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Subhyoid ectopic thyroid with mediolateral location in a developmentally retarded child with hypothyroidism in an iodine deficiency region

Hakan DEMIR,* Fatma BERK,* Serkan ISGOREN,* R. Okan ERDINCLER,*
Ercüment CIFTCI** and Cumali AKTOLUN*

*Departments of *Nuclear Medicine and **Radiology, Kocaeli University Medical School, Kocaeli, Turkey*

The authors present a case of subhyoid ectopic thyroid with mediolateral location diagnosed by technetium-99m pertechnetate scan. The ectopia was associated with hypothyroidism and developmental retardation.

Key words: thyroid, thyroid ectopia, thyroid scan, hypothyroidism, developmental retardation

INTRODUCTION

ALTHOUGH it is a rare congenital abnormality, ectopic thyroid tissue is most commonly seen in the neck either in a median or a lateral location. Its location in relation to the tongue, salivary glands, glottis or hyoid bone determines its terminological definition, lingual thyroid being the most common form.[1–5] Median location is more common than lateral location with the latter being more frequently associated with malignant diseases.[6–8] With a higher incidence in females, it can be seen at any age, but it is more commonly detected before the age of 20, probably due to dramatic physiologic hormonal changes before this age. Ectopic tissue is the only functioning gland in a some of the patients, whereas in others one or more ectopic glands are seen in addition to the actual thyroid gland in its expected location in the neck.[2,3,9–13] Calcification, benign hyperplasia or malignant transformation can be seen in the ectopic tissue.[6–8]

Hormonal changes can cause enlargement in size or deficiency or excess in circulating hormone levels, resulting in evident clinical signs and symptoms.[11,13–15] Dysphagia, dysphonia and "fullness in the mouth" are the most common clinical conditions seen in the clinical history and on examination.[16] Of the hormone related disturbances, the most unfortunate cases are children with undetected, subtle hypothyroidism causing developmental retardation.[9,10,12,13] In this report, we present a case of subhyoid ectopia of the thyroid in a developmentally retarded child with clinically subtle hypothyroidism.

CASE REPORT

A 4-year-old girl had been complaining of constipation and coarseness of hair for a long time. Although she had been examined a couple of times, mental and physical development retardation was overlooked by both parents and the physicians who treated the constipation medically. When the patient was referred to Kocaeli University Hospital, mental and physical development retardation was noted, and accordingly hypothyroidism was included in the differential diagnosis. The patient was referred to the Nuclear Medicine Department for functional assessment of the thyroid gland.

Hormone measurements revealed increased thyroid stimulating hormone (TSH > 100 μIU/m*l*, normal range 0.270–5.010 μIU/m*l*) and total T4 (TT4 = 26.70 μmol/*l*, normal range 58.05–174.41 μmol/*l*). A thyroid scan was scheduled for her. She was injected with 74 MBq (2 mCi) Tc-99m pertechnetate intravenously. Thyroid scan, performed with a pin-hole collimator, demonstrated no uptake in the usual site of the thyroid gland. The scan was repeated but no thyroid tissue was seen in the neck. Due to our awareness of a high incidence of congenital endocrine abnormalities including ectopic thyroid in our service area,[9,10] we repeated the scan with a parallel hole collimator. A well-defined small area of uptake near the left submandibular salivary gland, located slightly left of

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For reprint contact: Cumali Aktolun, M.D., M.Sc., Department of Nuclear Medicine, Kocaeli University School of Medicine, Derince, Kocaeli, TR-41900, TURKEY.
E-mail: aktolun@hotmail.com

