anteriorly to the hyoid bone

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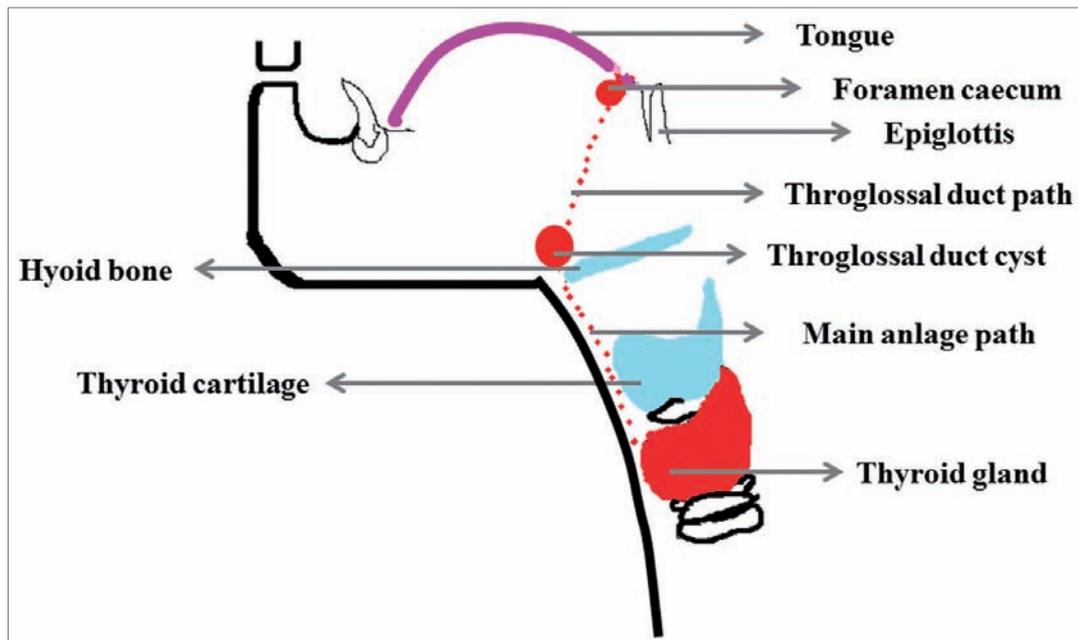


Figure 1. The developmental process of the thyroid gland during the embryonic period. Descent of the thyroid gland during embryological development can be visualized from the thyroglossal duct cyst or remnant/ectopic tissue to its ultimate position, in the pretracheal region.

and thyroid cartilage, which undergoes atrophy prior to definitive thyroid formation (Fig. 1). The two lobes of the thyroid gland normally lie on either side of the trachea, anterior to the second and third tracheal rings, and are joined by an isthmus. Lateral anlagen derived from two fourth branchial pouches give rise to two superior parathyroid glands and the lateral thyroid. The ultimobranchial bodies, derived from the fifth branchial pouches, migrate downward on each side of the neck to produce the parafollicular C-cells (3, 4).

Rarely, during development, all or part of the thyroid precursor cells fail to descend along the normal pathway, and the final gland lies in an ectopic location. During the fetal period, maternal antibodies against thyroid antigens are responsible for the etiology of the arrested descent of the thyroid anlage from its original location at the dorsum of the tongue down to its normal location (5). An ectopic thyroid is a rarely detected anomaly, occurring in approximately 1 in 3000 to 1 in 10 000 people (6). In 90% of cases, the ectopic thyroid is found at the base of the tongue; this is known as a lingual thyroid. However, it may be found in any location along the pathway of migration, from the foramen caecum to the mediastinum. Other non-cervical and non-mediastinal locations of ectopic thyroid tissue are the parotid gland, gallbladder, porta hepatis, proximal

segment of the small bowel mucosa and mesentery, pancreas, adrenal glands, uterus, vaginal wall, pituitary gland, and iris (7–10). In the neck, common locations of ectopic thyroid tissue are the tongue, submandibular region, parotid space, lateral neck region, and midline (11–14).

Goitrogenic enlargement as well as adenomatous and malignant transformation may occur in ectopic tissues, as in the thyroid gland. According to the English-language literature, follicular carcinoma is the most commonly reported tumor of ectopic tissues. In 30%–70% of reported cases, the ectopic tissues were functionally hypothyroid; only a few cases were hyperthyroid (6).

In patients who present with thyroid ectopia, the presence of a normally located thyroid should also be confirmed by physical examination and radiological investigation before surgery. In cases lacking a normal thyroid gland, surgical excision of ectopic tissue(s) can result in acute thyroid insufficiency, a potentially catastrophic condition.

A goitrogenic thyroid usually grows down into the anterior-superior mediastinum. Although extension cranially into the retropharyngeal region is extremely rare (Figs. 2 and 3), any retropharyngeal involvement is usually attributable to direct extension. In an occasional case, it is caused by ectopic thyroid tissue in this region. Cross-sectional imaging techniques can help make differential diagnoses.

