

CASE REPORT

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An unusual presentation of diffuse sclerosing variant of papillary thyroid carcinoma

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Abstract

Background: The diffuse sclerosing variant of papillary thyroid carcinoma is a rare pathologic entity. It is characterized by a tendency for rapid growth and a higher incidence of cervical lymph node and distant metastases.

We report an unusual presentation of diffuse sclerosing variant of papillary thyroid carcinoma presented as a single thyroid mass of a small size.

Case presentation: We experienced a case of a diffuse sclerosing variant of papillary thyroid carcinoma in a 30-year-old woman. The preoperative findings on the cytology and ultrasound were suggestive of a malignant thyroid nodule. This case was confirmed by surgical excision. The management included surgery, radioiodine ablation, and suppressive levothyroxine therapy.

Conclusions: We would like to highlight that an early diagnosis associated with an appropriate management leads to a good prognosis.

Keywords: Diffuse sclerosing, Papillary carcinoma, Ultrasonography, Thyroid, Surgery

Background

Papillary thyroid carcinoma (PTC) is the most common type of thyroid carcinoma accounting for approximately 80% of all thyroid gland malignancies.

The diffuse sclerosing variant (DSV) is a rare subtype of papillary thyroid carcinoma [1]. It was first described in 1985 by Vickery et al. as an unusual form of PTC, for which they proposed the term “diffuse sclerosis variant” [2, 3]. According to the World Health Organization classification, DSV occurs in young female adults and less commonly in children [1, 2]. It is characterized by an aggressive clinical behavior with a higher frequency of cervical lymph nodes and distant metastasis [4]. Histologically, it presents as a usually diffuse involvement of one or both thyroid lobes, without the formation of a

dominant mass. In addition to classic nuclear features of PTC, extensive squamous metaplasia, numerous psammoma bodies, patchy to dense lymphocytic infiltrates, and small papillary structures within dilated lympho-vascular spaces and stromal fibrosis lead to the diagnosis of DSV [3, 4].

Herein we report an unusual presentation of diffuse sclerosing variant of papillary thyroid carcinoma presented as a single thyroid mass of a small size.

Case presentation

A 30-year-old female presented to our department with a 1-month history of a cervical mass. She had not experienced hoarseness, dysphagia, odynophagia, or breathing difficulty. Her past medical history was free from prior radiation therapy and a family history of thyroid diseases. She was clinically euthyroid. At physical examination, we found a single mass of the right thyroid lobe, small-sized (off 1 cm), without palpable cervical lymph nodes.

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Cervical ultrasound examination revealed an ill-defined hypo-echogenic mass of the thyroid gland right lobe, measuring 1.5 cm in diameter, with diffused micro-calcifications without extra capsular extension associated with an enlarged right cervical lymph node of 1 cm in sector IIa (Fig. 1).

Thyroid function tests were normal. Fine needle aspiration of this nodule indicated Bethesda IV anomalies which is associated with malignancy in 15–30% of case. The patient underwent a right lobectomy. Per-operatively, we noticed a mal limited indurated nodule of the right thyroid lobe with adherence and enlarged central lymph nodes. Due to the preoperative findings, a frozen section examination of the nodule was indicated to label the histological type of the thyroid malignancy. It confirmed the

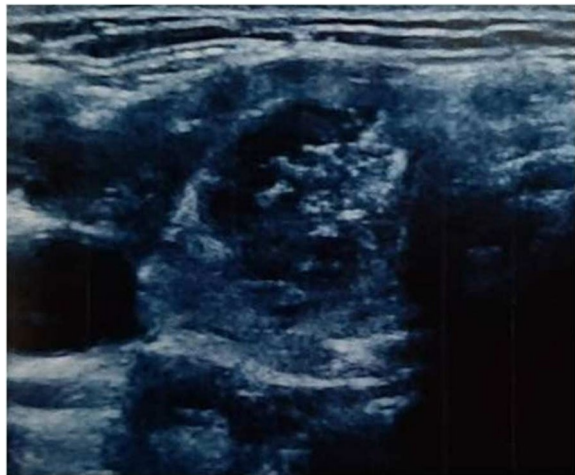


Fig. 1 Cervical ultrasound examination revealing an ill-defined hypo-echogenic mass of the thyroid gland right lobe, measuring 1.5 cm in diameter, with diffused micro-calcifications

diagnosis of papillary carcinoma; then, a total thyroidectomy, a bilateral central lymph node dissection, and a right lateral lymph node dissection were performed. The postoperative course was uneventful.