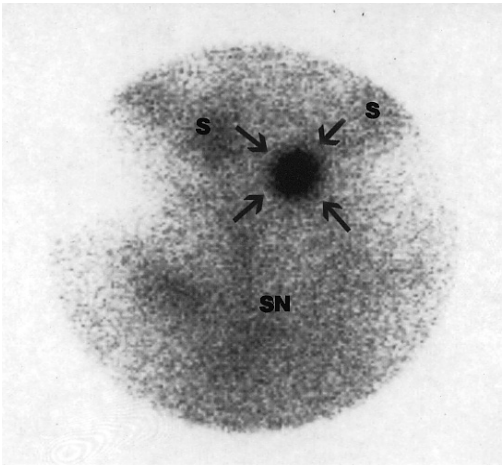
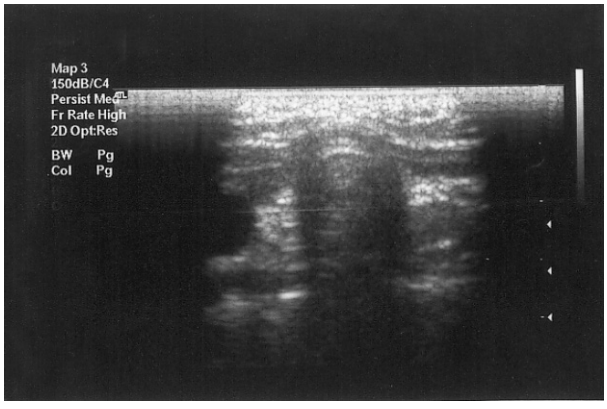


Fig. 1 After failing to see any uptake in the usual thyroid location on pin-hole image, the patient was imaged again using a LEAP collimator. A well-defined area of uptake is evident nearby the left submandibular salivary gland, located slightly to the left of the midline (*arrows*), but no uptake at the thyroid location on the thyroid Tc-99m pertechnetate scan.

Fig. 3 Contrast enhanced CT image showed a hyperdense, well-circumscribed tissue suggesting ectopic thyroid located at the subhyoid region confirming scintigraphic diagnosis.



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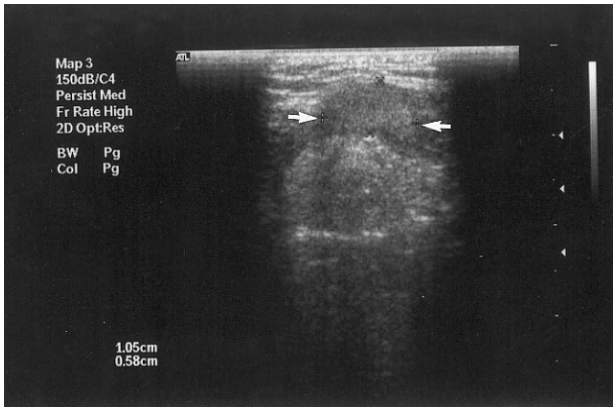

B

Fig. 2 Transverse sonogram of the neck at the level of thyroid gland (A) showed no normal thyroid tissue. A suspicious tissue for ectopic thyroid gland in subhyoid region (*arrows*) was observed, but a firm sonographic diagnosis of ectopic thyroid tissue could not be made (B).

the midline was detected (Fig. 1), and no radioactivity accumulation at the usual thyroid location was seen. Scans were repeated after sipping water and the same appearance persisted. After the thyroid scan, ultrasound (US) and contrast enhanced computerized tomography (CT) were scheduled in order to confirm the diagnosis of ectopic thyroid as we tried to avoid invasive diagnostic methods including biopsy. US demonstrated no thyroid tissue in normal thyroid localization but did not help us to definitely confirm the ectopic thyroid tissue as ultrasonographic examination has serious limitations in the subhyoid region (Fig. 2). After careful consideration of risk/benefit analysis, a contrast CT was performed. It showed a contrast enhanced hyperdense focus suggesting ectopic thyroid tissue in the subhyoid region, located slightly left of the midline (Fig. 3).

Supplementation of thyroid hormone was started. Constipation and hair coarseness problems disappeared gradually in the first month. No surgical intervention was considered with current clinical findings.

DISCUSSION

Thyroid ectopia is a rare clinical entity, and its diagnosis is still a challenge requiring the use of several non-invasive methods before a histologically proven diagnosis is maintained.[9,10,18–20] Its significance stems from the fact that it may present itself in different clinical pictures. It may sometimes cause only enlargement related complaints but in other cases hormonal disturbances including hyperthyroidism, hypothyroidism and developmental retardation can be the dominant symptoms. Enlargement related changes include diffuse or nodular hyperplasia.[11,15]

Although extremely rare, cancer of ectopic tissue is of significant clinical importance due to difficulties involved in the diagnosis and therapy.[6,8] In addition, malignant transformation of ectopic tissue has to be differentiated from cervical lymph node metastasis of a malignant tumor in the actual thyroid gland in the normal location as their clinical management is almost totally different.

