

Thyroid Sarcomatoid Carcinoma – Showing Positive Scan on ^{18}F FDG PET-CT

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ABSTRACT

A 47 years old woman presented with a longstanding multinodular goiter (MNG) for 10 years with history of Fine needle aspiration cytology (FNAC) proven nodular goiter at her initial diagnostic workup and no further follow up. Recently, she noticed rapidly growing swelling of neck with occasional shortness of breath and pain within the mass. A thyroid scintigraphy at the National Institute of nuclear Medicine & Allied Sciences (NINMAS) revealed multinodular goiter with a dominant cold nodule in left lobe. Patient underwent total thyroidectomy with left sided neck dissection and clearance of cervical lymph nodes. Histopathology reported metastatic papillary carcinoma with spindle cell differentiation. Immunohistochemistry findings were suggestive of sarcomatoid carcinoma presumably with a spindle cell differentiation at metastatic site. Post thyroidectomy imaging with ^{18}F -fluorodeoxyglucose Positron Emission Tomography-Computed Tomography (^{18}F -FDG PET-CT) revealed irregular lobulated soft tissue area with intense FDG avidity in neck region. Diagnostic whole body scan (WBS) with ^{131}I showed no uptake of radioiodine anywhere in the body. Patient received chemotherapy but survived only two months after that.

Keywords: Sarcomatoid tumor, ^{18}F -FDG PET-CT, Cold nodule, Spindle cells.

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INTRODUCTION

Sarcomatoid carcinoma is a type of aggressive carcinoma, that is a combination of carcinoma (cancer that begins in the skin or in tissues that line or cover internal organs in the body) and sarcoma (cancer of the bone, cartilage, fat, muscle, blood vessels, or other connective or supportive tissue). The majority of sarcomatoid carcinomas of thyroid are anaplastic carcinomas. Sarcomatoid carcinoma resembles spindle cells, which are sarcoma-like, and characterized by an admixture of anaplastic epithelial and mesenchymal

components. It is found in the head and neck region including the larynx, tongue, nasal cavity and thyroid gland (1). We report a case of MNG which gradually transformed into thyroid sarcomatoid carcinoma and short survival of the patient.

CASE REPORT

A 47 years old woman was referred to NINMAS with a request for an ^{131}I WBS and thyroid scintigraphy. The patient had history of neck swelling for long 10 years which was cytologically diagnosed as a case of MNG. Both the history and the growth of the goiter were unremarkable for a decade. After 10 years, she presented with a rapidly growing neck mass associated with occasional shortness of breath and pain within the mass. High resolution ultrasound (HRUS) of neck showed a fairly big multinodular goiter with the biggest solid, heterogenous nodule in left lobe measuring about 5 X 4.5 cm. $^{99\text{m}}\text{Tc}$ thyroid scan confirmed MNG with a dominant cold nodule in left lobe (Figure 1).

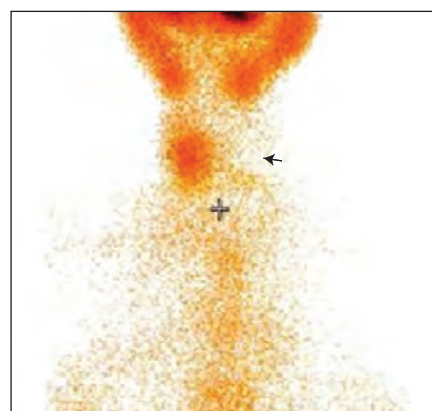


Figure 1: $^{99\text{m}}\text{Tc}$ thyroid scintigraphy image showing patchy tracer uptake in a multinodular goiter with a dominant cold nodule (arrow) in the mid-lateral area of left lobe

Patient underwent total thyroidectomy with neck dissection for the clearance of neck lymph nodes of level V. Histopathology reported well differentiated papillary thyroid carcinoma (classical) with metastatic lymph nodes having spindle cell differentiation. Differential diagnosis was spindle cell neoplasm as a separate entity. Immunohistochemistry (IHC) findings were suggestive of sarcomatoid carcinoma with presumable spindle cell differentiation at metastatic site. Molecular marker cytokeratine/pankeratin was positive. Beside these, Vimentin (a structural protein encoded by the VIM gene in humans) was also positive. However, S 100 and Thyroid Transcription Factor 1 (TTF 1) were negative. Whole body ¹⁸F-FDG PET-CT scan showed absent thyroid gland (thyroidectomized), irregular lobulated soft tissue density mass (Figure 2)

