An unusual presentation of the TENIS syndrome

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Abstract

Introduction: Papillary carcinoma is the commonest subtype of thyroid cancer. Patients are treated with radioactive iodine (RAI), and monitored with RAI scans, serum thyroglobulin levels, and cervical ultrasonography. Elevated serum thyroglobulin despite a negative RAI scan is termed the TENIS syndrome (Truncated Expression of the NIS; NIS represents the sodium-iodide symporter). Alternative imaging is then indicated.

Method/Result: A female Caucasian patient with papillary thyroid carcinoma had been followed up at our Thyroid Cancer clinic. A diagnostic scan revealed residual thyroid tissue. RAI ablation of the thyroid remnant was performed with 80 mCi of RAI. Two- and five-year follow-up scans were clear. However, thyroglobulin levels remained high, thus a diagnosis of TENIS syndrome was made.

An F-18 FDG PET/CT scan was performed which demonstrated inhomogeneous liver uptake (SUV > 6). Abdominal CT confirmed multiple liver lesions; ultrasound-guided FNA of these revealed several groups of malignant cells.

Conclusion: Persistently elevated thyroglobulin levels in the presence of a negative whole-body RAI scan are of great concern. In this patient with unusual liver metastases from papillary thyroid cancer, and relatively low thyroglobulin levels despite significant metastases, F-18 FDG PET/CT has once more proved its efficacy in the evaluation of patients with the TENIS syndrome.

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1. Introduction

Thyroid carcinoma, though a rare tumour in the setting of general oncology, is the commonest endocrine tumour (90%), and papillary carcinoma is its commonest subtype (70–80%) in iodine-replete populations.1,2 It is commoner in females (M:F ratio of 3:1) and has a peak age incidence of 30–50 years. Papillary thyroid carcinoma typically spreads to locoregional lymph nodes and to the lungs.
Following total thyroidectomy and radioactive iodine ablation, patients are monitored with whole-body iodine scans and serum thyroglobulin levels. In this setting, thyroglobulin is a sensitive tumour marker, even more so under the stimulation of thyroid stimulating hormone (TSH) (98% compared to 80% under TSH suppression). A varying proportion of patients (up to 56%) may exhibit elevation of thyroglobulin in the presence of a negative I-131 whole-body scan. This condition has been termed the TENIS syndrome (Truncated Expression of the NIS, NIS being an acronym for the sodium-iodide symporter). The syndrome has been attributed to several possible aetiologies, such as small dose of radioiodine used for the diagnostic scan, “stunning” of radioiodine (RAI) uptake by functional thyroid tissue, dedifferentiation of the carcinoma, loss of function mutation of the NIS, iodine ingestion/administration blocking RAI uptake, absence of immunoreactive cytoplasmic thyroglobulin and thyroxine, or to tumour size below gamma camera resolution. When compared to other alternative imaging agents, F-18 FDG PET/CT imaging, with a sensitivity of 85–94% and a specificity of 90–95%, has been declared by a consensus as the most suitable modality for detecting tumour spread and recurrence in such instances. This claim is substantiated by other studies. It was also observed that the sensitivity of FDG PET imaging increased in patients with the TENIS syndrome.

2. Method/case report

A 41 year old female patient known with papillary thyroid carcinoma had been followed up at our thyroid cancer clinic. Following completion thyroidectomy, she had a diagnostic radioiodine scan, which revealed the presence of residual thyroid tissue. RAI ablation of the thyroid remnant was performed with 80 mCi I-131 in December 2000. The patient then

<table>
<thead>
<tr>
<th>Table 1</th>
<th>Thyroglobulin, antithyroglobulin antibody and thyroid hormone assays at 5th year follow-up.</th>
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<tbody>
<tr>
<td>Tg (ng/ml)</td>
<td>Anti-Tg Antibody (ng/ml)</td>
</tr>
<tr>
<td>On T4 suppression</td>
<td>0.5</td>
</tr>
<tr>
<td>Off T4 suppression</td>
<td>10</td>
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</tbody>
</table>

* Tg: Thyroglobulin.
** TSH: thyroid stimulating hormone.

Figure 1  Whole-body radioiodine-131 scan.

Figure 2  Hepatic metastases as seen on patient’s F-18 FDG PET/CT scan.
underwent follow-up scans at 6 months, 1 year, 2 years and 5 years afterwards, which were all clear (see 5-year scan, Fig. 1). However, a sudden increase in the thyroglobulin (Tg) levels was noted at her five-year follow-up (Table 1); the patient was thus diagnosed as having the TENIS syndrome.

3. Result

PET/CT imaging (Figs. 2 and 3) demonstrated inhomogeneous uptake in the liver, with focal increased activity in the right lobe; in segments 4a, 5 and 8 (SUV 6.73). It also showed a pedunculated inhomogeneous mass arising from segment 5 of the liver (SUV 5.09). Abdominal CT scan findings were consistent with multiple liver lesions. Ultrasound-guided FNA biopsy of the liver metastases revealed several groups of malignant cells compatible with metastatic papillary thyroid carcinoma. The patient was counselled regarding her prognosis and maintained on l-thyroxine 150 µg daily.

4. Conclusion

Serum thyroglobulin is an established tumour marker for monitoring papillary thyroid carcinoma, after total thyroidectomy and lymph node clearance. Persistently elevated thyroglobulin levels in the presence of a negative whole-body RAI scan are of great concern. In this patient with unusual liver metastases from papillary thyroid cancer, and relatively low thyroglobulin levels despite significant metastases, F-18 FDG PET/CT has once more proved its efficacy for further evaluation of patients with the TENIS syndrome.

References


