CASE REPORT

Giant Anaplastic Thyroid Carcinoma of Sarcomatoid Type with Obstructive Symptoms: A Case Report

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Abstract

Aim: To report the surgical approach of a giant thyroid tumor despite the histological type.

Background: Although salt for human consumption contains iodine; in some populations far from civilization stone salt (noniodized salt) is still used, the latter plus a lack of medical attention, leads to the development of goiter with excessive growth and possible malignant transformation.

Case description: We report the case of surgical treatment of a giant tumor of the thyroid gland in a 55-year-old female patient who was admitted to the emergency department with respiratory distress and difficulty swallowing food. She underwent an emergency total thyroidectomy due to airway obstruction. During the surgical treatment, adhesions of the tumor to the cervical esophagus and to the aorta artery in its projection towards the mediastinal region were found.

Conclusion: Anaplastic thyroid carcinoma is the rarest and most dangerous of all types of thyroid cancers, so extreme caution should be exercised in the treatment choice according to the patient's case.

Clinical significance: Although the literature establishes the management of sarcomatoid anaplastic thyroid tumors, surgery remains the first option in severe cases with obstructive symptoms.

Keywords: Anaplastic thyroid carcinoma, Case report, Emergency thyroidectomy, Goiter.

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Introduction

Giant thyroid tumors are defined as any thyroid with a weight greater than 500 gm and a diameter greater than 100 mm.[1] Most commonly they are asymptomatic during their course, however, they can produce compressive symptoms such as dyspnea, dysphagia, and orthopnea when they reach a critical size and displace the regular anatomy.[2] The most critical complication occurs when the trachea is displaced or infiltrated by the tumor causing acute airway occlusion or life-threatening hemorrhage.[3] The evolution of thyroid cancer can be imminent, cases have been described in which anaplastic thyroid cancer (ATC) transforms into differentiated thyroid cancer (DTC), and in which both exist simultaneously.[4] Unlike DTC, anaplastic un-DTC cells are less likely to stain positive for thyroid transcription factor 1 (TTF-1), nor do they have positive staining for thyroglobulin in the anaplastic component of the tumor.[5] The evolution of this anaplasia is unusual, as anaplastic thyroid carcinoma accounts for <1–4% of all thyroid cancers.[6] In terms of histology ATCs are classified into three patterns—sarcomatoid, giant cell, and epithelial, all are characterized by inflammatory infiltrate, macrophages infiltrating the tumor, necrosis, and a high mitotic rate.[7] The sarcomatoid type, is an advanced cancer with infiltration to surrounding organs, blood vessels, and neck skin and can metastasize to the lung or brain.[8] It is considered the most aggressive of thyroid carcinomas, as it often presents with metastatic spread to regional lymph nodes and distant sites.[9] Cases of giant ATC are rarely reported as they are usually detected early for proper management. To our knowledge, this is the first reported case of giant anaplastic carcinoma in a 55-year-old woman with an unusual presentation of sarcomatoid-type ATC with obstructive symptoms.

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Case Description

A 55-year-old female patient from the Peruvian highlands presented to the emergency department with an anterior cervical tumor of 30 years of evolution, which was causing obstructive symptoms such as dyspnea, dysphagia, and dysphonia. She had no significant oncologic history; however, she reported having consumed noniodized salt for almost the first 2s of her life.

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Physical examination revealed a large cervical tumor corresponding to the thyroid gland occupying the entire anterior cervical region from the submental region to the supraclavicular region, predominantly right (Fig. 1). Laboratory tests showed normal serum thyroid stimulating hormone (TSH), T3, T4 levels, as well as complete blood count, biochemical profile, and coagulation tests.

