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Case Report

Incidentally detected ectopic thyroid in juxta cardiac location—Imaging and pathology

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ABSTRACT

Ectopic thyroid gland is a developmental anomaly that results from the arrest of thyroid tissue along its path of descent from the floor of mouth to the pre tracheal position in the lower neck. It is typically found along the thyroglossal duct with the base of the tongue being the most common site. Apart from mediastinal extension of goiter, the incidence of true intrathoracic ectopic thyroid tissue is rare. Presence of ectopic thyroid has been reported not only in the chest but also in the abdomen and pelvis. Pericardial and intracardiac locations are extremely uncommon and right ventricle location is predominant among the described cases. We describe a case of incidentally detected ectopic thyroid tissue in a rarer location—adjacent to the left atrium. The patient, who had undergone a nephrectomy for renal oncocytoma 5 years ago, presented with unintentional weight loss and left sided flank pain, prompting a workup to rule out abdominal malignancy. Findings on the computed tomography (CT) scan of the abdomen and pelvis prompted further investigation including a chest CT which showed a heterogeneously enhancing mass near the left atrium. Given its location, further radiological investigations played an important role in eliminating the differential diagnosis of paraganglioma. The mass was surgically resected and discovered to be a hyperplastic thyroid nodule on histologic examination.

Case report

A 69-year-old Caucasian male presented to the clinic complaining of left sided flank pain and unintentional weight loss of 10-15 pounds over the last couple of months. He complained of 5 years of periodic chest pain which was intermittent, nonexertional, and without discomfort or radiation. He had had a negative stress test 1 year ago. The patient denied shortness of breath, orthopnea, soft tissue edema, or palpitations. On physical examination, no abnormality was detected.

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Five years ago he had undergone a right partial nephrectomy for a renal oncocytoma.

Given the past history and current presentation, the possibility of a new or recurrent malignancy was considered. Consequently, a computed tomography (CT) scan of the abdomen and pelvis with intravenous (IV) contrast was performed. No significant abnormality was found in the abdomen or pelvis. However, a heterogeneously enhancing mass was partially seen in the mediastinum, posterior to the aortic root, on limited images of the lower chest. Retrospective review of previous abdomen CT scans dating back 4 years showed the mediastinal mass to be grossly stable on limited images of the lower chest. A subsequent transthoracic echocardiogram showed a mass abutting the left atrium; the report raised the possibility of myxoma. In order to better visualize the mass, a CT scan of the chest with IV contrast was performed. It revealed a well-defined, high attenuation (HU 184) mass measuring about 4.5 cm × 3.3 cm × 2.7 cm, located posterior to the root of aorta and extending superiorly behind the tubular portion of the ascending aorta (Fig. 1a). The patient was still in the department and a limited noncontrast scan was performed to image the mass (HU 72) that confirmed a true contrast enhancement (Fig. 1b). On the coronal reformatted image, the lesion was located between the roof of the left atrium and the right pulmonary artery (Fig. 1c). The left atrial roof consists of the upper wall of left atrium and upper pulmonary veins opening into the left atrium. The fat plane between the lesion and the adjacent left atrium was maintained, which ruled out the possibility of atrial myxoma. The fat planes with other mediastinal structures including the aorta were also maintained. There were no other mediastinal nodules, pericardial effusion, or other abnormalities in the chest.

The patient did not have systemic symptoms related to excess adrenaline or serotonin secretion (ie, flushing, palpitations, or hypertension). Subsequently, an I-123 metaiodobenzylguanidine (MIBG) scan was performed with single-photon emission computed tomography (SPECT). There was no radiotracer uptake in the mass (Fig. 2). Given the size and enhancement of the mass, its juxta-cardiac location, and the patient's history of a renal malignancy, the mediastinal mass was surgically resected using a sternotomy approach. During surgery, the mass was easily separable from the adjacent pericardium and did not adhere to any other mediastinal structures. Pathologic examination of the lesion showed thyroid tissue with hyperplastic nodules without any malignant features (Fig. 3). Pre and postsurgery thyroid profiles were within normal limits: [presurgery thyroid profile: FT4–1.03 ng/dl; TSH–1.196 mc int units/ml; postsurgery thyroid profile: FT4–0.94 ng/dl; TSH–1.87 mc int units/ml]; (normal range: FT4–0.93-1.70 ng/dl; TSH–0.27-4.20 mU/l). Retrospective review of CT images showed normal size, location, and appearance of the native thyroid gland. No other foci of ectopic thyroid were seen in the mediastinum.