Lingual thyroid

A lingual thyroid has been reported to be a nodular soft tissue mass on the dorsal aspect of the tongue, at the foramen caecum. A lingual location is found in 90% of all ectopic thyroid tissues, and it was first described by Hickman in 1869 (15). The incidence of this condition has been reported to be 1 in 100 000. However, lingual thyroid tissue, varying in size from a microscopic focus to a few centimeters, has been found in more than 10% of autopsy cases (16). In about 70% of these cases, a normal thyroid gland was absent from the pretracheal region (17). Lingual thyroid is more common in women, with reported female-to-male ratios ranging from 3:1 to 7:1 (6). The most widely accepted theory of its etiopathogenesis is the non-descent of the midline anlage (3). Lingual thyroids may be discovered incidentally in asymptomatic patients. In lingual thyroid cases, symptoms, such as dysphonia, dysphagia, a lump-in-throat sensation, dyspnea, and chronic cough, are related to the anatomical location and amount of ectopic tissue. US may show the absence of a thyroid gland in normal pre-tracheal locations, but it is less specific than thyroid scintigraphy for differentiation of ectopic tissue from other pathologies. US usually reveals a round, soft tissue mass near the tongue base at the level of the hyoid bone. Thyroid scintigraphy can reveal iodine uptake in the tongue base. CT and MRI reveal normal

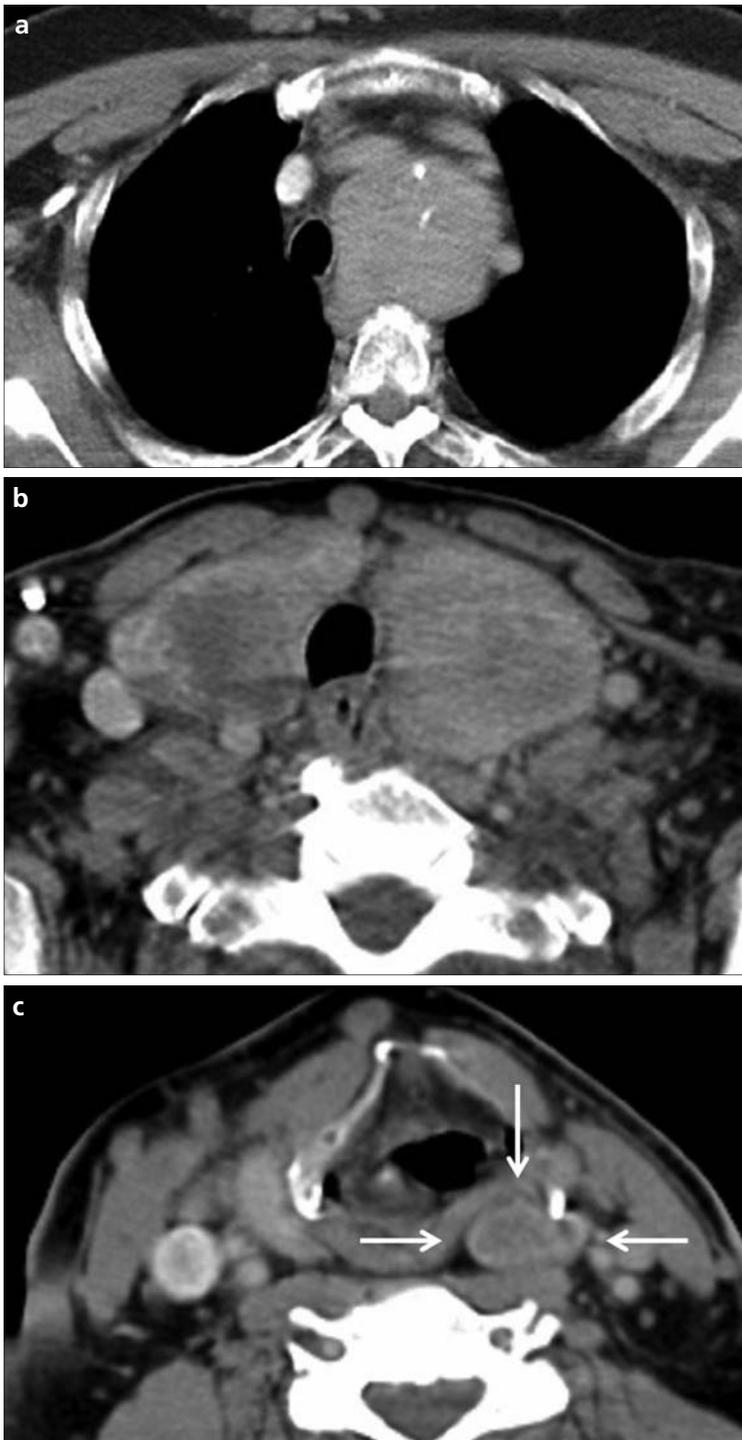


Figure 2. a–c. A 62-year-old woman with goitrogenic enlargement of the thyroid gland. Contrast-enhanced CT images (a–c) show goitrogenic enlargement, parenchymal heterogeneity, and calcification of the thyroid gland. The thyroid gland extended to the mediastinum (a) with unilateral retropharyngeal elongation at the level of the left piriform sinus (c, arrows).

