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Case Report

A Rare Papillary Thyroid Carcinoma Variant; How Aggressive We Should Be? Case Report

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Abstract

Cribriform Morular Variant (CMV) of Papillary Thyroid Carcinoma (PTC) is a rare type of thyroid papillary neoplasm. It is more prevalent in patients with Familial Adenomatous Polyposis syndrome (FAP), however sporadic CMV-PTC has been described. Due to a low prevalence of this condition, recent consensus guidelines do not state a specific therapeutic approach for this variant. We present a 20-year-old woman with PTC-CMV treated with a total thyroidectomy alone, with no evidence of recurrence at the one year follow up. More aggressive treatments that include lymph node dissection and adjuvant radio ablation have been described. This case shows that patients with this variant can be treated with a more conservative approach similar to that of variants that are at low risk for recurrence.

Keywords: Thyroid cancer; Cribriform modular variant; Management

Introduction

Cribriform Morular Variant (CMV) of Papillary Thyroid Carcinoma (PTC) is a rare type of thyroid papillary neoplasm, which is more prevalent in patients with Familial Adenomatous Polyposis syndrome (FAP), however CMV-PTC without FAP has been described [1]. Although the relation between both conditions is strong, the incidence of PTC has remained low (0.4-1.3%) [2,3]. CMV-PTC is characterized by its cribriform pattern with solid areas and spindle cell component [4]. Long term management is unclear. The latest American Thyroid Association [ATA] guidelines for differentiated thyroid cancer do not give any recommendation regarding therapeutic approaches for this specific variant [5]. This case report describes a 20-year-old woman with PTC-CMV treated as a low risk carcinoma.

Case Presentation

This is the case of a 20-year-old woman with past medical history of FAP. She came to our endocrinology clinics due to discomfort upon swallowing liquids and solids and sensation of neck fullness since 3 months ago. She had been diagnosed with FAP 6 months before our evaluation. Upon initial encounter, the patient denied any symptoms suggestive of thyroid dysfunction. Physical examination was remarkable for a mildly enlarged thyroid gland and a palpable non-tender nodule in the right side of the neck. Thyroid function tests were normal. Thyroid ultrasound revealed a solid hypoechoic nodule on the right thyroid lobe measuring 1.2cm x 1.0cm x 0.9 cm without calcifications, irregular borders, extra thyroidal extension. A Fine Needle Aspiration Biopsy (FNAB) was performed and showed a Papillary Thyroid Carcinoma with cytoplasmic and nuclear positive stain to b-catenin, suggestive of CMV-PTC (Figure A,B,C,D,E). Due to these findings and FAP history, a total thyroidectomy was performed. Surgical pathology report showed a well-delimited, unifocal, 1.5cm x 1.0cm x 0.9cm CMV-PTC, without angiolymphatic or perineural invasion. Postoperatively, the patient was treated with levothyroxine suppression with a target TSH of 0.5-2.0 ng/mL. Due to prior reports that support less invasive treatment, no lymph node dissection or radioactive iodine treatment was given. One year after her surgery, the patient remains without evidence of loco regional recurrence.

Discussion

Familial adenomatous polyposis syndrome is an autosomal

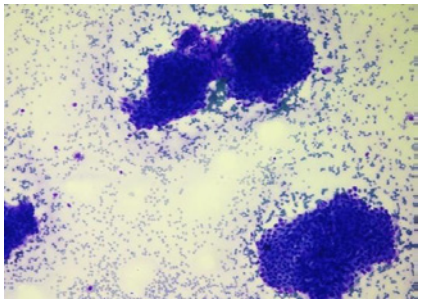

Figure 1A: Giemsa stain x 40, showing and hyper cellular aspirate composed of follicular cells with squamous morula, with absence of intervening fibro vascular stroma and colloid.

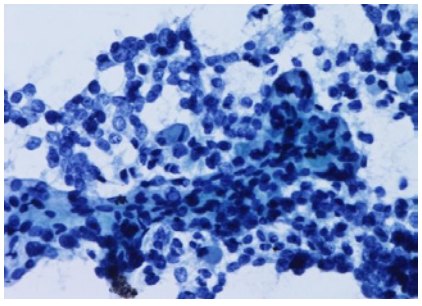

Figure 1B: Tissue fragment of follicular cells forming complex branching papillary structure. The upper part the cribriform pattern is observed with punch out spaces devoid of colloid. Fusiform, plasmacytoid and cuboidal follicular cells are present (Papanicolaou stain x 400).

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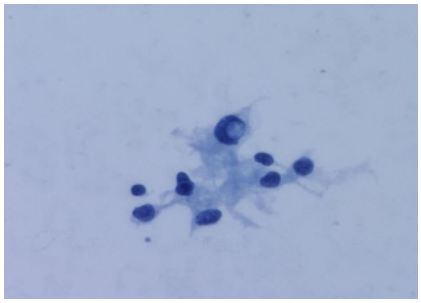

Figure 1C: Papanicolaou stain showing fusiform and cuboidal follicular cells. The follicular cells show variation in size; an intranuclear pseudo inclusion is observed in the upper middle cell.