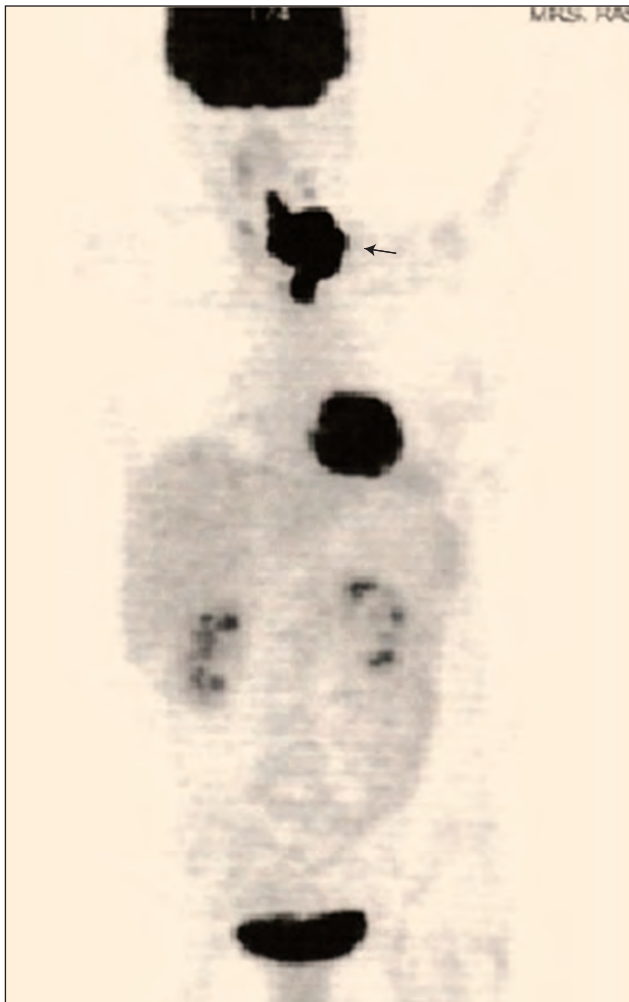


Figure 2: Sagittal section image of the whole body ¹⁸F- FDG PET-CT with intense metabolic activity (arrow) in the left side of the neck

with intense FDG avidity at the left paratracheal region measuring about 8 X5X6 cm and SUV max: 26.0 (Figure 3, 4 & 5) and appeared to be invading the trachea and pushing the trachea towards right.

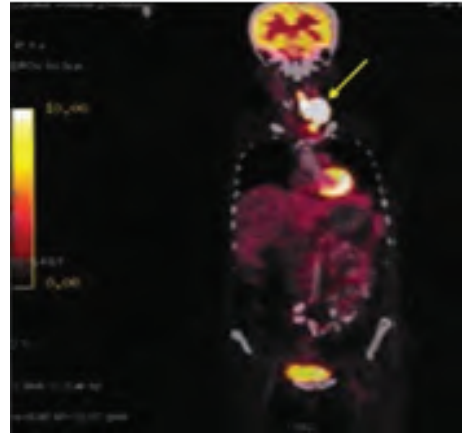


Figure 3: Coronal section image of the whole body ¹⁸F-FDG PET-CT with intense metabolic activity (yellow arrow) in the left paratracheal region of the neck

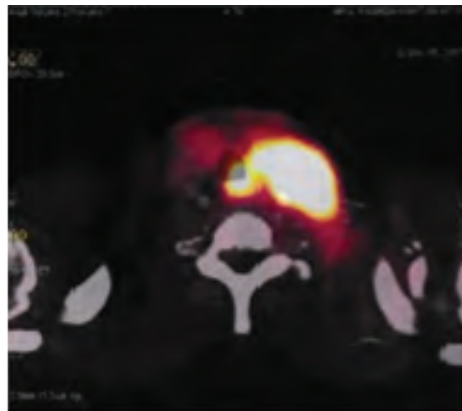


Figure 4: Transverse section image of neck showing intense FDG uptake in 4th cervical vertebral region

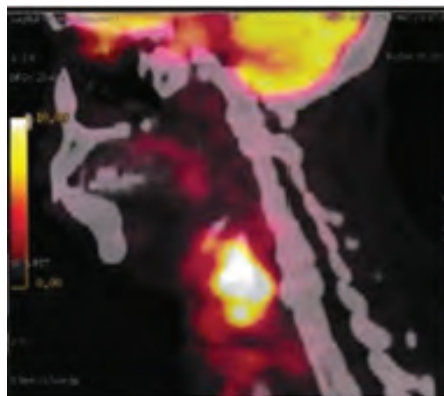


Figure 5: Sagittal section image of neck region showing intense FDG avidity in an irregular shaped area