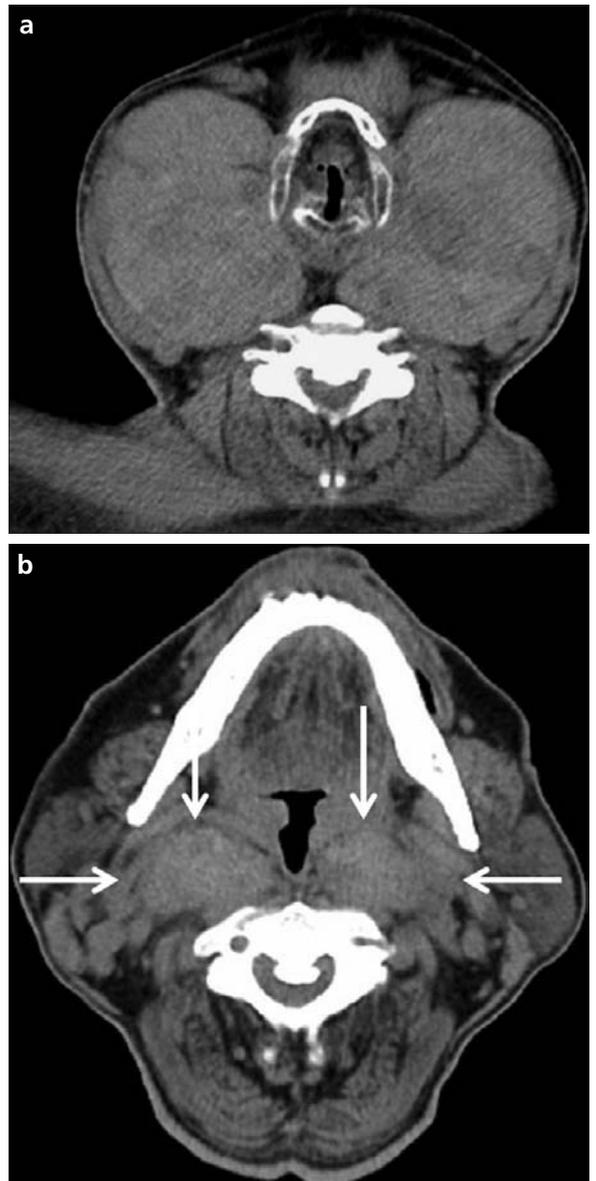


Figure 3. a, b. A 51-year-old woman with goitrogenic enlargement of the thyroid gland and progressive dyspnea, wheezing, and stridor. Unenhanced CT images (a, b) show prominent goitrogenic enlargement and parenchymal heterogeneity of the thyroid gland and mild stenosis of the larynx (a). Bilateral retropharyngeal extension of the thyroid gland was caused by narrowing of the oropharynx at the level of the mandible (b, arrows).

thyroid tissue with or without nodule formation at the tongue base (Fig. 4). Suppression therapy is used to decrease the dimensions of ectopic tissue, but surgical resection is recommended for symptomatic cases.

Thyroglossal duct cyst

Thyroglossal duct cyst is the most common congenital mass of the mid-line neck. Most cases are diagnosed in childhood; nearly 40% of diagnoses are



Figure 4. a–d. A 72-year-old man with a recently diagnosed lingual thyroid. Unenhanced CT image (a) shows a round, well-defined, and heterogeneously dense soft tissue mass at the tongue base (arrows). T2-weighted MR image (b) shows slightly increased signal intensity in the lesion (arrows). Contrast-enhanced T1-weighted MR image (c) shows strong heterogeneous enhancement of the mass (arrows). The airway passage is nearly obstructed by the lingual thyroid at the oropharynx in all images. A transverse US (d) reveals a smooth hypoechoic tumor with cystic areas.

made in young adulthood, and usually involve an enlarged painless mass. In some patients, this anomaly can be complicated by an infection or a neoplasm arising from thyroid rests or the epithelium of the cyst. Thyroglossal duct rests can be located anywhere along the course of the duct, from the base of the tongue to the suprasternal region. Most cysts are located adjacent to the hyoid bone. Cysts located adjacent to the tongue base are lined with stratified squamous epithelium, while cysts located adjacent to the thyroid gland are lined with cells similar to those of the thyroidal acinar epithelium. More than half of all thyroglossal duct cyst walls contain ectopic rests of thyroid tissue (17).

On contrast-enhanced CT images, a thyroglossal duct cyst appears as a fluid-like mass with uniformly thin

peripheral rim enhancement (Fig. 5) (18). With MRI, a thyroglossal duct cyst presents as a hypointense lesion on T1-weighted images and a hyperintense lesion on T2-weighted images; ectopic thyroid tissue also shows marked gadolinium enhancement (Fig. 6). Septations are rarely observed (19). On US examination, a thyroglossal duct cyst usually appears as a cyst-like anechoic mass with a thin outer wall in this characteristic location. When thyroid remnant tissue is present in the thyroglossal duct cyst, iodine uptake can be detected in scintigraphy. This condition must be differentiated from necrotic lymph nodes, external laryngocele, abscess, branchial cleft cyst, cystic hygromas, dermoid cysts, and lymphangiomas (18). A final diagnosis can be made based on histopathological examination.

Midline thyroid ectopia

A midline ectopic thyroid is a congenital anomaly in which a remnant of thyroid tissue lies in the anterior aspect of the neck from the tongue to the diaphragm. Extra-lingual thyroid tissue is usually located in the anterior cervical region. Anterior midline ectopia is most commonly derived from thyroglossal duct remnants. During development, the main anlage of the thyroid gland is adjacent to the heart. The effect of the descending heart is influenced by the thyroid gland descent, which is caused by various anomalies of the thyroid position (20). In addition, incomplete descent of the main anlage may lead to the presence of a pyramidal lobe or levator glandulae thyroideae without ectopia (21).