Macroscopic examination showed a white firm, ill-defined nodule, measuring 1.4 cm in greatest diameter with a rubbery cut surface. The surgical specimens were included and examined histologically. There was a diffuse involvement of the thyroid by a widely invasive tumor associated with dense fibrous stroma. The tumor had a papillary pattern with solid cribriform areas and extensive squamous metaplasia (Fig. 2). Tumor cells showed a characteristic nuclear atypia of papillary thyroid carcinoma. Numerous psammoma bodies were observed in addition to a marked lymphocytic infiltration with the formation of germinal centers within the fibrous stroma. Vascular invasion by tumor cells was seen (Figs. 3 and 4). Microscopic examination revealed multiple metastatic cervical lymph nodes, either. The final diagnosis was the DSV of PTC. She underwent radio ablative therapy followed by suppressive levothyroxine therapy. There was no residual uptake on subsequent 123-Iodine isotope scanning. There was no evidence of recurrence after 5 years follow-up.

Discussion

DSV is an uncommon variant of PTC accounting for 1.8% of all PTCs [1]. Compared with classical PTC, it is characterized by younger patient age and larger mean size of the tumor with a higher tendency for cervical lymph node metastasis and distant metastasis [5].

Definitive diagnosis is made on the basis of the presence of several pathological features including a diffuse firm enlargement of the thyroid gland with scattered islands of papillary carcinoma, extensive lymphatic permeation, and lymphocytic infiltration, extensive

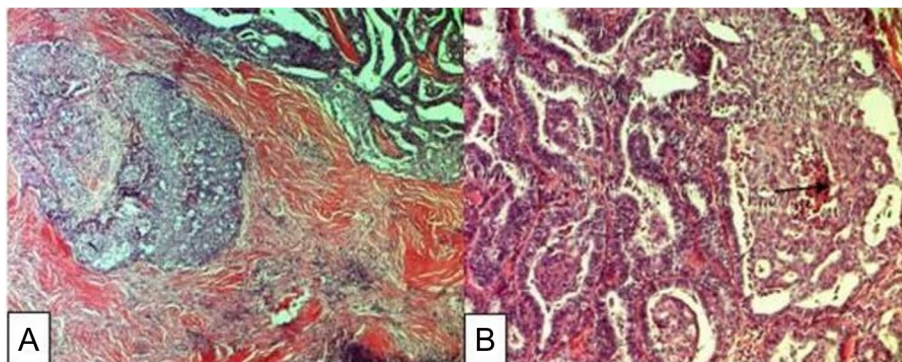


Fig. 2 High magnification of tumor pattern. **A** Tumor exhibit papillary pattern with solid and cribriform areas (HE \times 50). **B** Extensive squamous metaplasia (indicated by an arrow) within cribriform and papillary structures (HE \times 100)

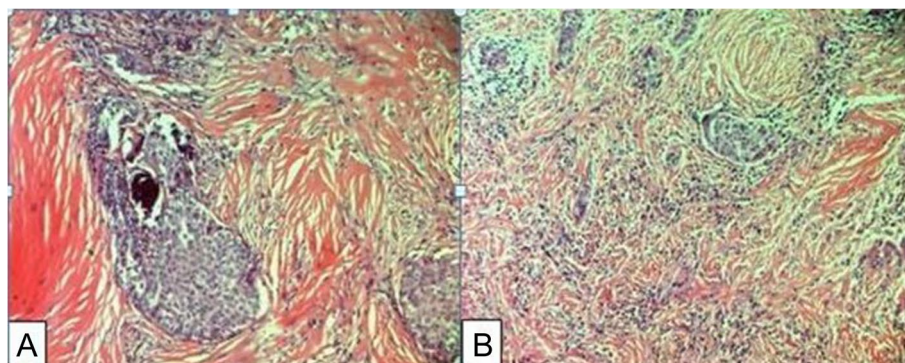


Fig. 3 Microscopic features of tumor stroma. **A** Diffuse severe hyaline fibrosis (HE \times 50). **B** Marked dense inflammatory infiltrate surrounding tumor sheets (HE \times 50)

