Refrence Number 4,5,6,9,11,13 Not site seen

Case Report

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A Rare Presentation of Thyroid Swelling Epitheliod Angiosarcoma: A Case Report

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Abstract

Thyroid angiosarcoma is a very rare malignancy that is mostly seemn in elderly of alpine regions, related to high prevalence of iodine deficient goiter. Its is a very aggressive tumour that can rapidly spread to the cervical lymph nodes, lungs, brain or can metastasize to the duodenum, small bowel and large bowel. Metastatic disease is associated with poor prognosis and limits the mean survival time to a few months after diagnosis and surgical treatment.

Keywords: Thyroid; Epitheloid angiosaroma; Distant spread.

INTRODUCTION

Thyroid angiosarcoma is a rare, aggressive, mesenchymal tumor of the thyroid gland with vascular differentiation. It mainly occurs in adult females; the highest incidence is reported in the 7th decade. Its prognosis is considered very poor with early metastases occurring at lymphnodes, lung, skin, bones, soft tissues, and with a mean overall survival of less than a year. Thyroid Angiosarcoma usually presents as a large and hemorrhagic thyroid mass that extends to local tissues, lymphnode, and distant sites. The non-neoplastic gland frequently shows multinodular goiter.

Thyroid angiosarcoma was originally described in iodine-deficient areas of the Alps and other mountain regions in association with endemic goiter. It accounts for up to 4.3% of all malignant thyroid tumors in Switzerland and its presence is documented in other mountain regions such as Austria and Northern Italy. Although there have been case reports of angiosarcoma in patients without goiter, many patients may not be aware of an underlying thyroid disease until a tumor is detected. Several cases have also been reported in non-alpine areas although with unknown incidence. The coexistence of AS with Hashimoto's thyroiditis or differentiated thyroid cancer has been reported.

CASE PRESENTATION

A 48 year old gentleman resident in assam with a known case of type II diabetes mellitus on OHA

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came in with complaints of lump in the neck, rapidly growing in size, associated with pain. No hyperthyroid or hypothyroid symptoms noted.

On examination patient was alert, awake, oriented and moderately build. No palpable cervical lymph nodes.

Neck findings: 4x3cm lump seen and palpable in the anterior aspect of neck more towards the left, solid in nature, irregular, tender, moves with deglutition and doesn't move with tongue protrusion.

FNAC showed mildly atypical features in a background of lymphocytic thyroiditis with hurthle cell hyperplasia.

640 slice CT neck contrast done which showed a lobulated hypoenhancing solid cystic mass lesion involving the left lobe and isthmus of thyroid gland measuring 4.8 x 3.6 x 4.8cm. The superior aspect of the mass consists of predominantly solid enhancing component and the inferior aspect of the lesion consist of non enhancing cystic components. Anteriorly the mass is infiltrating the strap muscles and closely abuting the left sternocleidomastoid. A few small left level II cervical adenopathy and above findings are highly suspicious for neoplastic thyroid mass.