Ectopic thyroid tissue is located primarily at the anterior aperture of the

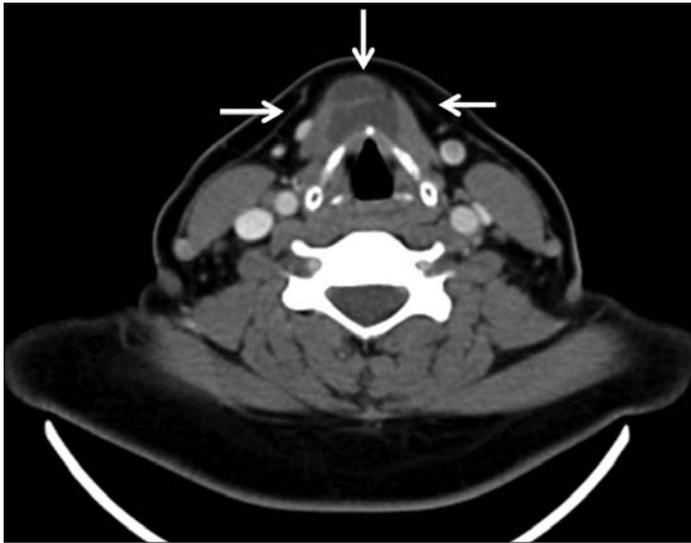


Figure 5. A 34-year-old man with a thyroglossal duct cyst. Contrast-enhanced CT image shows a cystic mass in the anterior midline of the neck, at the level of the thyroid cartilage (arrows). The cyst contains thin septations. Histopathological examination revealed a thyroglossal duct cyst.

thyroid cartilage, in our experience. In our opinion, this location is more common than is appreciated in the general population. The radiological architecture of this tissue is the same as that of the normal thyroid gland, but ectopic thyroid tissue is usually smaller in size, with an ovoid configuration. Ectopic thyroid tissue is subject to the same goitrogenic and neoplastic disorders as normal thyroid tissue, and goitrogenic or neoplastic involvement alters the radiological appearance of ectopic tissue.

In most examples, ectopic thyroid tissue is reported as an incidental finding on US, CT, or MRI (Figs. 7–9). On CT, ectopic tissue is detected as variously sized homogeneous soft tissue at characteristic locations. On MRI, ectopic tissues show the same signal intensity as the thyroid gland.