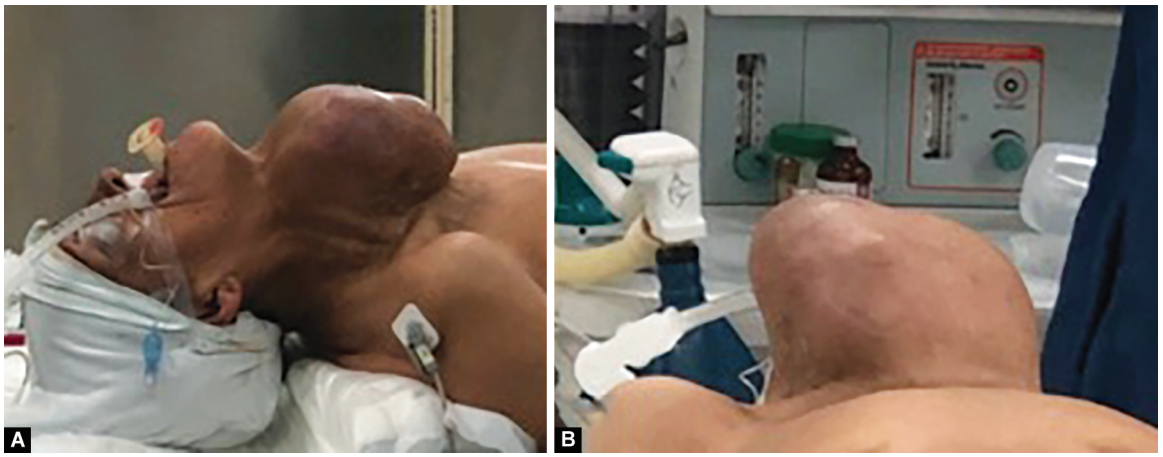
The neck computed tomography scan showed a large lobulated tumor measuring 158 × 133 × 116 mm, with a volume of approximately 1350 CC and central calcifications. Contrast administration showed peripheral uptake and central necrosis; displacement of adjacent structures to the left side and airway compression (Fig. 2A). Chest X-ray showed two mass-like nodular images in the pulmonary parenchyma, which showed metastasis (Fig. 2B).

A total thyroidectomy was performed, where adhesions of the tumor were found to all the parathyroid muscles and to the great vessels of the neck, which were resected; however, toward the mediastinal region a firm adhesion of the tumor was found in the aorta artery and the brachiocephalic trunk, which could not be completely released (R2 resection). In addition, the airway was freed. The patient's evolution was favorable, with a resolution of dyspnea and ingestion of food without difficulty. The patient did not present hypocalcemia or recurrent laryngeal nerve palsy, so she was discharged 4 days postoperatively without complications (Fig. 3). However, the patient died 4 months after discharge due to pulmonary metastases which caused respiratory distress syndrome.

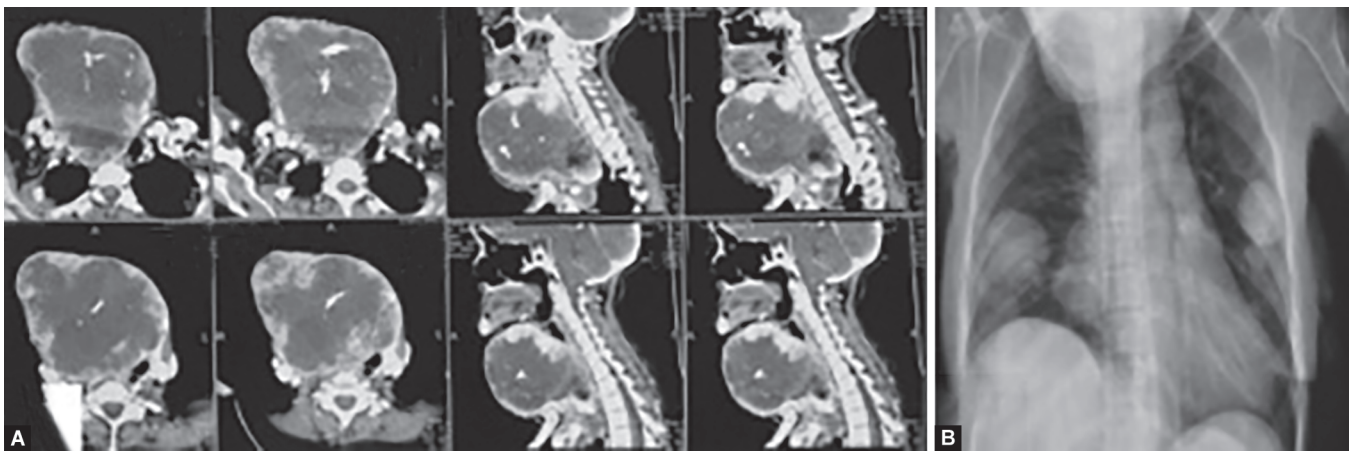
Microscopic examination revealed a bilobulated tumor of 1050 gm, whitish-brown tissue with purulent material accompanied by necrosis in 30%. The microscopic study revealed a malignant neoplasm of pleomorphic pattern, with



