# Diffuse Sclerosing Variant of Papillary Thyroid Carcinoma : A Case Report

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We present a rare case of a 26-year old woman with diffuse sclerosing variant of papillary thyroid carcinoma. The patient was referred to our hospital with diffuse enlargement of the thyroid accompanied with palpable bilateral cervical lymph nodes. Ultrasonography showed a heterogeneous pattern with ill-defined hypoechoic areas in both thyroid lobes. There were multiple small punctate echogenic foci. Fine-needle aspiration cytology revealed typical signs of papillary carcinoma. The patient underwent a total thyroidectomy using a bilateral modified neck dissection. Pathological findings demonstrated diffuse involvement and continuous infiltration of the tumors to both thyroid lobes, lymph nodes and cervical soft tissue. Postoperatively, 100 mCi of <sup>131</sup>I was administrated to the small amount of residual thyroid tissue. The patient is free from recurrence one year after the operation. (Kitakanto Med J 2006; 56: 237~239)

Key Words : papillary thyroid carcinoma, diffuse sclerosing variant.

# Introduction

Different morphologic variants of papillary thyroid carcinoma have been described. Classical and mixed (papillary and follicular) variants are often seen and have a better prognosis. Diffuse sclerosing variant of papillary thyroid carcinoma (DSPC) is rare, has a more aggressive clinical course and a less favorable prognosis. In this report, we present a case of DSPC and discuss its clinico-pathological features.

### **Case Report**

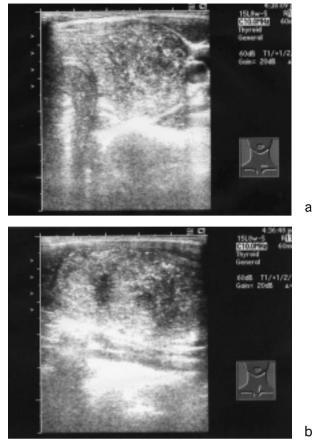
A 26-year-old woman with diffuse enlargement of the thyroid with no symptoms of pain or pressure was referred to our hospital. On physical examination, the thyroid gland was diffusely enlarged, and firm on palpation. Bilateral cervical lymph nodes were also palpable. The thyroid function was within normal limits on laboratory examination. The serum concentration of thyroglobulin (Tg) was 20.5ng/ml (reference range < 30), and anti- thyroglobulin antibody (anti -TgAb) was strongly positive. Ultrasonography (US) showed a heterogeneous pattern with ill-defined hypoechoic areas in both lobes of the thyroid. The margins were irregular and there were multiple small punctate echogenic foci (Fig. 1). Fine-needle aspiration cytology (FNAC) was performed in the hypoechoic areas of both lobes, and pathological examination showed typical signs of papillary carcinoma

A total thyroidectomy was performed using a bilateral modified neck dissection. The thyroid gland was diffuse and enlarged (Fig. 2a) and exhibited a solid cut surface with diffuse small granular and reticular calcifications without any distinct nodular mass (Fig. 2b). Pathological findings demonstrated diffuse involvement and continuous infiltration of tumors to both thyroid lobes, lymph nodes and cervical soft tissue. Heavily lymphocytic infiltration,

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**Fig. 1** Ultrasonogram showed ill-defined hypoechoic areas with a heterogeneous internal pattern and multiple small punctate echogenic foci in both lobes of the thyroid (a : a cross section, b : a vertical section).

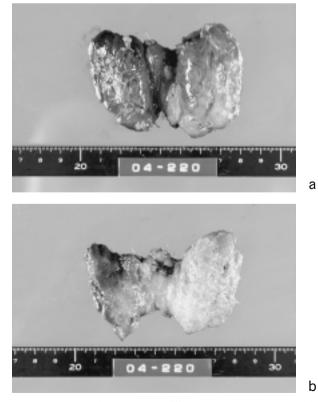


Fig. 2 The thyroid gland was diffusely enlarged (a) and exhibited a solid cut surface with diffuse small granular and reticular calcifications without distinct nodular mass.