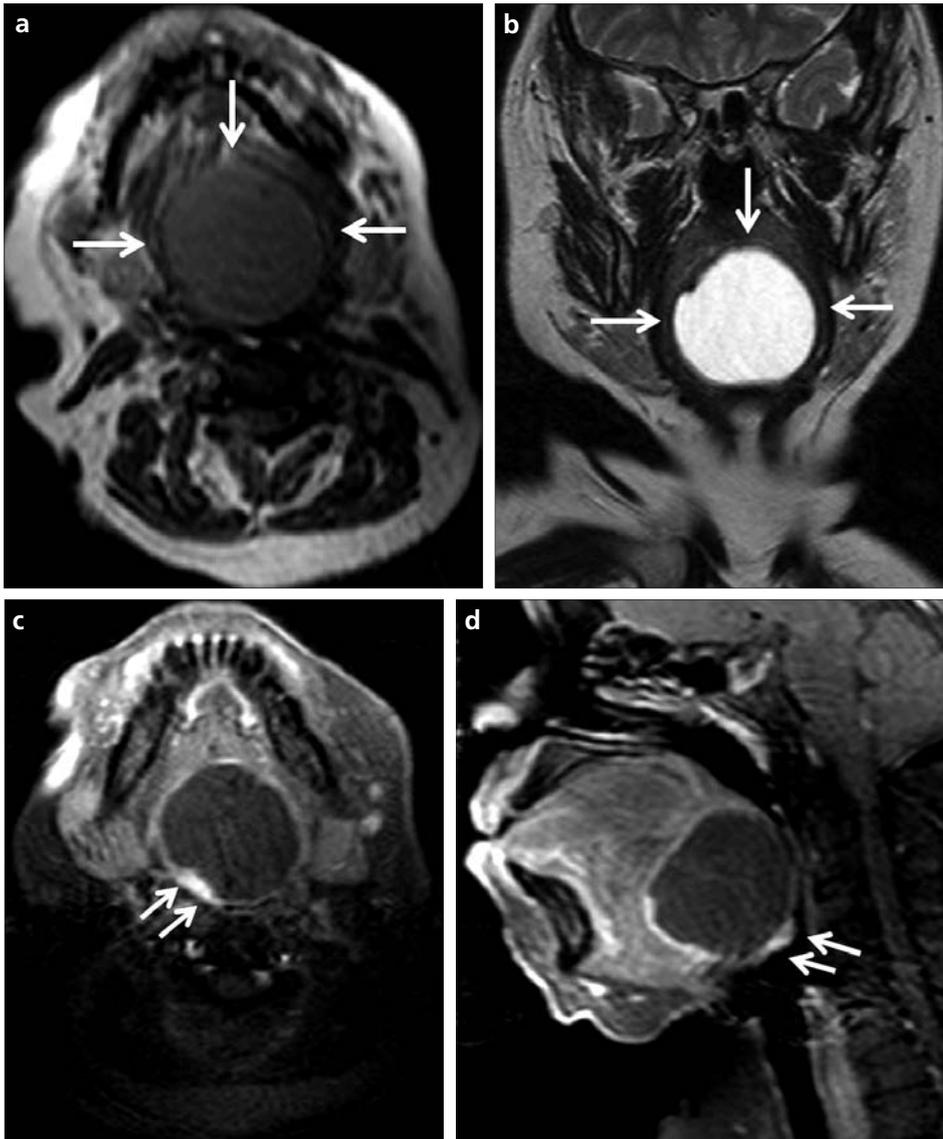


Figure 6. a–d. A 48-year-old woman with a giant thyroglossal duct cyst. Axial T1-weighted MR image (a) shows a well-defined cystic mass in the floor of the mouth at the tongue base, the classic location for a thyroglossal duct cyst. The increased signal intensity of the cyst is due to either proteinaceous content or a prior hemorrhage (a, arrows). Coronal T2-weighted MR image (b) reveals high hyperintensity of the cyst, with mural thickening (arrows). The axial (c) and sagittal (d) contrast-enhanced T1-weighted MR images with fat suppression show mild rim enhancement of the cyst with strong enhancement of the thickened wall due to residual thyroid tissue (arrows).

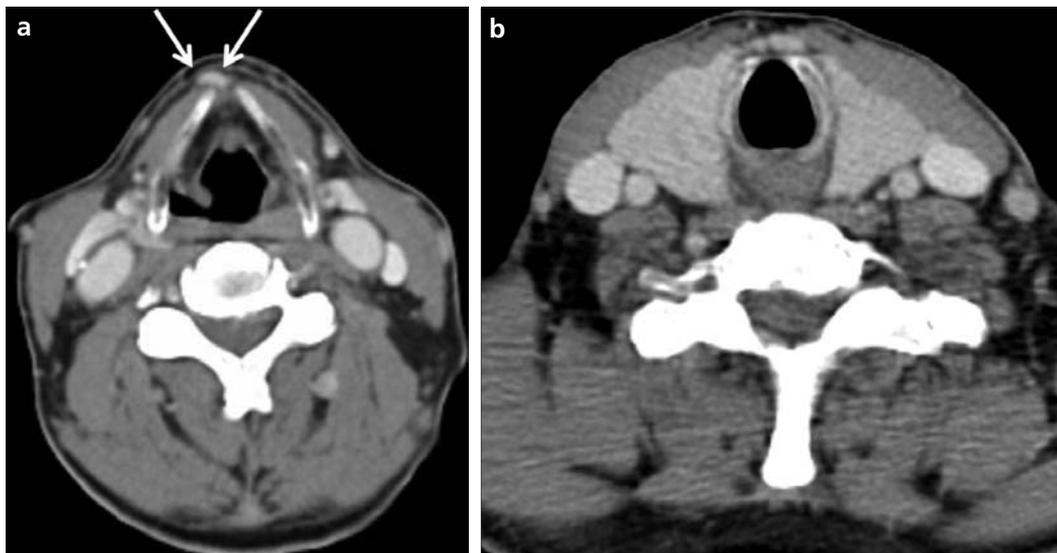


Figure 7. a, b. A 44-year-old woman with midline thyroid remnant tissue. Contrast-enhanced CT image (a) shows a small, strongly enhanced mass between the strap muscles at the anterior aperture of the thyroid cartilage (*arrows*). The thyroid gland has a normal appearance in the lower neck (b).