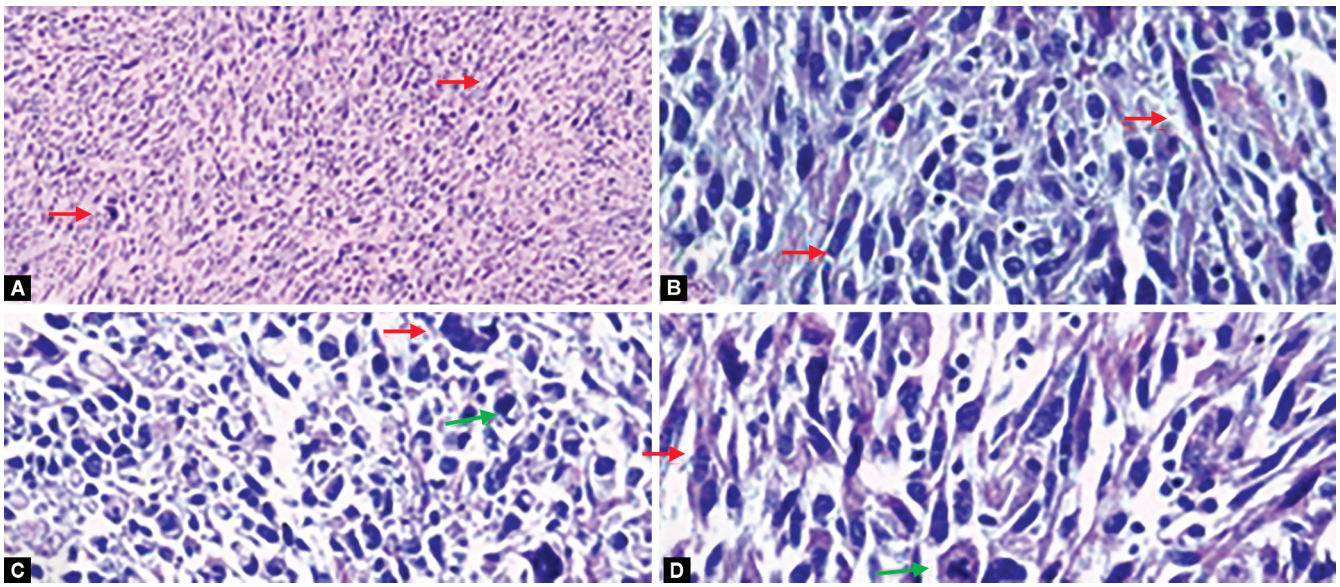
Fig. 3: Postoperative image



Figs 1A and B: (A) Right side view; (B) Bottom to top view



Figs 2A and B: (A) Tomography of the neck, axial, and sagittal section, showing a tumor of 158 × 133 × 116 mm, with an approximate volume of 1350 CC; (B) Chest X-ray with nodular images



Figs 4A to D: (A) Histopathology of the tumor shows undifferentiated cells in the thyroid gland made up of large, spindle-shaped cells with a sarcomatoid appearance. Hematoxylin-eosin stain 100× magnification; (B) Malignant spindle cells are observed (red arrow). Resembling high-grade pleomorphic sarcoma. Hematoxylin-eosin stain 400× magnification; (C) The image shows anaplastic and giant cells (red arrow). Large polygonal cells with high nuclear pleomorphism (green arrow). Hematoxylin-eosin staining, 400× magnification; (D) In this figure, spindle cells with pleomorphic nuclei predominate (red arrow) simulating high-grade sarcoma mitotic figures are evident (green arrow). Hematoxylin-eosin stain, 400× magnification

lymphovascular and perineural invasion present; glandular capsule, extraglandular soft tissues, and surgical margins compromised by the malignant neoplasm. It showed in all fields sarcomatoid cells with large nuclei, pleomorphic, with evident mitotic activity, in addition to giant cells. The stroma was variable in appearance showing fibrosis with hyalinization, desmoplasia, and areas of necrosis. Most of the tumor area consisted of undifferentiated cells (Fig. 4).

