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Primary Squamous Cell Carcinoma of the Thyroid: A Case Report and Review of the Literature

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Abstract **Case Report**

Primary squamous cell carcinoma of the thyroid or Herrenschmidt tumor is an extremely rare entity that accounts for less than 1% of all types of thyroid neoplasms. Moreover, it has an aggressive behavior and unfavorable prognosis that resembles anaplastic thyroid carcinoma. The management of this tumor is mainly based on radical surgery. However, various other therapeutic approaches have been used, either exclusively or in combination with surgery. We report the case of a 60 years female patient, who presented with a cervical mass, that has been evolving for 2 months, causing intermittent dysphonia and dyspnea. Computed Tomography of the neck showed a goiter involving the isthmus and right lobe of the thyroid, infiltrating the aerodigestive tract, filling the thoracic inlet, and associated with cervical lymph nodes. Biochemical tests including serum thyroid-stimulating hormone (TSH), T4, thyroglobulin, and calcitonin were quite normal. An Echoguided thyroid biopsy was performed, which the histological and immunohistochemical study revealed a squamous cell carcinoma of the thyroid. The metastatic workup was negative. The case was discussed in a multidisciplinary deliberation meeting that concluded that the tumor is unresectable, and the patient was proposed for palliative radiotherapy and supportive treatment. The post-therapeutic follow-up was marked by size progression of the neck mass and persistent dyspnea. The patient died of airway compromise after 4 months.

Keywords: Primary, Squamous cell carcinoma of the thyroid, PSCCT, Herrenschmidt tumor, Radiotherapy.

INTRODUCTION

Primary squamous cell carcinoma of the thyroid (PSCCT) or Herrenschmidt tumor was originally described by von Karst in 1858 [1]. It is an extremely rare entity that accounts for between 0,7 to 3,4 % of all types of thyroid neoplasms [2]. Moreover, it has an aggressive behavior and unfavorable prognosis that resembles anaplastic thyroid carcinoma. This tumor is usually found locally advanced at the time of diagnosis, with infiltration of the aerodigestive tract and neck vessels. The management of PSCCT is mainly based on radical surgery. However, various other therapeutic approaches, including radiotherapy and chemotherapy, have been used, either exclusively or in combination with surgery [3].

CASE PRESENTATION

We report the case of a 60 years female patient with no medical history. Who presented a painless swelling in the right anterior cervical region that has been rapidly increasing in size for 2 months, causing intermittent dysphonia, dyspnea, and episodes of aspiration. These symptoms were evolving in the context of preservation of the general condition, with no signs of dythyroidism.

On clinical examination, the patient had right-sided sub-hyoid cervical hard mass, measuring about 8cm, painless on palpation, and fixed to deep structures, with no inflammatory surface or fistula. Moreover, she had spontaneous distension of the right external jugular vein and infracentimetric right sub-maxillary lymph nodes. The rest of the examination was unremarkable.

Cervical Ultrasonography revealed a significant thyroid hypertrophy, with an oval nodule in the right lobe, containing a macrocalcification, and measuring 3,90 cm * 3,60 cm, graded ti-rads 5 . The left lobe contains 3 ovular nodules that are graded ti-rads 3 and a right cervical lymph node measuring 2,84 cm. Computed Tomography (CT) of the neck showed a goiter that measures 44x44x55mm involving the isthmus and right lobe of the thyroid, with a

macrocalcification. This tumor fills the thoracic inlet, infiltrates the laryngeal structures, and is also associated with middle and lower right jugular lymph nodes (Figure 1, Figure 2).

Biochemical tests including, serum thyroid-stimulating hormone (TSH), T4, thyroglobulin, calcitonin, and calcium were quite normal.

An Echoguided thyroid biopsy was performed. Which the histological study revealed a poorly differentiated squamous cell carcinoma (SCC), and on immunohistochemistry (IHC), the tumor cells stain positive for Cytokeratin7 (CK7), CK19, P63, with overexpression of Ki-67 that was about 60%, but they stain negative for CK19, CK20.

In search of a primarily contiguous spread of a squamous cell carcinoma (SCC) from the upper aerodigestive tract, or metastatic disease that has spread from its original site, Other tests were performed a full-body CT scan showed, increase in the size of the same tumoral process measuring 87*61*79 mm, which invades the larynx, and responsible for compression of the aerodigestive tract. Moreover, it was associated with right internal jugular vein thrombosis, right jugular, and supraclavicular lymph nodes. Pan-endoscopy, esophagoscopy, and bone scintigraphy did not reveal any abnormalities.