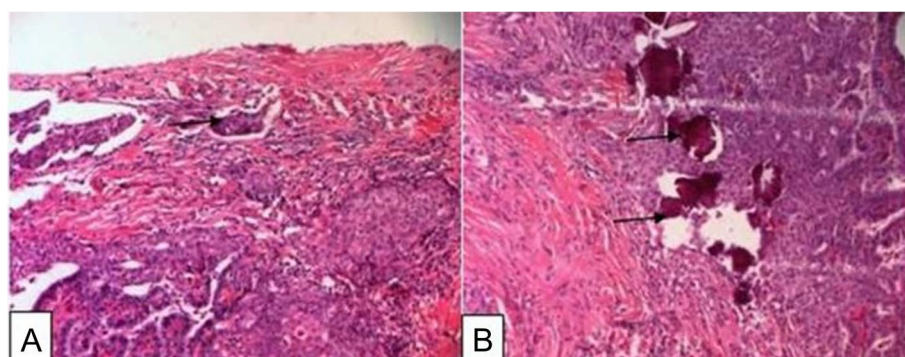


Fig. 4 Other tumor particularities. **A** Presence of lymphatic emboli within tumor (HE \times 50). **B** Numerous psammomatous bodies (indicated by an arrow) are observed (HE \times 50)

squamous metaplasia, a large number of psammoma bodies, and a prominent fibrosis. These histological features are essential in distinguishing PTC from benign pathologies such as Riedel's and Hashimoto's thyroiditis [6, 7]. The preoperative diagnosis of DSV remains difficult, and it can be diagnosed preoperatively based on a combination of typical imaging findings and careful cytological examination [8, 9].

Cervical ultrasonography (US) is generally considered as the most accurate imaging modality for the characterization of the thyroid gland pathologies. The US features of DSV are heterogeneous echotexture, solid composition, ill-defined margins, and scattered microcalcifications [6, 8, 10].

In our case, cervical ultrasound examination revealed an ill-defined mass of the thyroid gland right lobe with diffused micro-calcifications associated.

However, the diffuse nature of DSV often mimics chronic thyroiditis particularly in the absence of a focal solid mass [6]. The preoperative neck US may help detect intrathyroidal extension and lymph node metastases in

DSV patients [6, 9]. The mean incidence of distant metastasis reported in DSV is 14.9% [5, 8], and the most common site is the lung. Early studies suggested that DSV had a poorer prognosis than classical PTC [10, 11]. It had also been reported that eradication required a more aggressive therapeutic approach.

In our case, a total thyroidectomy, a bilateral central lymph node dissection, and a right lateral lymph node dissection were performed. More recent studies revealed that DSV has a similar prognosis to patients with classical PTC [1, 8], and then the treatment should be the same radical surgery including total thyroidectomy and extensive lymph node dissection, radio-iodine ablation, and suppressive hormone therapy [12, 13].

Conclusion

DSV is one of the most aggressive forms of papillary thyroid cancer. Therefore, it is important to diagnose this as quickly as possible to ensure appropriate treatment. The management includes total thyroidectomy associated

with lymph node dissection and postoperative radioiodine therapy. A close follow-up is necessary.

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Authors' contributions

Authors' Contributions: MG: data analysis and interpretation and final approval of the version to be published. JH: conception of the work, data collection and drafting the article. LC: data collection. NA: Analysis and interpretation of the data and drafting of the article. ME and MA: data collection. MB, KW and MA: critical revision of the article. All authors read and approved the final manuscript.

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Consent for publication

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Competing interests

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