Clinical case

Ectopic thyroid tissue in the anterior mediastinum with a normally located gland: A case report

Une thyroïde ectopique médiastinale antérieure en présence d'une thyroïde cervicale :
à propos d'un cas

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Abstract

Ectopic thyroid tissue occurs in about 7 to 10% of the general population. Localization in the anterior mediastinum is rare: seven cases reported in the literature, most associated with thyroid dysfunction. We report the case of a 77-year-old woman who presented ectopic thyroid tissue in the anterior mediastinum and a normally-located normally-functioning gland.

Résumé

L'ectopie thyroïdienne se définit comme la présence de tissu thyroïdien hors de sa localisation anatomique habituelle : la région cervicale infrathyroïdienne. Sa prévalence est estimée de 7 à 10 % dans la population, mais sa localisation médiastinale antérieure est rare : sept cas décrits dans la littérature et son association à une thyroïde cervicale fonctionnelle exceptionnelle. Nous rapportons l’observation d’une patiente de 77 ans présentant cette particularité anatomique.

1. Introduction

The discovery of ectopic thyroid tissue is exceptional, with an incidence of about one case in 300,000. Localization in the mediastinum is very rare. Over the last 30 years, only seven cases have been described in the literature, including five involving the anterior mediastinum and two the posterior mediastinum [1,2]. Thyroid tissue appears during the fourth week of embryonic development on the medium surface of the developing pharyngeal floor: the foramen cecum. It then descends along the thyroglossal duct to reach, at the seventh week of gestation, its final anterotracheal position. Ectopic thyroid tissue can be located all along this migration route, in the neck, pharynx, larynx, esophagus, trachea, aorta, mediastinum, or tongue, the most common localization of ectopic thyroid tissue (90% of cases) [3]. The molecular mechanisms implicated in this dysgenetic process are not fully elucidated. Mutations of genes regulating thyroid gland development have been postulated [4]. We report a fortuitous discovery of ectopic thyroid tissue in a 77-year-old patient undergoing explorations for chronic dyspnea.

2. Case report

A 77-year-old woman with dyspnea was referred for emergency care. Past history included hypertension treated with irbesartan and dyslipidemia treated with fenofibrate. Clinically, the patient presented dyspnea (NYHA II) which had worsened progressively over 1 month but was not associated with functional impairment or alteration of the patient’s general health status. The physical examination found polypnea (>15/min) with no overt signs of respiratory failure, heart failure or thromboembolism. Oxygen saturation was 95% (FiO2 60%) and blood pressure 130/70 mmHg. There was no fever.

Blood tests showed normal cell counts, no signs of inflammation, normal LDH level (145 U/L), and non-compensated respiratory alkalosis (pH 7.47, PO2 73 mmHg, PCO2 26 mmHg,
bicarbonates 18.9 mmol/L, base excess −4.8 mmol/L, alkaline reserve 19.7 mmol/L). Angiotensin converting enzyme level was normal. Search for tuberculosis was negative: PPD measured at less than 5 mm; negative gastric aspirations. The electrocardiogram showed a regular sinus rhythm with normal repolarization.

The transthoracic echocardiography showed a non-dilated left ventricle with no hypertrophy and no dilatation of the right cavities or pulmonary hypertension; the pericardium was normal and the ventricular function satisfactory (ejection fraction 68%). Computed tomographic angiography ruled out pulmonary embolism but fortuitously revealed a right lateral superior mediastinal mass in the pretracheal retrocaval space measuring 43 × 35 × 27 mm. The mass was polylobular and exhibited heterogeneous enhancement with contrast injection. There were no signs of compression of adjacent organs (Figs. 1–3).

Mediastinoscopy was performed to obtain a biopsy. Pathology examination of the biopsy specimen showed thyroid tissue with variably-sized vesicles boarded with squamous cuboid epithelium, zones of interstitial fibrosis and no signs of malignancy, nuclear atypia or mitosis. Neck ultrasound showed a normally positioned thyroid gland measuring 18.7 mm with discrete heterogeneous structure, no nodules and no lymph node enlargement. The patient was euthyroid (TSH 1.80 µU/ml, free-T4 17.8 pmol/L, free-T3 4.3 pmol/L). CEA level was normal (2.1 ng/ml) as was thyrocalcitonin less than 2 ng/L. The autoimmune work-up was negative (antithyroglobulin, antithyreoperoxidase, antithyrotropin receptor antibodies). Iodine-123 scintigraphy showed homogeneous tracer uptake in the thyroid gland in the neck and confirmed the presence of ectopic thyroid tissue in the superior mediastinum.

Complete resection of the mediastinal mass was performed. There was no sign of malignancy. The surgeon did not note a channel linking the ectopic tissue with the thyroid gland.

3. Discussion

Several diagnostic hypotheses including malignant mediastinal adenopathy related to a lymphomatous process or metastases from locoregional (lung, esophagus, breast, thyroid) or distant (uterus, kidney, gut) cancers would be compatible with the clinical presentation and ultrasound findings in this elderly patient despite the absence of signs of compression, the lack of any impact on general status, and regular gynecological check-ups. Considering the patient’s age and the lack of compressive features, nodal tuberculosis could be a putative diagnosis, much like sarcoidosis. A bronchial cyst could also be discussed despite the discordant radiographic aspect. A histology study was necessary to obtain the diagnosis of ectopic thyroid tissue.

One of the unusual features of this case is the combination of ectopic thyroid tissue in the mediastinum and a normally functioning thyroid gland in the neck. This is a rare situation and has not been described in the literature. Similarly, for submandibu-
lar ectopic thyroid tissue, association with a normally positioned gland has been reported in only one case [5]. Rereading the computed tomography scans of our patient did not reveal any vascular malformation in the neck or mediastinum which might have favored excessive migration. Similarly, no evidence of abnormal ascending migration of the thyroid during embryogenesis could be found.

Most patients with ectopic thyroid tissue remain symptom free, although some patients exceptionally develop signs of compression of the upper airways, hypothyroidism, or more rarely hyperthyroidism [4,6–8]. In our patient, since a cardiorespiratory disorder had been ruled out and considering the normal blood tests and the lack of compression, the dyspnea was attributed to physical deconditioning in a context of progressive loss of autonomy.

Although the risk is generally higher than for a thyroid gland in a normal anatomic position, it is exceptional for ectopic thyroid tissue to degenerate (papillary, follicular, Hürthle cell, anaplastic carcinoma). Thirty cases of malignant degeneration of ectopic thyroid tissue in the tongue have been described [9–11]. There is no complementary test capable of ruling out the presence of a neoplastic process. Iodine-123 scintigraphy is useful for diagnosis and should be performed preoperatively: a positive scintigram is in favor of the presence of thyroid tissue but a negative scintigram cannot rule out the diagnosis [12,13]. The gold standard treatment for ectopic thyroid tissue in the mediastinum is surgical resection, especially in the presence of signs of local compression: trachea, esophagus, superior vena cava [12–14]. Resection is usually achieved via thoracotomy or median sternotomy. Similarly, ectopic thyroid tissue within a nodal tissue in the neck should be removed due to the risk of metastasis from a well-differentiated thyroid carcinoma [15].

Our case suggests that ectopic thyroid tissue should be entertained as a tentative diagnosis in patients presenting a mass in the anterior mediastinum, even in those with a normally functioning gland in an anatomic position since the potential for degeneration requires systematic surgical resection.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

References