People living in the Karadeniz (Black Sea) region of Turkey have higher a incidence of thyroid diseases resulting from endemic iodine deficiency. Kocaeli University is situated in the western Karadeniz region, and therefore attracts a large number of patients from other parts of the Karadeniz region. It is our observation[9,10] that congenital thyroid abnormalities including ectopia and resultant developmental retardation are common in this region, and they might be associated with endemic iodine deficiency. This observation indeed needs confirmation through screening studies in a larger number of patients.

Clinical presentation determines the diagnostic tools to be employed in the work-up of these patients. If hormonal 'imbalance is the dominant clinical feature, the patient first undergoes serum analysis of thyroid hormones and some *in vivo* tests including T3 suppression test. These are followed by scintigraphic and/or ultrasonographic evaluation of the thyroid gland.[9,10,20,21] Radioactive iodine (I-131, I-123) and Tc-99m pertechnetate scanning are well known methods for evaluating the size, distribution, and activity of thyroid disease from other masses in the head and neck and superior mediastinum that are not of thyroid origin. Radioactive iodine is the isotope of choice for mediastinal imaging. Some statements in the literature say that the high photon energy of I-131 is necessary for penetration of the sternum. I-123 is cyclotron produced, has a short half-life, and contains radiopharmaceutical impurities of I-124 and I-125, depending on the method of production.[22] In children, Tc-99m pertechnetate is generally used, because it is trapped by the thyroid gland but not organified. It is thought to be safer and cheaper than I-131 and I-123 because it is a pure gamma emitter as opposed to a beta emitter, non-cyclotron product, and it has a half-life of only 6 hr.[23] In addition, we do not use I-123 routinely for thyroid scanning because of the high cost and availability problems. For those reasons we performed thyroid scanning with Tc-99m pertechnetate.

In many cases, a final diagnosis of ectopic thyroid is achieved at this stage, whereas more sophisticated and possibly more invasive diagnostic procedures are needed in a certain subgroup of patients, mainly in those whose primary clinical presentation is a result of enlarged ectopic tissue, causing a mass of unknown nature in various sites in the head and neck region. Malignant transformation of ectopic thyroid and metastatic enlargement of regional lymph nodes should be excluded in any patient with a mass of unknown nature.[24] It may therefore be necessary to use CT scan, MR imaging and FNA or excisional biopsy in these patients.[25–27]

In our patient, Tc-99m pertechnetate scan suggested an ectopic thyroid, and radioiodine scan with iodine-123 is the method of choice to confirm this diagnosis, but because we had a temporary radioiodine shipment problem the time, instead we had to use CT scan for confirmation and a detailed picture of the relation of this cervical mass to the other structures in the region. Considering the complex anatomic structure of the neck with several vessels, lymph nodes, muscles and bones, we performed contrast CT scan since we did not want to repeat the CT scan without contrast in such a young patient if we failed to visualize the relation of this cervical mass to the other structures (particularly the blood vessels). Although our clinical decision was in favor of performing CT scan with contrast in this patient, in similar clinical situations in future we recommend using radioiodine scan only, and if a CT is necessary, one should prefer plain CT since the thyroid tissue has an attenuation coefficient higher than that of muscle, which is thought to be due to high iodine content in thyroid tissue, suggesting that a plain CT scan without contrast would have been sufficient in this case.

As a general rule, thyroid ectopia of lateral location requires a more rigorous approach as the possibility of malignancy is more common than with head and neck masses of median location.[5–7,10] Considering the age of the patient and the excellent response to hormone supplementation, we avoided invasive sampling methods.

Once the presence of thyroid tissue of either a benign or malignant nature is detected, hormone suppression, surgery and/or iodine-131 therapy can be utilized in accordance with the final clinical diagnosis of the patient, availability of these treatment options and the patient's individual needs. Generally, ectopia causing dysphagia, dysphonia and severe cosmetic problems is treated by surgery regardless of its location unless it invades nerves and vital blood vessels.[28] Recent refinements in hormone and radioiodine therapy have significantly reduced the need for extensive surgical procedures even in patients with huge masses. Well-differentiated malignant tumors of the ectopia require a similar therapeutic approach with that applied in actual thyroid cancers.

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