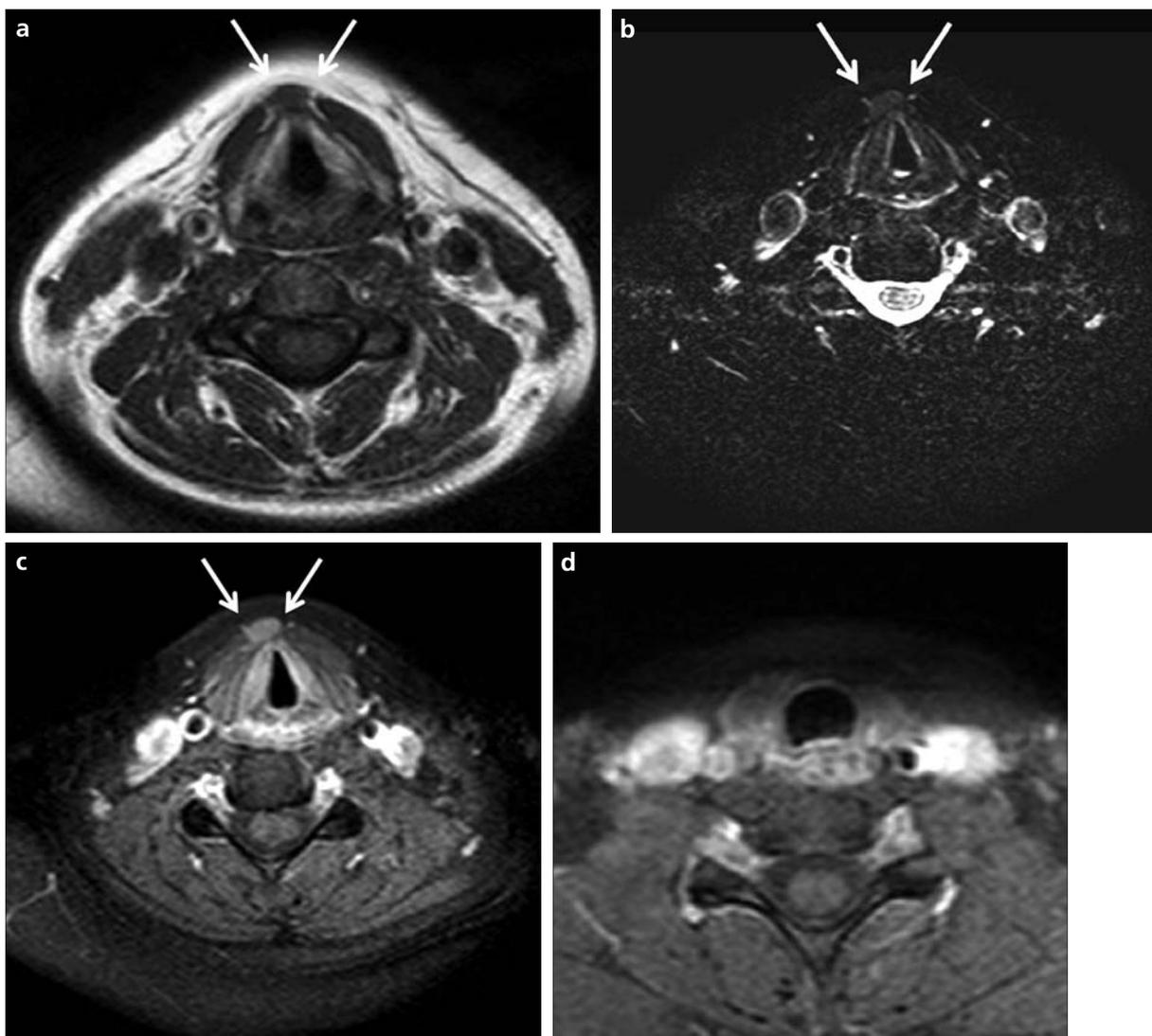


Figure 8. a–d. A 39-year-old woman with midline thyroid remnant tissue. T1- (a) and T2-weighted (b) MR images show a superficial lesion with intermediate signal intensity in the right paramedian region, at the anterior aperture of the thyroid cartilage (*arrows*). Contrast-enhanced T1-weighted MR images (c, d) show strong homogeneous enhancement of the mass (c, *arrows*). This lesion has the same signal intensity and enhancement pattern as the thyroid gland in all sequences.