Discussion

Ectopic thyroid tissue is a rare developmental anomaly occurring in 1 in 100,000-1 in 300,000 people [1]. Ectopic thyroid may be classified into 2 types. Type 1 ectopic thyroid is present in the absence of thyroid gland at the normal location anterior to the trachea. Type 2 refers to ectopic thyroid in addition to thyroid gland present at its normal location [2]. We have presented an instance of type 2 ectopic thyroid in this report.

The thyroid gland is located in the subcutaneous plane of the anterior neck between the 2nd and 5th tracheal rings. It is the first of the body's endocrine glands to develop, on approximately the 24th day of gestation. The gland originates as a proliferation of endodermal epithelial cells on the median surface of the developing pharyngeal gut between the 1st and 2nd pharyngeal pouches. The thyroid primordium penetrates the underlying mesoderm and descends, anterior to the pharyngeal gut, as a bilobed diverticulum [3,4]. When present, ectopic thyroid tissue usually lies along the normal path of embryological descent and therefore occurs in the midline anywhere from the foramen cecum at the base of the tongue to the anterior midline of the neck. Lingual thyroid is the most common location of ectopic thyroid—accounting for 90% of cases [5]. Ectopic thyroid has been reported not only in the chest (eg, within thymus, trachea, esophagus, lung, and ascending aorta) but also in the abdomen and pelvis (eg, within liver, gall bladder, duodenum, pancreas, and vagina) [3]. Although relatively rare, instances of ectopic thyroid occurring in the heart have been reported and may be due to the anatomical relationship between developing thyroid primordium and the bulbus cordis of the developing heart. Due to this association with the bulbus cordis, ectopic thyroid may develop in the right ventricle when the heart and great vessels descend from the neck to the chest during development [6,7]. Therefore, cases of ectopic thyroid in the heart are generally restricted to the right ventricle—unlike our patient whose ectopic tissue arose near the roof of the left atrium.

Ectopic thyroid tissue is often discovered incidentally since it is usually benign and the patients are euthyroid [8]. Patients with large nodules may present with symptoms due to mass effect on adjacent structures [9]. Given the wide variety of possible locations of ectopic thyroid and its benign nature, it may be incidentally detected on CT scans done for nonrelated reasons. Various modalities have been deemed useful for evaluation. Ultrasound has advantages of being noninvasive with lack of radiation exposure. It is extremely useful for diagnosing a cervical ectopic thyroid [10]. Computed tomography is a very sensitive modality for detecting extracervical ectopic thyroid. A CT scan without contrast often shows a slightly increased attenuation (70 HU+/− 10) in comparison to skeletal muscles due to the presence of higher iodine content within thyroid tissue [11]. Contrast enhanced CT scan shows an avid enhancement of the normal thyroid tissue. MRI scans have also been shown to be helpful in visualizing ectopic thyroid with T1 showing isointense to mildly hyperintense and T2 showing mild hyperintensity of normal thyroid tissue compared to muscle. Radionuclide Iodine uptake scans (thyroid scintigraphy) are considered excellent for visualization of ectopic thyroid tissue [11]. It may also help to determine if there is metastasis of malignant thyroid tissue even though this is uncommon in ectopic thyroid [12]. Ultimately, however, a significant proportion of asymptomatic ectopic thyroid cases are ultimately diagnosed only by histological analysis after the mass is excised [6].

