Primary Thyroid Sarcoma: A Case Report

Keywords: m*alignant fibrous histiocytoma, sarcoma, thyroid gland, thyroid cancer, undifferentiated pleomorphic sarcoma*

ABSTRACT

Background. Undifferentiated pleiomorphic sarcoma (UPS) of the thyroid is rare. The site of origin is still unknown but UPS has been thought to arise from the fascia surrounding the thyroid gland. The tumor frequently presents as a firm, fixed, aggressively enlarging neck mass associated occasionally with pain, dysphagia, and cough - a clinical course that is closely similar to that of anaplastic thyroid cancer. It is often identified in its advanced stage, with distant metastasis, and has poor prognosis. This paper presents one of the less than 30 cases that have been documented in the English language ever since it was first identified in 1989. In the Philippines, there has been no published report of a primary thyroid sarcoma.

Case Presentation. The author describes a 73-year-old female who presented with a rapidly enlarging neck mass, dysphagia, and weight loss. CT scan revealed a complex enhancing mass in the right thyroid gland measuring 16.5cm x 12.5cm with amorphous calcifications. She underwent tumor debulking, tube gastrostomy, tracheostomy, and radiation therapy. Biopsy revealed a malignant tumor exhibiting high-grade cellular features characterized by storiform pattern and irregular fascicles with variable cellularity. The cells were markedly pleiomorphic, bizarre-looking, and with marked atypia in a background of collagenous stroma. Multinucleated tumor giant cells and atypical mitotic figures were also present. All of these favored the diagnosis of undifferentiated pleomorphic sarcoma. She was discharged well after the initial admission, but succumbed two months after, exhibiting the dismal course and prognosis of this rare thyroid cancer.

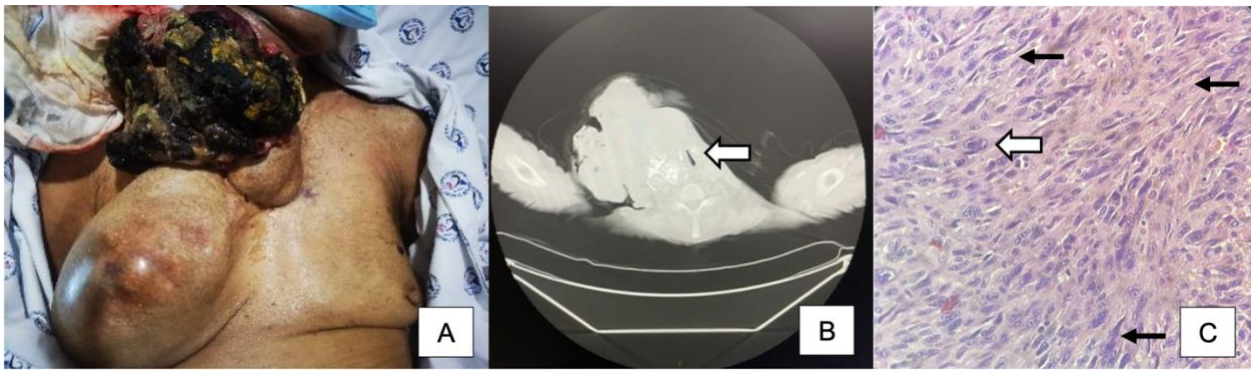

Figure 1. 73-year-old female with a huge neck mass. *(A) Patient who developed a rapidly enlarging neck mass, dysphagia, dyspnea, and weight loss; initially diagnosed with a multi-nodular non-toxic goiter with benign cytology by aspiration biopsy twenty years ago; three months prior to admission, the thyroid mass had progressed to approximately 20 x 15 cm. (B) CT scan of the neck: leftward displacement and compression of the laryngeal lumen (arrow). (C) Histopathology, HPO: irregular fascicles (black arrows) with variable cellularity in storiform pattern, multi-nucleated tumor giant cell (arrow).*

Learning Points. There is still no consensus for the management of thyroid sarcoma but surgery plays a central role; this involves thyroidectomy plus excision of involved tissue with or without neck dissection. The recommendation for surgical margins of head and neck sarcomas is to do a complete tumor resection with as wide a margin as is feasible, with the least morbidity possible; but due to the anatomical and functional complexity of

the head and neck and the aggressiveness of sarcomas, resections with adequate margin are not easily obtained. Since surgical treatment alone gives poor results, adjuvant radiotherapy and/or chemotherapy is often utilized even if chemotherapy's role for UPS remains unknown. The few existing studies on the advantages of chemotherapy detect no differences in overall survival.

Conclusion. Primary thyroid sarcoma is a rare malignancy that shares similar clinical features with anaplastic thyroid cancer. Similarly, it also carries a poor prognosis. Histopathology is important in arriving to final diagnosis. Due to the aggressive growth and infiltration of this kind of tumor into the adjacent structures, airway and nutrition may be compromised by the time of presentation. The mainstay of treatment is surgery, radiotherapy, and clinical management of the malignancy's complications; therefore, a multi-disciplinary team composed of surgical specialists and clinicians is crucial for the improved survival of these patients