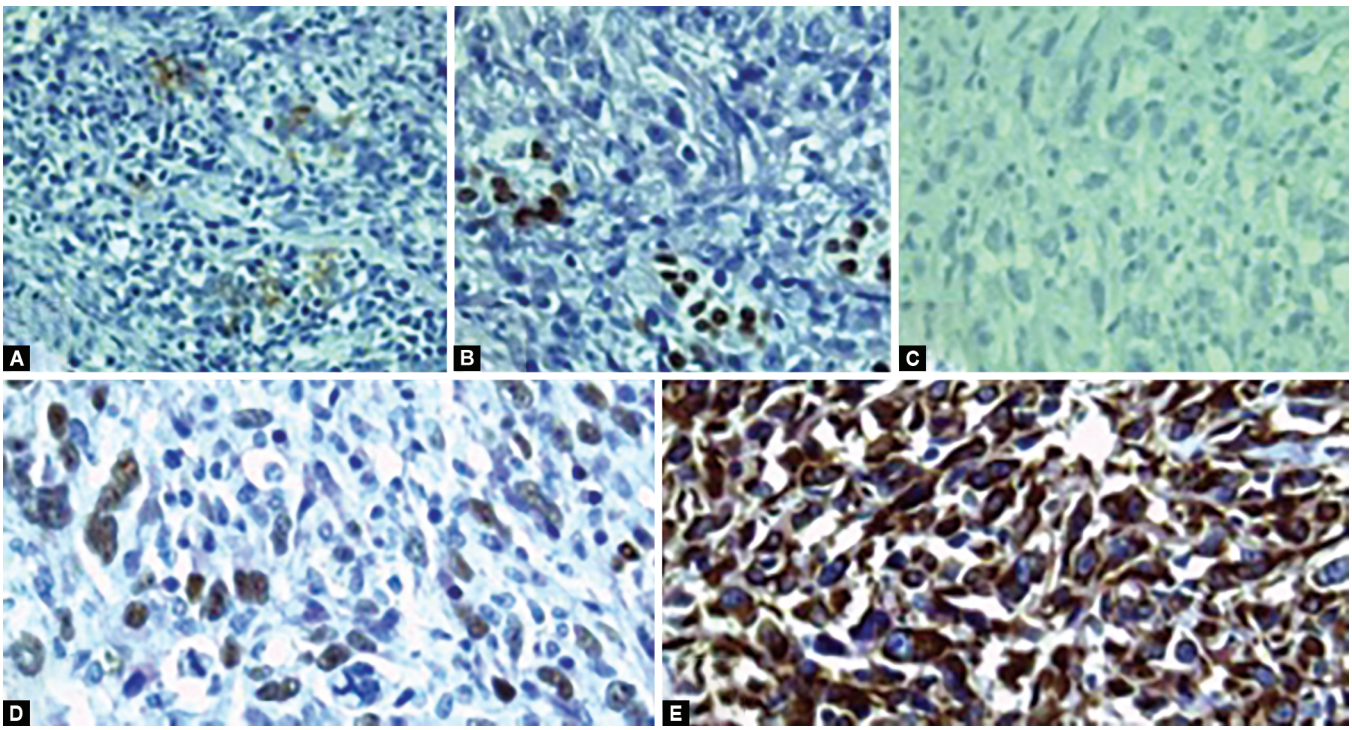
The tumor was removed from the thyroid lobe. The labeling for keratin and TTF-1 was positive in a certain number of cells, whereas the reaction for thyroglobulin was negative. p53 labeling was positive, as well as vimentin with a strong intensity in the spindle cell component. Therefore, the immunohistochemical profile of the tumor concludes an ATC of sarcomatoid type (Fig. 5).

Discussion

In relation to the diagnosis of thyroid tumors, there is no method to predict which thyroid neoplasms will undergo an anaplastic transformation, although it is acceptable that undifferentiated or ATC develops from a well-differentiated one.[10] In the evolution of these tumors, the role of TSH in the evolution of anaplastic thyroid carcinoma is postulated,[11] so it is to be expected that patients with anaplastic thyroid carcinoma present abnormal levels of TSH, However, it has been demonstrated that the growth and metabolic activity of undifferentiated thyroid tumors is independent of TSH function[10] as in the case of our patient in which the evolution of the tumor was observed with normal levels of TSH, T3, and T4. The histopathology of this ATC shows undifferentiated cells in the thyroid gland constituted by large cells with a spindle-shaped appearance like sarcomatoids that respond to hematoxylin-eosin staining, thus concluding with the diagnosis.