This case was discussed in a multidisciplinary deliberation meeting, which concluded that the tumor is unresectable, and even tracheostomy could be unfeasible due to the thoracic inlet filling the patient was proposed for palliative external beam radiation therapy (EBRT), anticoagulant therapy, and supportive treatment. She received radiotherapy on the thyroid gland, with a total dose of 30 Grays (Gy), distributed in 10 fractions, over a period of 2 weeks. The post-therapeutic follow-up was marked by size progression of the neck mass and persistence of dyspnea the patient died of airway compromise after 4 months.

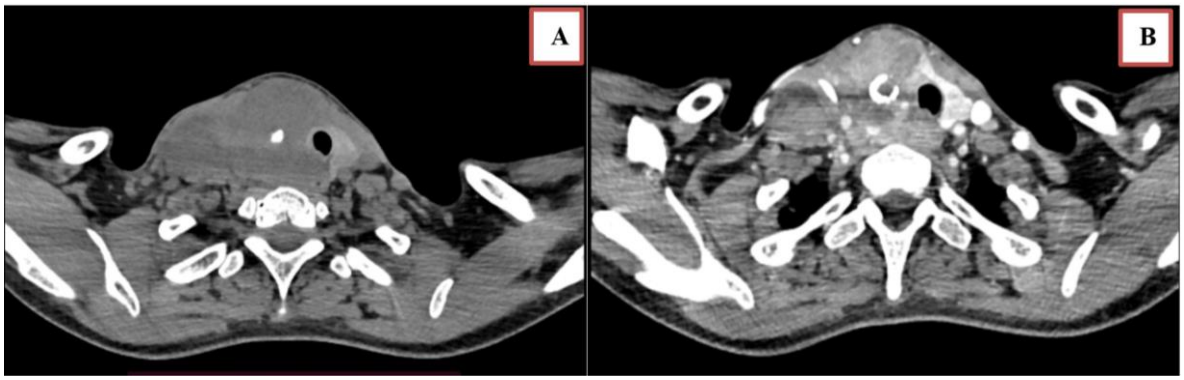

Figure 3: Cervical CT scan in axial section showing the tumoral process of the thyroid with central macrocalcification and infiltration of the aerodigestive tract. Associated to a right jugular lymph node. (A: without injection of contrast, B: with contrast injection)

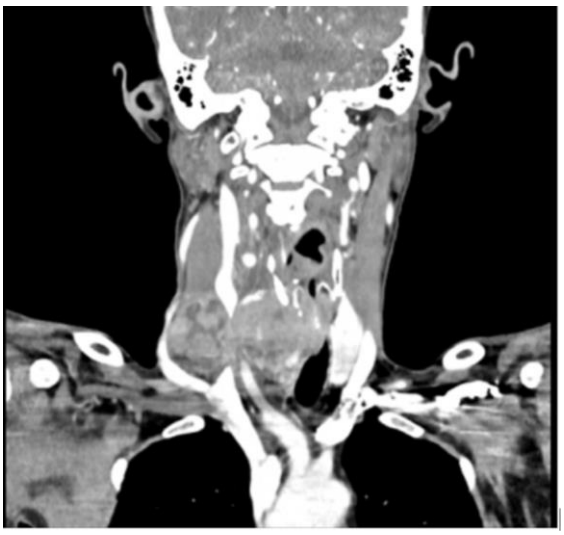

Figure 4: Cervical CT scan with contrast injection in coronal section showing the tumoral process of the thyroid infiltrating the aerodigestive tract, and a right lower jugular lymph node

DISCUSSION

Primary squamous cell carcinoma of the thyroid (PSCCT) is an extremely rare malignancy accounting for less than 1% of all thyroid cancers [4]. It is a very aggressive entity that resembles anaplastic thyroid carcinoma, which usually manifests as a locally advanced disease with an unfavorable prognosis [3, 5]. It usually appears between the fifth to sixth decades of life, with the mean age of diagnosis at 68 years. Moreover, Women are more likely to be affected than men [6, 7].

The pathogenesis of primary SCC within the thyroid remains controversial since The squamous epithelium is absent in the thyroid gland [3]. Though, three theories have been proposed. The first one is the embryonic nest theory that was proposed by Goldberg and Harvey, who suggested that Squamous cells are generated from embryonic vestiges such as the thyroglossal duct, thymic epithelium and ultimobranchial body [8]. The second theory suggests

that the origin of squamous cells emerges due to squamous metaplasia, which can occur as a result of inflammation or Hashimoto's thyroiditis [9]. The third hypothesis is the dedifferentiation theory which suggests that SSC originates from potential dedifferentiation from other types of existing thyroid carcinomas, such as papillary, anaplastic, or medullary carcinoma [10].