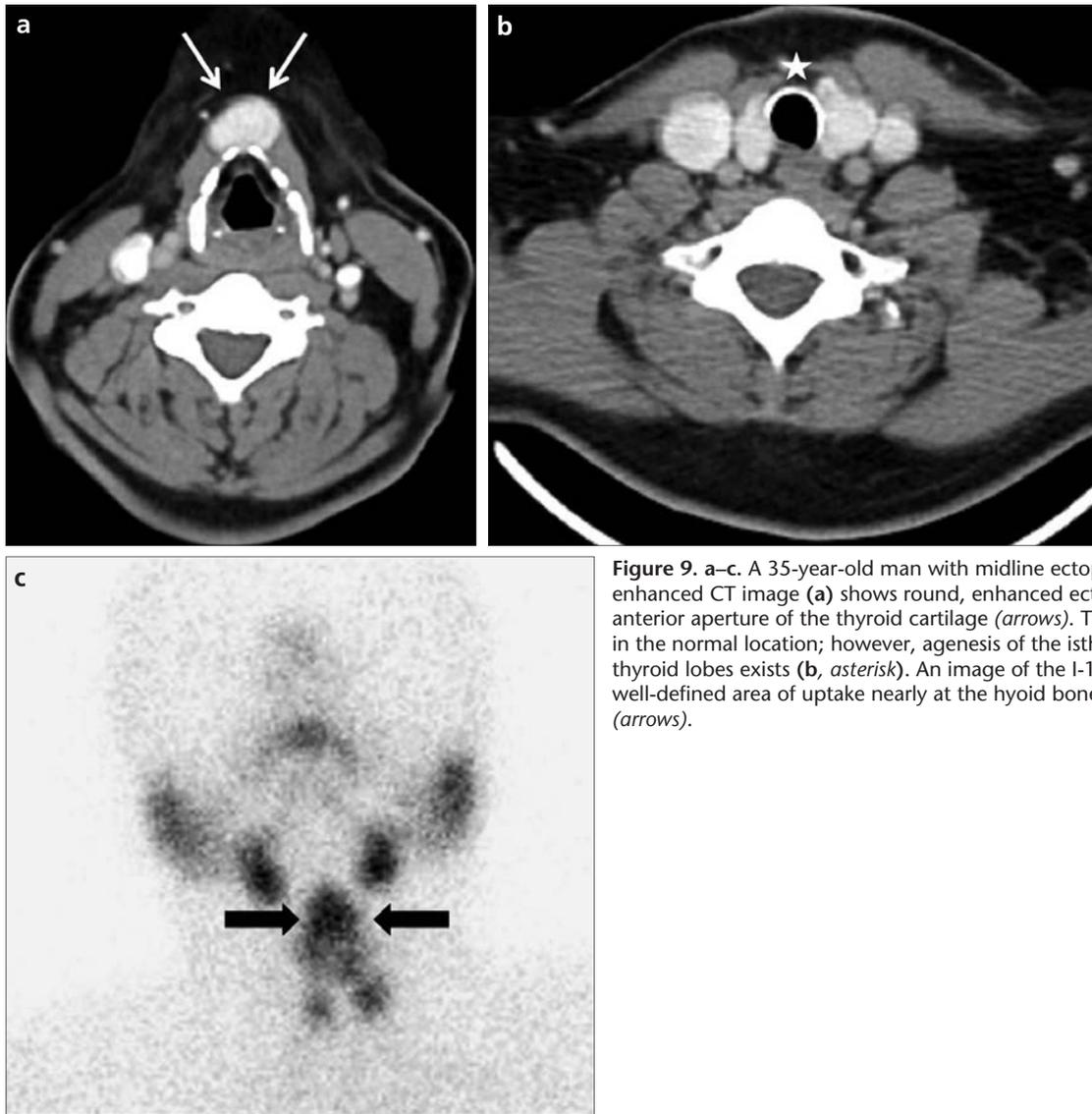


Figure 9. a–c. A 35-year-old man with midline ectopic thyroid tissue. Contrast-enhanced CT image (a) shows round, enhanced ectopic thyroid tissue at the anterior aperture of the thyroid cartilage (arrows). The thyroid gland is located in the normal location; however, agenesis of the isthmus with hypoplastic thyroid lobes exists (b, asterisk). An image of the I-131 scan (c) illustrates a well-defined area of uptake nearly at the hyoid bone, located at the midline (arrows).

tissue may appear as a lymph node or solid mass lesion. Thyroid scintigraphy shows iodine uptake by ectopic tissues.

Lateral thyroid ectopia

Lateral ectopia is an unusual developmental anomaly of the thyroid gland. Arrested descent of the lateral anlagen during the fetal period causes ectopic thyroid tissue on the lateral neck region, between the submandibular space and the proper location of the thyroid gland (22). Ectopic tissue usually extends into the parapharyngeal space along this path (23). Patients are typically young adults who present with a slowly growing painless neck mass. It is important to exclude malignancy, which shows a prominent enhancement pattern similar to that of lateral ectopic thyroid tissue. The

differential diagnosis includes metastasis from a primary thyroid carcinoma, other metastatic lymph node involvement, and paragangliomas, particularly glomus caroticum and vagale.

The role of sectional imaging techniques in the diagnosis of a neck mass is well established. MRI and CT characteristics of ectopic tissues are similar to those of the normal thyroid gland, showing a well-defined mass, either in continuity with the normally located thyroid gland or with approximately equal signal intensity. Contrast-enhanced imaging reveals intense enhancement (Figs. 10 and 11). On US examination, ectopic tissue usually appears as a soft mass near the submandibular gland in the lateral neck region. Thyroid scintigraphy shows iodine uptake by the ectopic tissue(s). In

the presence of goitrogenic or neoplastic disorders, the radiological appearance of ectopic tissue becomes heterogeneous and unpredictable.

Rare locations of thyroid ectopia in the head and neck

Ectopic thyroid is rarely located in the neck, sublingual fossa, submandibular space, parotid space, larynx, and orbita.

Conclusion

The imaging characteristics and locations of thyroid ectopia and aberrant thyroid tissue vary widely. Thyroid ectopia are most often identified as incidental findings on CT or MRI. In our experience, small midline thyroid ectopia is not rare and should be considered in the differential diagnosis of solid, enhancing neck lesions.

