ORIGINAL RESEARCH PAPER

Oncology

KEY WORDS: Thyroid gland ; Sclerosing Mucoepidermoid carcinoma of thyroid with eosinophilia; Hashimoto thyroiditis;

A RARE CASE OF SCLEROSING MUCOEPIDERMOID CANCER WITH EOSIOPHILIA OF THYROID GLAND-A CASE PRESENTATION

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ABSTRACT

Sclerosing mucoepidermoid carcinoma of the thyroid with eosinophilia (SMCE) is a rare neoplasm of the thyroid which resembles the salivary mucoepidermoid cancer.It has been reported predominantly in women and that too in a background of Hashimoto's thyroiditis. In the World Health Organisation Classification of endocrine and neuroendocrine tumours of the thyroid published in 2022, Sclerosing mucoepidermoid cancer with eosinophilia of thyroid is listed under thyroid tumours of uncertain histogenesis where the cell lineage is unclear. Here we report a case of a 50 year old lady who presented with an anterior neck swelling and underwent total thyroidectomy. Histopathology and immunohistochemistry had confirmed the diagnosis. Being rare, there is no consensus regarding the optimal treatment guideline. Even though this condition was previously considered to be a low grade malignancy, metastatic cases have been reported recently. Therefore in view of the close surgical margins, our patient underwent adjuvant postoperative radiotherapy. She is now disease free and on follow up.

INTRODUCTION

Sclerosing mucoepidermoid cancer with eosiophilia is a rare malignancy of the thyroid characterized by nest or strands of epidermoid tumor cells with squamous differentiation, rare mucous cells, prominent sclerotic stroma, eosinophilic and lymphoplasmacytic infiltration, and a background of chronic lymphocytic thyroiditis in the non-neoplastic thyroid gland.(1)Even though Sclerosing mucoepidermoid carcinoma of the thyroid was first reported in 1991(2), It has been recently recognized as a separate entity. So far approximately 50 cases have been reported in the literature (1) Although it was considered a slow growing benign tumor, recently metastatic cases have also been reported which show a locally aggressive clinical picture.(3,4,7,9). In published cases, the Sclerosing mucoepidermoid cancer with eosiophilia of thyroid gland predominantly occurs in women, although rarely it has been reported in men too(1).It usually occurs in a background of Hashimoto's thyroiditis.(5). Pathological classification-

Overview of the 2022 World Health Organisation Classification ofThyroid neoplasms(6)

Developmental abnormalities
1.Thyroglossal duct cyst
2.other congenital thyroid abnormalities
Follicular cell derived neoplasms
1.Benign tumours
A.Thyroid follicular nodular disease
B.Follicular Adenoma
C.Follicular adenoma with papillary architecture
D.Oncocytic adenoma of the thyroid
2.Low risk neoplasms
A.Non -invasive follicular thyroid neoplasm with papillary like nuclear features
B.Thyroid tumours of uncertain malignant potential
C.Hyalinizing trabecular tumour
3.Malignant neoplasms
A. Follicular thyroid carcinoma
B.Invasive encapsulated follicular variant papillary carcinoma
C.Papillary thyroid carcinoma
D.Oncocytic carcinoma of the thyroid
E.Follicular-derived carcinomas,high grade
Differentiated high-grade thyroid carcinoma
Poorly differentiated thyroid carcinomas
F.Anaplastic follicular cell derived thyroid carcinoma
Thyroid C- cell- derived carcinoma
1.Medullary thyroid carcinoma
Mixed medullary and follicular cell derived carcinomas
Salivary gland type carcinomas of thyroid
1.Mucoepidermoid carcinoma of the thyroid
2.Secreatory carcinoma of salivary gland type
Thyroid tumours of uncertain histogenesis
1.Sclerosing mucoepidermoid carcinoma with eosinophilia
2.Cribriform morular thyroid carcinoma
Thymic tumours within the thyroid
1.Thymoma family
2.Spindle epithelial tumour with thymus like elements
3.Thymic carcinoma family
Embryonal Thyroid Neoplasms
1.Thyroblastomas

Case Presentation-

A 50 year old hypothyroid woman on thyroxin for the past 10 years presented with an anterior neck swelling of 2 months duration. There were no other swellings elsewhere in the body. Routine blood investigations like Complete blood count, serology and biochemistry were normal. A pan endoscopy ruled out other primaries in the upper aero digestive tract.