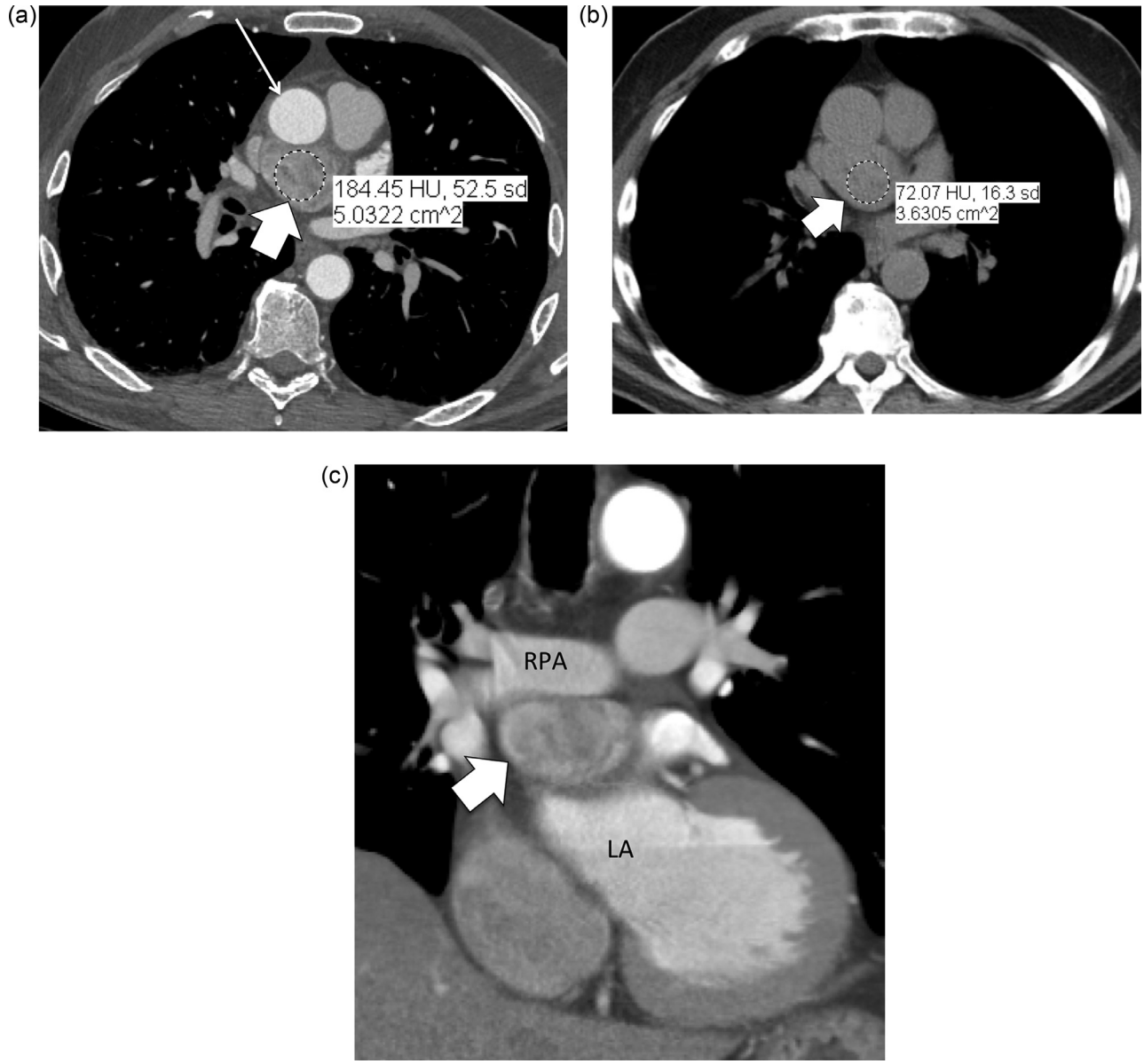

Fig. 1 – Axial contrast enhanced (a) and noncontrast (b) computed tomography images show a heterogeneously enhancing mass (block arrow) in the middle mediastinum behind the aortic root extending superiorly behind the tubular portion of the ascending aorta (thin arrow). Coronal CT image (c) shows location of the mass (block arrow) between roof of the left atrium (LA) and right pulmonary artery (RPA) with maintenance of fat planes.

In our case, the chest CT with IV contrast showed a high attenuation mass posterior to the aortic root. Sometimes complex pericardial or foregut duplication cysts can show higher attenuation due to proteinaceous contents or milk of calcium and can be confused with enhancing lesions on contrast study. Although, the degree of higher attenuation in this case (HU 184) and heterogeneous appearance is unusual for a complex pericardial cyst, a foregut duplication cyst remained a possibility. However, subsequent noncontrast CT was helpful to rule it out as the mass showed intermediate attenuation (HU 72) and confirmed a true contrast enhancement. The differential diagnosis for contrast enhancing mediastinal masses includes aneurysm, carcinoid, paraganglioma, unicentric Castleman's disease, ectopic thyroid, and parathyroid tissue, and certain metastases such as melanoma, renal cell cancer, thyroid cancer, and high grade sarcoma. As this patient's mass lacked communication with the aorta or tracheobronchial tree, the differentials of aneurysm and carcinoid, respectively, were excluded. Although our patient had a history of renal malignancy (oncocytoma), the abdomen CT scan was normal and the mediastinal mass had been grossly stable over the last 4 years, making metastasis extremely unlikely.