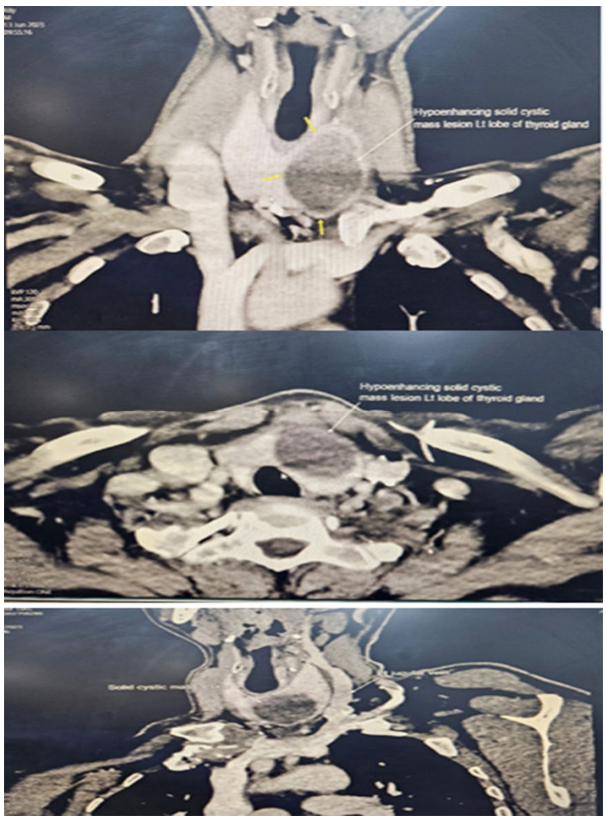

Fig. 1: 640 Slice CT neck contrast

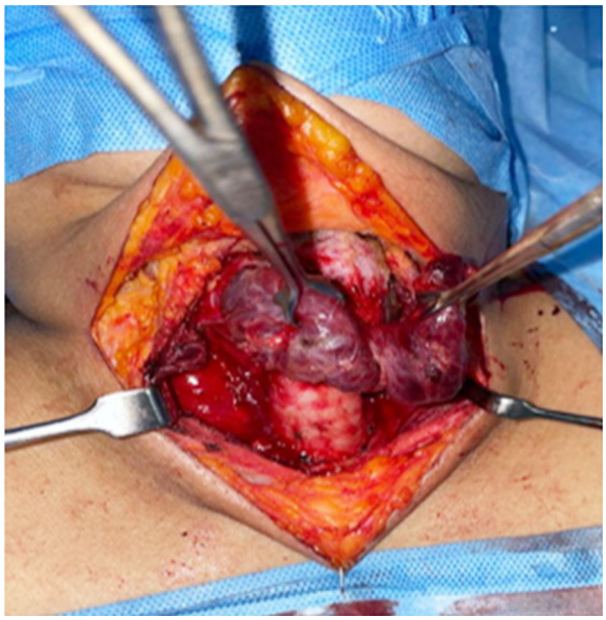

Fig. 2: Intraoperative image of thyroid mass

There was a 4*3*4 cm well encapsulated thyroid mass, solid cystic in type, irregular in shape. Total thyroidectomy was performed, the mass was sent for histopathological evaluation and IHC.He tolerated the procedure well. Post operative period was uneventful. Patient was discharged on the day of surgery with drain.

IHC: left lobe of thyroid: PAN cytokeratin, vimentin, CAM 5.2, ERG - positive Impression: IHC findings in favour of an epitheliod angiosarcoma left lobe of thyroid.

HPE: Unifocal, left lobe of thyroid and isthmus, poorly differentiated thyroid carcinoma. Tumour necrosis present. Angioinvasion present. Lymphatic invasion present. Extrathyroid invasion present (macroscopic and histologically performed). Margins positive for tumour (left lobe). Regional lymph node status: not submitted. Pathological stage classification: AJCC eighth edition: pT3b pNn(not assigned).

DISCUSSION

Thyroid gland vascular tumors range in severity from benign hemangiomas, like infantile hemangioendothelioma, which spontaneously regresses, to angiosarcoma, which is severely aggressive locally and has a dismal prognosis. The original geographical range for angiosarcoma was restricted to the mountains. These tumors are now also occasionally recorded from non-mountainous

regions such as northern France, Hong Kong, and US coastal areas. In this instance, we are covering an Indian case.

Thyroid angiosarcoma is a very uncommon tumor that is highly seen in European Alpine regions where it represents 10% of all thyroid neoplasms.[1] This predilection for mountainous regions has been explained by iodine deficiency leading to thyroid goiters. In the English literature, few non-alpine thyroid angiosarcoma have been reported, suggesting the presence of other etiological factors not yet known, involved in the pathogenesis of this disease.[2] The patients' ages ranged from 50 to 88 years, with a female predominance (Female male ratio, 9:3).[3] Clinical and radiological features are nonspecific and include thyroid mass with compression symptoms or signs related to distant metastasis. On gross examination, the tumor is usually a single nodule commonly filled with bloody fluid and compressing thyroid.

Angiosarcomas are primarily found in older females and have a lengthy history of goiter with a recent, sharp growth in size. Both the pathologist and the clinician/radiologist face significant challenges when dealing with cases of persistent nodular goiter. Numerous disorders are manifested by such lesions. Angiosarcoma on fine needle aspiration cytology closely resembles the number of distinct lesions, leading to a false positive. As a result, in these situations, the final diagnosis is typically determined only after the removed tumor has undergone immunohistochemistry and histopathology.

Immunopositivity for vascular markers for example, CD31, CD34, factor VIII related antigen, and absence of epithelial markers, greatly helps in diagnosis. But the distinction between angiosarcoma and anaplastic sarcomatoid carcinoma is difficult. Immunonegativity for thyroglobulin supports a diagnosis of angiosarcoma which is not found anaplastic carcinomas, confirming that these lesions are unrelated malignant tumors. In our case also, the pathologist had a difficulty in diagnosing the tumor on cytopathology however the diagnosis was established with HPE & IHC.

Some authors doubt the existence of thyroid angiosarcoma, because they are of the opinion that the reported cases were being classified as anaplastic carcinomas with angiomatoid features. They are of the opinion that angiosarcomas are transitional tumors. These tumors show a variable appearance of mesenchymal metaplasia with both epithelial and endothelial differentiation. Angiosarcoma is the extreme in the spectrum of endothelial differentiation. WHO classification of thyroid tumors, published in 2004 has added the entity of poorly differentiated carcinoma and a variety of rare thyroid malignancies, such as angiosarcoma to its four traditional major tumor groups (papillary, follicular, medullary, and anaplastic carcinoma).

Aside from the diagnosis, the tumor's high recurrence rate and locally aggressive and destructive behavior present treatment challenges for a physician. In terms of adjuvant therapy, some argue that following radical surgical excision of the tumor, chemotherapy and radiotherapy are necessary; others contend that if the patient receives chemotherapy and radiotherapy and the tumor is not surgically corroded, these treatments only serve as palliative measures because radical surgical excision of the tumor is always the preferred course of action.

Regarding prognosis of the tumor, poor outcomes occur mostly with extracapsular tumor spread and distant metastasis. Data on survival are sparse. Goh et al. showed a 5-year survival rate of 33.3%.[7] But most of the patients die in less than 6 months regardless of the treatment with a few surviving up to 5 years.[8,10] Entirely intrathyroid tumors generally have a longer survival than those with extrathyroidal extension.[12] Cases from non-mountainous, non-endemic goiter areas have a better prognosis.[10,12]

CONCLUSION

Thyroid angiosarcoma is a rare thyroid tumor that might be difficult to differentiate from anaplastic carcinoma and metastatic angiosarcoma. In the most challenging circumstances, the problem can be resolved by utilizing a large panel of antibodies in conjunction with epidemiological and clinical data.

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