Radiological Findings

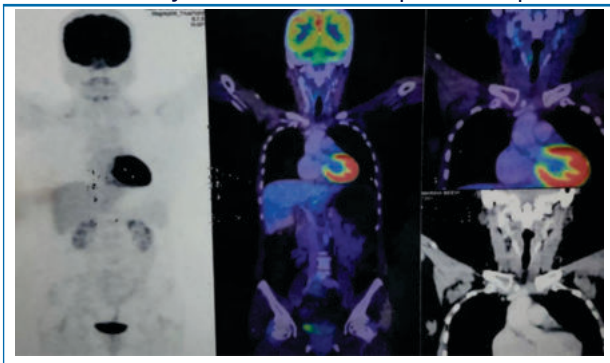
An initial Ultrasonography of the neck showed a Thyroid Imaging Reporting And Data System (TIRADS) II lesion with a few internal calcifications measuring 2.2x1.4 cm. Ultrasound guided Fine needle aspiration showed florid lymphocytic thyroiditis suggestive of Hashimoto's thyroiditis. A repeat Ultrasound after 5 months showed poorly defined nodule in the right lobe measuring 25x29mm with clusters of micro calcifications Thyroid Imaging Reporting and Data system (TIRDAS)IV.After pathological confirmation of the disease a Positron emission tomography -computed tomography scan was taken which showed no other metastasis elsewhere. This helped to rule out metastasis from other sites. Usually sclerosing mucoepidermoid cancer with eosiophilia's are cold on radio nucleotide scans.

Clinical Diagnosis

Primary localized thyroid malignancy

Differential Diagnosis

1. Follicular cell derived neoplasms
2. Thyroid C- cell- derived carcinoma
3. Other rare histologies of primary thyroid malignancy.



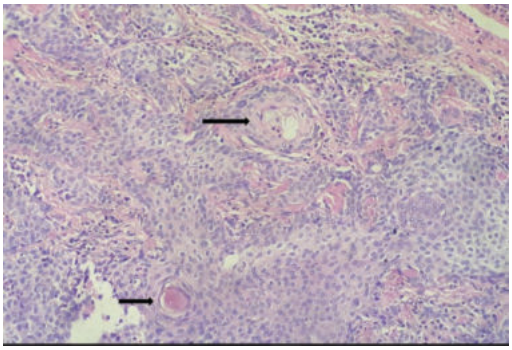
Picture1-PET scan showing no FDG avid areas.This helped to rule out metastasis from other sites.

PATHOLOGICAL DISCUSSION

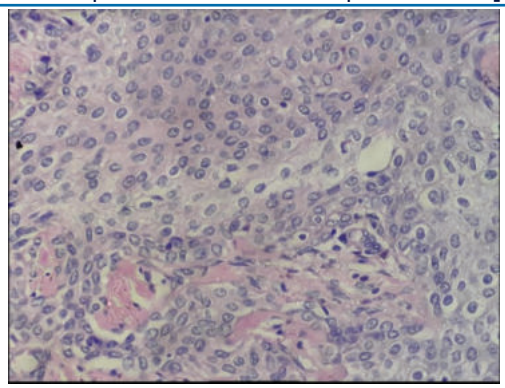
Uequivocal cytological diagnosis is not an easy task. Guided fine needle aspiration cytology showed scattered aneucleate squames singly and in clumps along with keratinized mature squamous cells. This patient underwent total thyroidectomy and histopathology was suggestive of sclerosing mucoepidermoid carcinoma with eosinophilia and predominant epidermoid component, with the neoplasm seen close to the inked margin. Adjacent thyroid showed Hashimoto's thyroiditis. Immunohistochemistry done showed that tumor cells are P40+, Cytokeratin 7-,thyroglobulin- , Thyroid Transcription Factor – and mucin – consistent with Sclerosing mucoepidermoid cancer with eosiophilia of thyroid.

Conventionally this condition is characterized by nests or strands of epidermoid tumor cells with squamous differentiation, rare mucous cells, prominent sclerotic stroma, eosinophilic and lymphoplasmacytic infiltration, and a background of chronic lymphocytic (Hashimoto) thyroiditis in the non-neoplastic thyroid gland. In view of the presence of keratin pearls Sclerosing mucoepidermoid cancer with eosiophilia of thyroid are sometimes confused with the more aggressive squamous cell carcinomas of the thyroid. Squamous cell carcinoma's may be primary , secondary or other thyroid neoplasms with squamous differentiation.