Multiple discrete FDG avid lymph nodes were seen on left side of the neck at level II (SUV max: 4.5). FDG avid (SUV max: 9.9) soft tissue mass lesion appeared to be involving the left vocal cord. FDG avid nodules were present in the right lower lung with bilateral mild pleural effusion and mild hepatosplenomegaly. Patient was clinically staged as stage IV. Unfortunately, there was no scope of radioiodine therapy as the diagnostic WBS with ^{131}I showed no uptake and reported as negative. Chemotherapy with injection Doxorubicin and cisplatin continued under oncologist's supervision but patient could tolerate only few and survived two months after the diagnosis.

DISCUSSION

Carcinoma of the thyroid gland arising from follicular cells comprises about 95% of all thyroid tumors. The most common forms of thyroid malignancy are papillary and follicular carcinoma and also known as "Well differentiated thyroid carcinoma" (WDTC) with an overall survival (OS) rate of more than 90%. On the other hand, Hurthle cell carcinoma have an OS of 1.2-10% and anaplastic thyroid carcinoma (ATC) only 2% - 5% of all thyroid cancers (2, 3). Reported death from thyroid carcinoma reaches to almost 40% due to the aggressive nature of anaplastic variant (3). Maximum patients present with local or distance metastases at the time of initial diagnosis. The common sites of metastases are lung (42%), followed by bone (32%) and brain ((9%). Microscopic appearance of anaplastic thyroid carcinoma is highly variable and frequently encountered patterns include **i)** Squamoid-looks like non-keratinizing squamous cell carcinoma, without follicles, papillae, or even trabeculae or nests. **ii)** Sarcomatoid-containing spindle cells and giant cells showing fascicular or stormiform growth pattern and **iii)** Giant cell (3, 4). Sarcomatous thyroid carcinoma does not form follicles, papillae, or even trabeculae or nests. Spindle cell carcinoma of thyroid is very rare. Immunohistochemistry is the gold standard for diagnosis and the most useful marker is keratin and vimentin (3, 5). The reported patient was diagnosed by immunohistochemistry. Molecular markers cytokeratine and vimentin were positive. Vimentin expression is a marker of mesenchymal differentiation, which may

be clinically useful to assess the disease aggressiveness. But S 100 and TTF 1 were negative in spindle cell thyroid carcinoma (5, 1). These two immunohistochemical marker were also negative in this patient. ^{18}F -FDG PET has a potential role in the clinical assessment of ATC. One study showed intense uptake on ^{18}F -FDG PET images in 8 out of 16 (50%) patients (6). Reported patient also had intense FDG uptake in the primary site i.e. thyroid bed and also metastatic areas. PET has a direct impact in diagnosis and management of ATC where diagnostic WBS with ^{131}I fails to determine the primary site and specifically localize metastases. Sarcomatoid thyroid carcinoma is a disease of the old age (average 66.5 years) with a M:F ratio of 1:3.1 (7, 8). Clinically ATC grows rapidly as a firm mass on top of a long standing goiter (9) and that matches with the mentioned case. Most of anaplastic thyroid carcinoma grow larger than 5 cm and usually cause hoarseness of voice, dyspnea or dysphagia (8, 9) which correlates with the reported patient who had long standing goiter more than 8 cm in size causing pressure effects in the neck. Undifferentiated anaplastic sarcomatoid carcinoma of the thyroid is known for its aggressive nature, which tends to invade the surrounding tissues like true sarcoma. Tracheal invasion of the described patient explains this type of aggressiveness as well. Multidisciplinary team approach of surgeon, radiation oncologist, chemo-oncologist, palliative oncologist may combinedly deal with the limited opportunities in a patient of ATC. Spindle cell carcinoma is a highly lethal variety with a median survival of 4-12 months from the time of diagnosis (8, 10, 11) which corresponds to the poor survival period of this case too. A large portion of ATC may develop from longstanding goiter or from preexisting incompletely treated papillary or follicular thyroid carcinoma (12). The reported patient had a longstanding slow growing goiter which turned out to be an aggressive one. Anaplastic sarcomatoid carcinoma of the thyroid is extremely rare, highly recurrent, locally advancing and entirely reflecting this case report.

CONCLUSION

From the reported case it may be concluded that sarcomatoid carcinoma of thyroid is iodine non-avid, highly

¹⁸F FDG avid having poor survival. Long standing MNG may transform to aggressive carcinoma like sarcomatoid tumor.

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