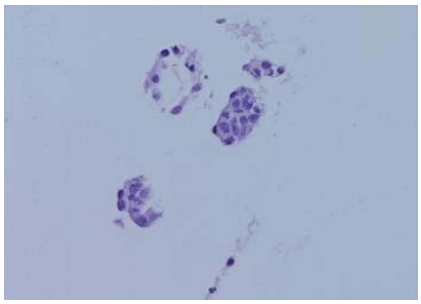

Figure 1D: Hematoxylin and eosin stain (x100) on cell block showing spindle to cuboidal follicular cells forming morules.

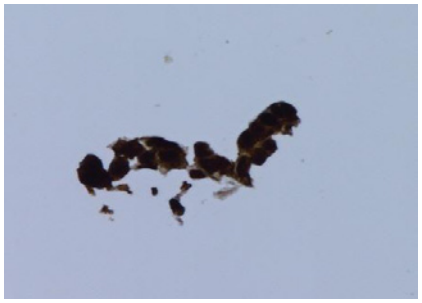

Figure 1E: The cell block was used for b-catenin staining showing a strong nuclear staining pattern by immunohistochemistry (x400).

dominant condition caused by a mutation in the Adenomatous Polyposis Coli gene (APC). It can present with several extra-intestinal manifestations in the thyroid, pancreas, liver and central nervous system among others. CMV-PTC is an uncommon variant and is highly associated with FAP syndrome [1]. It was first described since 1949 by Crail et al. [6]. Since then, roughly 200 cases have been reported in the literature. In contrast to other PTC variants, data is scarce regarding therapeutic approaches.

Surgery is the mainstay of therapy in thyroid carcinoma. The approach depends the type of PTC and extent of disease. Recent ATA guidelines recommend that tutors from 1cm to 4cm without extra thyroidal extension or lymph node involvement can be treated with lobectomy or total thyroidectomy [5]. The use of radioactive iodine ablation is not recommended since it does not add benefit in terms of survival or recurrence. Historically radioactive iodine ablations routinely used as adjuvant therapy to surgery in patients with CMV-PTC [7,8]. However most recent reports suggest a more conservative approach is equally effective [9-11]. Recent clinical practice guidelines do not recommend prophylactic lymph node dissection for tumors with low aggressive features since do not increase long term survival. However this approach have been used is some case series of CMV-PTC with great results [12,13] (Table 1).

Our case shows the usefulness of molecular markers to confirm the diagnosis and therefore help guide management. It also demonstrates that less invasive procedures may be equally curative for these patients while avoiding the potential risks of radioactive iodine or lymph node dissection. The scarcity of available data has limited the ability to come up with specific guidelines. However, our report aims to contribute to current literature and support the use of more conservative approaches in patients with CMV-PTC. Hopefully, it will also help in bringing uniformity to treatment recommendations.

Although some clinical evidence exists about the management for CMV-PTC, no standard of care has been defined. This is mainly because the evidence is based on small studies and case series. This case highlights an important unresolved issue regarding the management approach for this uncommon variant of PTC. We recommend the use of molecular markers in the histopathology diagnosis. lobectomy

Table 1: This table shows the difference case series that have compared the different treatments for CMV-PTC.

Author	Patients [n]	Surgery [total/subtotal thyroidectomy]	Radiotherapy	Complications	Recurrence	Neck Dissection
Jagidar et al. [7]	3	Total = 3	3	none	None	None*
Cameselle Teijeiro et al. [8]	4	Total = 4 Subtotal= 1	2	none	None	None*
Tomoda et al. [9]	7	Total =7	0	none	None	None*
Miyauchi et al. [10]	18	N/A	0	none	None	None*
Cruz Correa et al. [11]	4	Total =2 Subtotal =2	0	none	None	None*
Nam Es et al. [12]	3	Total =3	0	none	None	Central Neck Dissection = 3
Miya et al. [13]	32	Total = 21 SubTotal = 11	0	none	2 (Subtotal thyroidectomy)	Central Neck Dissection = 32

*N/A: Unavailable, all the patients underwent surgery, surgery approach not describe
*None: Not mention in the study
Total: Total thyroidectomy
Subtotal: Subtotal thyroidectomy
None: No complications or recurrence occurred
Subtotal thyroidectomy**: Recurrence occurred in 2 patients within the subtotal thyroidectomy group.

approach for the sporadic cases and total thyroidectomy for the FAP associated cases. In our opinion lymph node dissection and radioactive iodine ablation as adjuvant therapies should not be performed given that it may cause an increase in intraoperative as well as postoperative complications without a significant improvement in patient prognosis.

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