Due to the location and hyper enhancing nature of our patient's mass, the possibilities of paraganglioma or solitary lymph node conglomerate such as from unicentric Castleman's disease were considered. Thoracic paragangliomas are

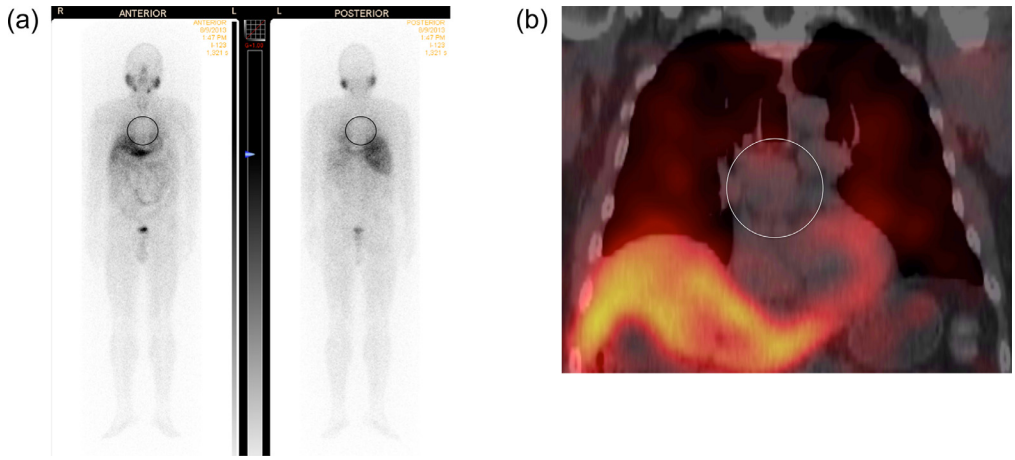

Fig. 2 – Planar (a) and coronal SPECT-CT (b) images from the I-123 MIBG scan show absence of tracer uptake within the mass (circle).

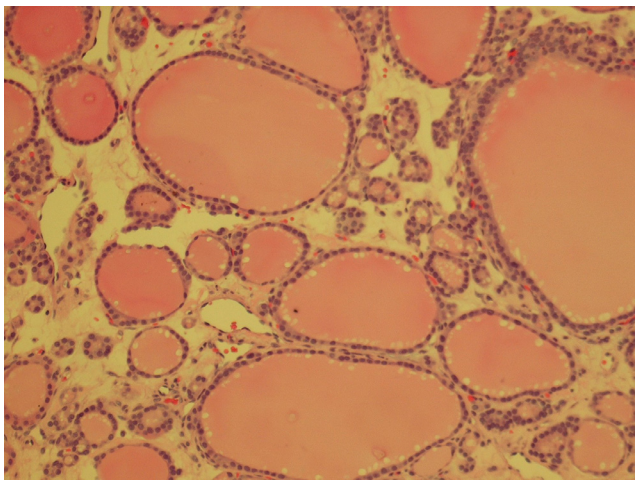

Fig. 3 – Photomicrograph of the nodule under higher magnification (x100) shows multiple active thyroid follicles.

rare and can be found along the paravertebral region, or in the pericardium, near the roof of the left atrium, anterior to the aortic root as markedly enhancing masses on contrast enhanced CT. Intrapericardial paragangliomas are slow growing tumors that arise from neural crest remnants and are often nonfunctioning. MIBG scintigraphy is positive in functioning paragangliomas with sensitivity of 90% [13]. ^{111}In pentreotide scan has a higher sensitivity of 94% [14]. In our case, only I-123 MIBG scintigraphy was performed and was negative. However, the possibility of a nonfunctioning paraganglioma could not be ruled out. Castleman's disease is a complex lymphoproliferative disease and seen as avidly enhancing lymphadenopathy. There are two clinical forms of the disease: unicentric and multicentric. Unicentric Castleman's disease is typically asymptomatic and occurs in the thorax. The intermediate attenuation of HU 72 (higher than skeletal muscles) on noncontrast CT and hyperenhancement after IV contrast is characteristic of thyroid tissue. However, ectopic thyroid tissue is commonly seen in the anterior mediastinum. Due to the location of the mass near the aortic root and normal thyroid function tests in our case, ectopic thyroid was not considered in the clinical differential and the diagnosis came as a surprise after surgical excision.

Treatment options for ectopic thyroid include resection, as was done in our case, especially in cases of large masses which impinge on surrounding structures. Functional hypo or hyperthyroidism, if present, is treated medically. In cases of type 1 ectopic thyroid tissue, complete excision will require the patient to receive lifelong thyroid hormone supplementation.

In our case, there were no specific imaging features which could resolve the differential possibilities in that location and the diagnosis came as a surprise upon pathologic examination.

Teaching point

Extracervical ectopic thyroid tissue is rare and can be present at various locations within the chest, abdomen, and pelvis. These foci are asymptomatic and may be detected incidentally on imaging. It is important for radiologists not only to be aware of these rare locations and imaging characteristics, but also the roles of various imaging modalities. CT without and with contrast shows higher attenuation and marked contrast enhancement respectively, whereas Iodine scintigraphy with single-photon emission computed tomography imaging provides functional and anatomical characterization.

Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:10.1016/j.radcr.2018.06.004.

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