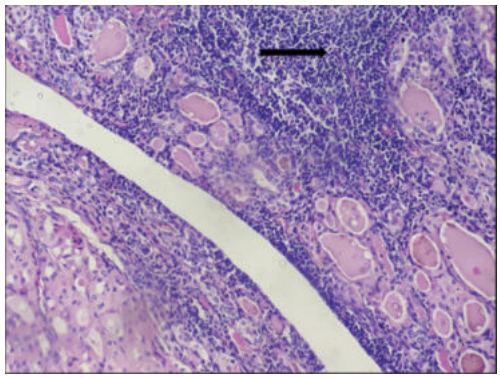
Sim et al compared Sclerosing mucoepidermoid cancer with eosiophilia with conventional mucoepidermoid cancer of thyroid gland and supported the former to be a distinctive entity [8].Usually both were positive for cytokeratin and negative for calcitonin. All cases of mucoepidermoid cancer were positive for thyroglobulin, whereas all cases of Sclerosing mucoepidermoid cancer with eosiophilia were negative.(9) Hunt et al revealed the expression of p63 in Sclerosing mucoepidermoid cancer with eosiophilia of thyroid gland suggesting a tumor origin of ultimobranchial body/solid cell nest rather than follicular epithelial cells. Few authors also suggested an origin from C-cells, parathyroid, ectopic salivary gland, and thyroglossal duct. (6, 11).With prominent eosinophilic infiltration and sclerosis This condition may also sometimes be mistaken for Hodgkin's disease especially when it presents as a nodal disease.(10)



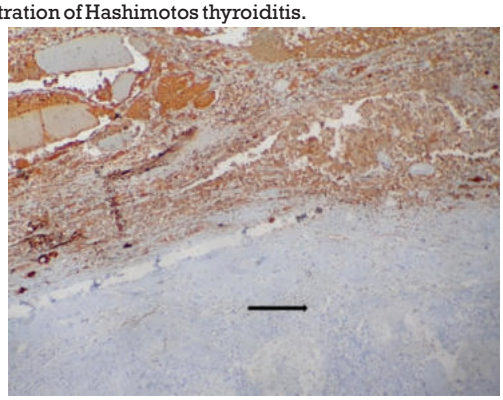
Picture2-Low power view of the thyroid showing keratin pearls



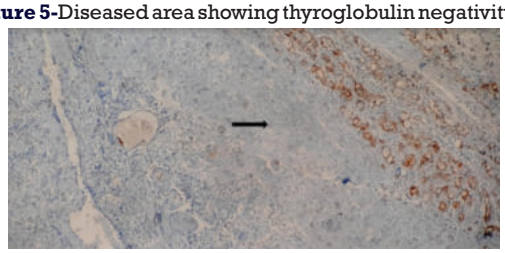
Picture 3-High power view showing typical thyroid follicles.



Picture 4-Adjacent areas in the thyroid showing lymphocytic infiltration of Hashimotos thyroiditis.



Picture 5-Diseased area showing thyroglobulin negativity



Picture 6-Areas in thyroid showing negative for Cytokeratin 7

DISCUSSION ON MANAGEMENT

Treatment guidelines have not been formulated in view of the rarity of the disease. As this condition was initially thought to be a low grade malignancy, a total thyroidectomy was done in all cases as a part of the treatment. Recently metastatic and aggressive varieties have been reported .In our patient, a total thyroidectomy was done.The resected thyroid tissue was yellow and firm .Histopathology was suggestive of sclerosing mucoepidermoid carcinoma with eosinophilia and predominant epidermoid component, with the neoplasm seen close to the inked margin. Adjacent thyroid showed Hashimoto's thyroiditis. Immunohistochemiatry showed tumour cells that are p40+, Cytokeratin 7 -, thyroglobulin- ,Thyroid Transcription Factor -and mucin – consistent with

Sclerosing mucoepidermoid cancer with eosiophilia of thyroid.In view of close margins at pathology the patient was treated with external beam radiotherapy of 45Gray in 20# as 1.12 Gray per fraction over 4 weeks followed by 15Gray as 6 fractions over 6 days as 1.25 gray per fraction .Patient is currently on follow up and disease free.

Newer molecular studies have shown involvement of the RAS-RAF-MEK-ERK signaling pathway in the pathogenesis. There fore BRAF inhibitors may prove to be useful in treatment of aggressive varieties of Sclerosing mucoepidermoid cancer with eosiophilia of thyroid with activating BRAF mutations.(12)

Final Diagnosis

Sclerosing mucoepidermoid carcinoma of the thyroid with eosinophilia.

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