The diagnosis of PSCCT is very challenging. It can only be confirmed after eliminating an SCC from an adjacent structure invading the thyroid, such as pharyngoesophageal and laryngotracheal, or from a metastasis that originates from other sites [4]. In that sense, the diagnosis is based on a combination of clinical, endoscopic, pathological, and radiological features [3].

Clinically, patients routinely present with rapidly growing neck mass with frequent expansions into surrounding structures and compression of the upper aerodigestive tract, typically causing neck pain, dysphagia, dyspnea, and dysphonia. Furthermore, It is usually accompanied with an invasion of cervical lymph nodes [3].

Cervical ultrasound is a simple and cost-effective examination that is usually used in the first place during the diagnosis process. It helps to suggest the malignant nature of the tumor based on its form and echogenicity; moreover, it guides the biopsy [11]. Computed tomography (CT) and magnetic resonance imaging (MRI) of the neck are very valuable. They allow to distinguish the thyroid mass from other neck masses and permit an accurate assessment of details, including calcification, necrosis, margins, and extra glandular extension. Also, they help analyze the adjacent structures for invasion or compression [3]. However, there are no specific radiological features of thyroid SCC, and they are similar to those seen in other thyroid malignancies. Before retaining the diagnosis of PSCC, the exclusion of a primary source for secondary Squamous Cell Carcinoma of the thyroid (SSCCT) is vital .this is based on phonendoscope and CT scans of the chest, abdomen, and pelvis.

From an anatomopathological perspective, Thyroid squamous cell carcinoma should be formed exclusively of squamous differentiated tumor cells [12]. Despite this, poorly differentiated carcinoma represents nearly one-third of all primary thyroid squamous cell carcinomas [13]. Immunohistochemistry may be very helpful in differentiating primary from secondary squamous cell carcinoma or coexisting papillary carcinoma of the thyroid [14]. The PSCC stains positive for cytokeratin, with a specific pattern that is positive for CK7 and CK19, but negative for CK1, CK4, CK10, CK13, and CK20 [15]. It also stains positive for Thyroid transcription factor 1 (TTF1) and thyroglobulin [6]. Additionally, overexpression of oncoprotein p53 and a high Ki-67 proliferation index may be observed [16]. These markers can help make the diagnosis and establish the prognosis, as they were reported as indicators of poor prognosis and higher risk of local recurrence [16, 17].

Various treatment regimens, including surgery, radiotherapy, and chemotherapy, have been used in managing PSCCT, either exclusively or in combination. Surgery represents the main treatment of PSCCT, which consists of total thyroidectomy and complete surgical resection of the tumor along with infiltrated adjacent structures and regional lymph nodes resection[3, 4, 17]. The surgery must be as complete as possible in order to ensure negative surgical margins since it has been reported to be the only factor correlated with a favorable prognosis [17, 18].

Some authors used different chemotherapy protocols as adjuvant treatment; nevertheless, the results were disappointing, as they reported an unconvincing or little benefit of chemotherapy [3, 19, 20]. Although PSCC has been documented to be relatively radio resistant , radiotherapy have been used in combination with chemotherapy as post operative adjuvant therapy [3], or alone for a palliative aim [21]. Tunio *et al*., recommended adjuvant radiotherapy (50-60 Gy) with or without chemotherapy after complete surgery , according to his review this regimen seems to have achieved better outcome and better survival rates [21], though he reported that radiotherapy and chemotherapy alone are ineffective in the other hand, Soufi *et al*., have reported that post operative radiotherapy is ineffective and it can be even dangerous, as it may causes inflammation and oedema leading more likely to accelerate respiratory distress [22]. Syed *et al*., concluded in his review that all the reported combination of treatment regimens including surgery , radiotherapy , and chemotherapy have no significant impact in enhancing outcome or extending survival rate, and he recommended instead a palliative approach which consists of tracheostomy, gastrostomy, supportive treatment and in some cases palliative surgery to relieve severe airway obstruction [3].

CONCLUSION

Primary squamous cell carcinoma of the thyroid is an extremely rare and very aggressive tumor. Besides surgery that represent the main treatment, radiotherapy and chemotherapy can also be used, with better outcomes when combined as an adjuvant to surgery. However, and Regardless of any treatment, the PSCCT appears to be associated with pejorative prognosis.

Abbreviations

SCC; Squamous cell carcinoma \ PSCCT ; Primary squamous cell carcinoma of the thyroid \ SSCCT ; Secondary Squamous Cell Carcinoma of the thyroid \ TSH ; Thyroid stimulating hormone \ CT ; Computed

Tomography \ MRI ;Magnetic resonance imaging \ IHC; Immunohistochemistry \ CK ; Cytokeratin \ EBRT; External beam radiation therapy \ Gy; Gray.

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