In this sense, the diagnosis of ATC is challenging, given that it overlaps morphologically with other malignant neoplasms that show anaplastic characteristics, in these circumstances, the diagnoses must be made from the biopsy.[7] Likewise, the rational use of thyroid-specific markers, as well as other tissue-specific markers, is useful to differentiate ATC from its imitators. In ATC, thyroglobulin and TTF-1 are usually negative (96 and 70% of cases, respectively).[12] In our case, the markers for keratin, TTF-1, p53, and vimentin were positive, and thyroglobulin was negative. The TTF-1 positivity can be explained by its intervention in the embryonic development of the thyroid.[13] Positive thyroglobulin is expressed in certain cases suggesting the possibility that this tumor developed from a well-differentiated thyroid tumor.[14] In relation to the use of the immunohistochemical markers vimentin and p53, the first was used because being an anaplastic carcinoma of sarcomatoid type, it presents a positive staining, cytoplasmic staining, especially in the spindle cell component; besides that, in general, vimentin confirms the tumors which are of mesenchymal origin. The use of this marker had already been used previously in different studies with this same type of tumor, which confirms its correct use since the positivity of spindle cells with vimentin is also mentioned.[15] The use of p53 can be distinguished from immunohistochemistry with predominance at the nuclear level. In the study, it was demonstrated that there was a strong positive nuclear staining of p53 which indicates the presence of mutation, this fact was presented previously in different case reports which also mention the existence of a genetic mutation, besides being the most common, within the ATC.[16] Therefore, according to the anatomopathological and immunohistochemical profile of the tumor, we conclude that it is an anaplastic thyroid carcinoma of the sarcomatoid type.

The staging of anaplastic cancers is considered stage IV cancers[17] IVA is intrathyroid anaplastic cancer, while IVB is anaplastic cancer with macroscopic extrathyroid extension or metastasis in the cervical lymph nodes and IVC with distant metastasis. In this case, as evidenced by the chest X-ray, we



Figs 5A to E: (A) Anaplastic (undifferentiated) carcinoma of the thyroid gland with focal keratin positivity; (B) Focal positivity to TTF-1 is observed in a thyroid sample; (C) Negative thyroglobulin; (D) Expression of the p53 protein in undifferentiated (anaplastic) carcinoma of the thyroid gland; (E) Undifferentiated (anaplastic) carcinoma of the thyroid gland with the presence of vimentin-positive cells

consider that the neoplasm presented was of the IVC type and that they are suggestive of pulmonary metastases. However, after advising the family, we decided not to intervene in the biopsy of the lung masses.

Hatch-McChesne and Lieberman reported that worldwide the population does not consume iodine in a range of 35–45% and also highlights that with a deficient consumption of iodine, the incidence of goiter ranges from 5 to 20%.[18] There are many areas in the highlands of Huancayo-Huancavelica (highlands of Peru) where "stone salt, not iodized" is still consumed, added to the low consumption of foods that may contain iodine (foods that come from the sea). This, together with the scarce consumption of food that may contain iodine (food from the sea), leads to cases of dyshormonogenetic goiters, which can evolve into follicular thyroid carcinoma and later into anaplastic thyroid carcinoma, as occurred in our patient due to the lack of iodine consumption in her first 2s of life.[19]

It is important to keep in mind that ATC is one of the most aggressive cancers and this leads to the fact that it is responsible for half of all deaths caused by thyroid cancers, representing 33–50% of the total.[20] Regarding the life expectancy of patients with ATC, a comparison was made with other case reports, and it was discussed that only 5–10% survive approximately 5 years and the average life expectancy after diagnosis does not exceed 3–6 months.[20]

Similarly in a case report where it is mentioned that a retrospective study of 25 years was made and that 83 clinical histories of patients with this type of tumor were analyzed; the specific survival rate of the disease was 33% for 1 year and 23% for 2 years.[21] In this case, the patient died 4 months postoperatively.

Conclusion

Anaplastic thyroid carcinoma is the rarest and most dangerous of all types of thyroid cancers due to its metastasis and poor prognosis, so extreme caution should be exercised in the treatment choice according to the patient's case.

Clinical Significance

Although the literature establishes clinical management of anaplastic sarcomatoid thyroid tumors, surgery remains the first option in severe cases with obstructive symptoms.

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