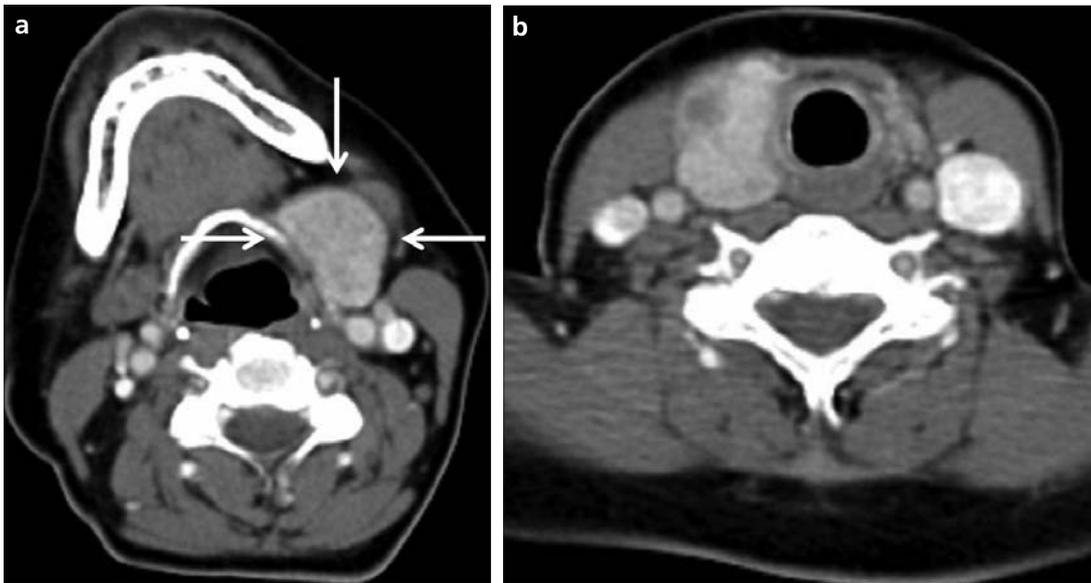


Figure 10. a, b. A 42-year-old man with lateral ectopic thyroid tissue. Contrast-enhanced CT images (a, b) show ectopic thyroid tissue in the submandibular and parapharyngeal regions at the hyoid bone level. The left submandibular gland is pushed anterolaterally by the ectopic tissue (a, arrows). The right thyroid lobe is visualized in the normal location and incidentally detected as a hypodense nodule in the right lobe. Agenesis of the isthmus and left thyroid lobe is noted (b).

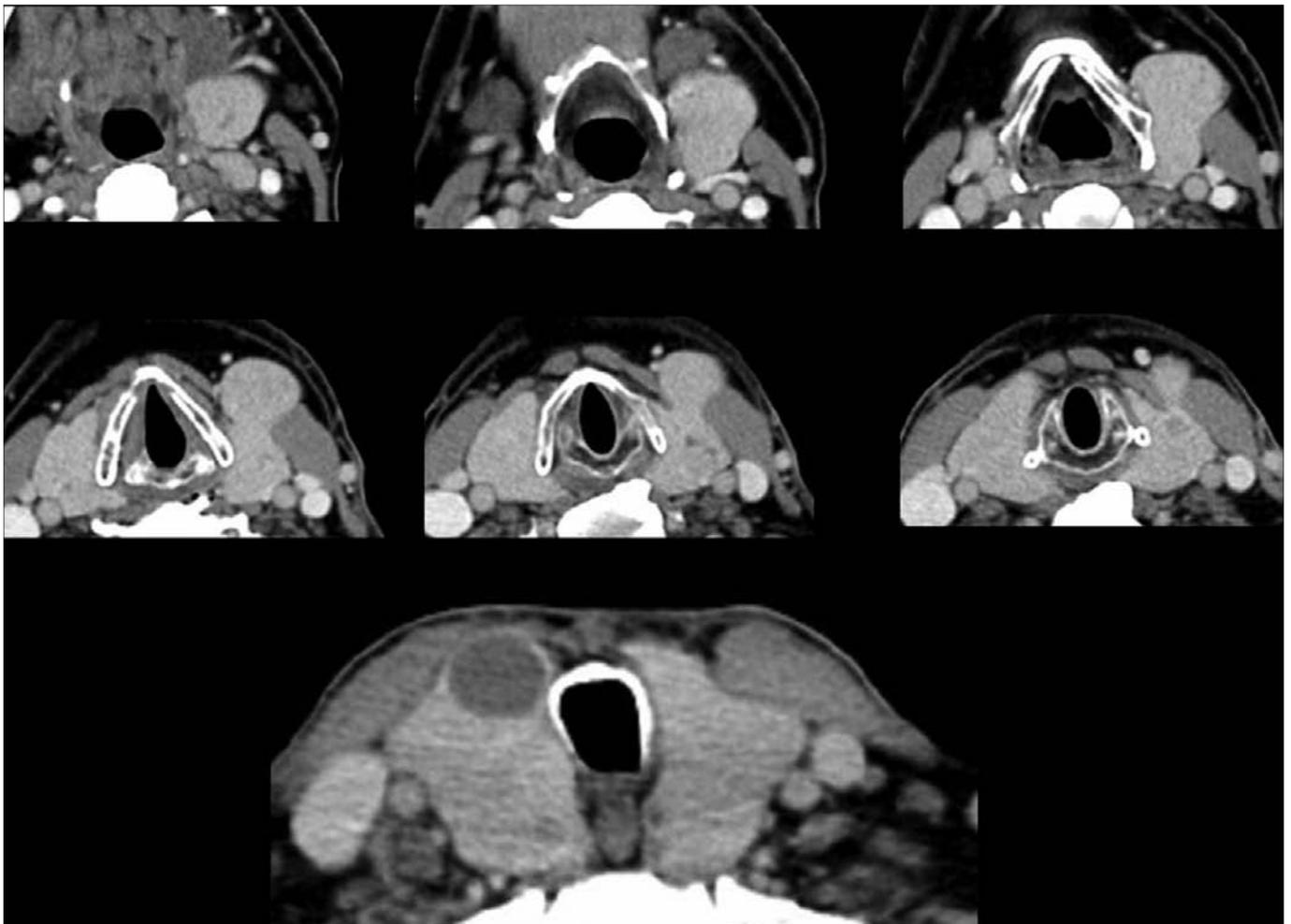


Figure 11. A 59-year-old man with lateral ectopic thyroid tissue. Contrast-enhanced CT image series show a homogeneous, dumbbell-shaped mass with uniform enhancement that extends into the submandibular region from the left thyroid lobe region. No isthmus is present.

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