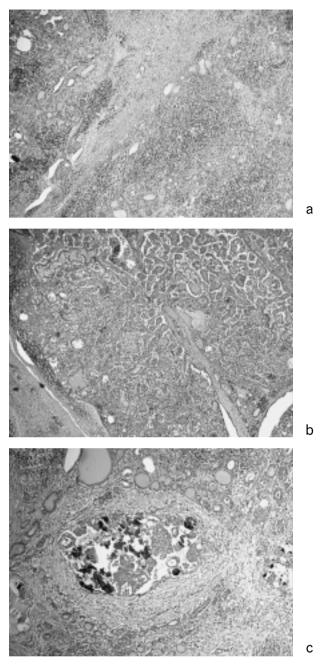


Fig. 3 Microscopic findings demonstrated diffuse sclerosing variant of papillary thyroid carcinoma. Lymphocytic infiltration, dense sclerosis (a,  $\times$  50), widespread papillary growth (b,  $\times$  50), psammoma bodies (c,  $\times$  100) and squamous metaplasia, were present.

dense sclerosis (Fig. 3a), widespread papillary growth (Fig. 3b), psammoma bodies (Fig. 3c) and squamous metaplasia were present. According to these findings, the case was diagnosed as a diffuse sclerosing variant of papillary thyroid carcinoma.

A <sup>131</sup>I whole body scan postoperatively showed uptake in the bed of the thyroid. 100 mCi of <sup>131</sup>I was administrated to the small residual thyroid tissue. The patient is free from recurrence one year after the operation.

# Discussion

The diffuse sclerosing variant (DSPC) is rare and comprises only 0.75-5.3% of all papillary thyroid carcinoma (PTC).<sup>1-3</sup> DSPC has prevalence in women similar to conventional papillary carcinoma, but the median age of occurrence is younger than for any other form.<sup>3,4</sup>

The preoperative diagnostic evaluation of patients with DSPC is problematic. Because DSPC shows a silent or painful enlargement of the thyroid gland, it has been reported that patients have been mistakenly diagnosed as having Hashimoto's thyroiditis or post -partum painful thyroiditis.<sup>2,4</sup> Therefore, the diagnosis is often delayed and this leads to a delay for surgical treatment. The functional status of the thyroid varies. FNAC is highly accurate in demonstrating a malignant pattern, suggestive of PTC, however, it may be inefficient in predicting pathologic variants of PTC such as DSPC.<sup>5,6</sup> In 1985, Vickery et al<sup>1</sup> proposed that a morphologic criteria is needed to make an accirate diagnosis of DSPC as an uncommon distinctive pathologic variant of PTC. Morphologic features of DSPC, such as sclerosis, psammoma bodies, patchy lymphocytic infiltration and squamous metaplasia are considered as anomalies within the spectrum of PTC. DSPC has a tendency to involve both of the thyroid lobes and lymphatic invasion. This is the reason for a high prevalence of regional lymph node metastases at presentation. Our case had all of these morphologic criteria.

DSPC is considered to have a worse prognosis than the classical PTC because of the delay in presentation, the higher incidence of lymph node involvement and its distant metastases. Some authors have reported that metastatic lymph nodes are present in 60-100% of cases.<sup>2-4</sup> Among the 62 cases reviewed by Chow et al<sup>3</sup>, distant metastases were seen in 17 patients (28%). Some reports, however, demonstrated that patients with DSPC have a similar prognosis as classical PTC.<sup>7-9</sup> It is difficult to draw any conclusions about the prognostic significance of this variant. In our case, we were unable to predict any prognostic implication of this rare variant since we had only a single case. Additional studies including a larger sampling with extended follow up would help to answer the queries on management.

Widespread intra-thyroidal growth, the presence of lymph node metastases, and the high incidence of distant metastases, justify a total thyroidectomy with bilateral modified neck dissection, followed by treatment with radioactive iodine to ablate the metastatic lesions. Tg is a sensitive and specific serum marker for detecting relapses in follow-up after surgery of PTC, however, its effectiveness is limited by the presence of anti-TgAb with may interfere with Tg assays. The high incidence of anti-TgAb in DSPC might render Tg less useful as a tumor marker after radioactive iodine ablation.<sup>3,7,11</sup> Thus, long-term follow up using a <sup>131</sup>I whole body scan is necessary in patients with DSPC.

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