**CASE REPORT**

**Diffuse Sclerosing Variant of Papillary Thyroid Carcinoma**

*Jyothi C.R.,* López A.C.,*Ejeckam G.C. and **Hussain A.A.*

Departments*Laboratory Medicine & Pathology and **Surgery
Hamad Medical Corporation, Doha, Qatar

**Introduction:**

Diffuse sclerosing variant of Papillary Thyroid Carcinoma (DSPC) is an unusual type of Papillary Carcinoma that occurs in young patients(1). Clinically it can be mistaken for Hashimoto’s Thyroiditis as it often presents as a diffuse firm thyroid swelling(2,3). It is important to recognize this variant as the patients invariably have lymph node metastasis at the time of diagnosis(3). Contrary to the previous reports of a poor prognosis of this variant when compared to the classical papillary carcinoma, recent reports indicate a good prognosis(2,3). We report a case of DSPC occurring in a young boy.

**Key Words:** Diffuse sclerosing variant, papillary thyroid carcinoma

**Case Report:**

An eleven-year-old boy presented with an anterior neck swelling to the general surgery department of Hamad Medical Corporation. The swelling was situated mainly on the left side of the thyroid, moving with deglutition, firm in consistency and measuring 2 x 3 cm in size. Complete Blood Count (CBC) was within normal limits. Free T4 was 9 pmol/L, TSH 18.95 mIU/L (0.30-5.60). Tc-99 perthyroid scintigraphy showed cold nodules in the left lobe of the thyroid gland.

Fine Needle Aspiration Cytology of the left lobe of the thyroid showed a few cells with nuclear grooves, intranuclear cytoplasmic inclusion, psammoma bodies, clusters of squamoid cells, few follicular epithelial cells and scanty colloid. A diagnosis of papillary thyroid carcinoma was made.

The left thyroid lobe was received for frozen section. It measured 3.5 x 3 x 1.5 cm weighed 8 gms and was reddish and slightly nodular. The cut surface was homogenous and pink. Light microscope examination showed diffuse stromal sclerosis and diffuse infiltration of the thyroid by lymphocytes forming follicles with germinal centers and islands of squamoid cells in vascular and lymphatic spaces but the striking feature was the presence of numerous psammoma bodies (Figure 1).

![Figure 1: Photomicrograph H&E x 10 showing abundant Psammoma bodies.](image-url)

The patient underwent total thyroidectomy with bilateral functional neck dissection. The right lobe weighed 6 grams and measured 3 x 2 x 1.5 cm. The surface was nodular and the cut surface showed a uniform brown appearance with faint nodule formation. Microscopically it was similar to the lesion in the left lobe. There were numerous clusters of papillary carcinoma cells with squamoid differentiation (Figure 2). The extra-capsular soft tissue was also infiltrated by tumor. One of the nine cervical lymph nodes on the right and 18 of the 26 lymph nodes on the left side contained metastases from the neoplasm.

Immunohistochemistry showed the tumor cells to be positive for thyroglobulin. The squamoid cells were positive for cytokeratin (Figure 3). The neoplastic cells were negative for calcitonin. S-100 positive Langerhan’s cells were scattered within the tumor. A mixed population of B and T lymphocytes was noted in the inflammatory infiltrate. The vascular spaces were lined by CD 34 positive cells.

**Discussion:**

DSPC was initially recognized as an aggressive variant of papillary carcinoma in 1985 by Vickery et al(5). Subsequently several reports of this variant have been published(1-3). DSPC...
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Figure 2: Photomicrograph H&E x 40 showing cells with squamoid differentiation in endothelial lined spaces.

Figure 3: Photomicrograph x 10 showing neoplastic cells positively stained for cytokeratin by Immunohistochemistry.

accounts for 1.6 to 3.4% of papillary thyroid carcinomas. Our case is the first of 108 papillary thyroid carcinomas examined in the past 15 years in the histopathology section of Hamad Medical Corporation.

DSPC has to be recognized and treated radically as most of the patients have lymph node metastasis at the time of diagnosis. It can be confused clinically with thyroiditis as the patients present with firm thyroid swelling and elevation of thyroid antibodies in some cases. This often results in the delay of treatment. However when the patient is young and abundant psammoma bodies are detected by ultrasonography or soft tissue X-ray of the neck, DSPC has to be considered in the differential diagnosis. Cytopathology can be diagnostic when a combination of clinico-pathological features of numerous psammoma bodies, lymphocytes, squamous metaplasia and absence of stringy colloid are noted with otherwise typical cytological features of papillary carcinoma in FNAB smears obtained from a diffusely nodular firm thyroid enlargement. Only a small number of cases of the usual type of papillary carcinoma show psammoma bodies in FNAB smear. Psammoma bodies can also be seen rarely in benign thyroid lesions like lymphocytic thyroiditis. Only 50% cases of classical papillary carcinoma show psammoma bodies in histopathology. The immunohistochemical findings in our case were consistent with literature reports.

The key morphological feature, wide spread lymphatic permeation of the thyroid, is clearly related to the incidence of nodal spread and post-therapy disease persistence. The diffuse lymphatic permeation also causes the diffuse and sometimes painful enlargement of the thyroid mimicking a thyroiditis clinically. The presence of elevated levels of antibodies in some cases complicates the issue. The lymphocytic infiltration and the raised antibody titers could be the result of an antigenic response to the tumor. As for the view that DSPC develops in a Hashimoto’s thyroiditis, Hurthle cells, which are a feature of Hashimoto’s thyroiditis, are absent in DSPC.

Prognosis has been reported as variable in different studies. Probably the youth of the patients contributes to a more favorable prognosis in spite of the lymph node metastasis. Prognostic significance of the various morphological features of DSPC is still unsettled.

There is still reluctance in diagnosing this variant of papillary carcinoma in cytology and frozen section as the clinical features mimic thyroiditis and the patients are usually young. Previous reports indicated a female predilection for DSPC but this condition is being recognized increasingly in children and young adults. A combination of numerous psammoma bodies, diffuse sclerosis, tumor cells in cleft-like lymphatic spaces, squamous metaplasia and intense lymphocytic infiltration in a young patient with diffuse thyroid enlargement points to the diagnosis of DSPC as it is a once seen never forgotten lesion.

References: