Case Report

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Unusual case of a midline neck swelling in a fifty year old female

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ABSTRACT

Goitrous enlargement of the thyroid can present as a massive enlargement of the neck. It can be so huge as to obscure detection of an underlying malignancy. Poorly differentiated carcinoma of the thyroid gland is very rare. A 50 year old female patient who was diagnosed preoperatively as having colloid goiter underwent near total thyroidectomy was diagnosed with poorly differentiated thyroid carcinoma on histopathological examination. This case report highlights the various architectural patterns, azzopardi phenomenon with emphasis on immunohistochemical stains and an aberrant rhabdoid phenotype of tumor cells along with review of literature.

Keywords: Poorly differentiated carcinoma, Thyroid, Rhabdoid, Azzopardi phenomenon

INTRODUCTION

Poorly differentiated thyroid carcinoma is a very rare, aggressive and often fatal tumor of the thyroid comprising of less than 1% of all endocrine malignancies. It was described originally by Langhans in 1907 under the term 'wuchernde struma' and characterized by locally aggressive behavior and a typical architectural pattern of forming insulae. Herein, we report an unusual case of midline neck swelling along with emphasis on review of literature.

CASE REPORT

A 50 year old female came to the Surgical OPD for a midline neck swelling since 10 years of duration. She also complained of stridor and sleep apnea. On examination, she had a thyroid swelling that moved with deglutination measuring approximately 10x10cm in its greatest dimensions [Figure 1]. A solitary non-tender right-sided submandibular lymph node was also palpable measuring 1×1 cm. On imaging, C.T. scan revealed a 9.7 \times 6.7 \times 8.2 cm hetero-dense lesion arising from the

thyroid gland displacing the carotid and internal jugular vein posteriorly and deviating the trachea to the left without any retrosternal extension. Multiple septae and calcifications within the lesion were noted [Figure 2]. The radiologist signed it out as long-standing goitrous enlargement of the thyroid gland and advised fine needle aspiration cytology (FNAC) correlation. Serum thyroid hormone levels were all within normal limits. (T3 – 1.59 ng/ml, T4 – 5.39 micrograms/dl, TSH – 1.77 microIU/ml).

FNAC performed showed scant cellularity and thin colloid mixed with blood which was inconclusive. The patient was posted for near total thyroidectomy. We received a specimen with a predominantly enlarged lobe measuring $10 \times 7 \times 6$ cm and a small lobe measuring 1×1 cm, the entire gland weighing 200 gm. External surface was smooth and partially encapsulated. Cut section showed brownish solid areas with variegated appearance. Areas of hemorrhage and necrosis were noted. On microscopic examination, H & E stained sections studied showed a partially encapsulated tumor comprising of tumor cells arranged in insulae, nests, cords, trabeculae

and syncytial sheets which was seen invading the capsule. The tumor cells had bizarre nuclei, with marked nuclear pleomorphism ranging from oval to spindle to large atypical shapes. Multiple tumor giant cells and atypical mitoses were noted. Few tumor cells had abundant eosinophilic cytoplasm with eccentrically placed nuclei reminiscent of rhabdoid differentiation. The tumor nests were separated by hyalinized stroma. A striking feature on microscopic examination was the presence of Azzopardi phenomenon [Figure 3].



Figure 1: Clinical photograph.



Figure 2: CT scan.

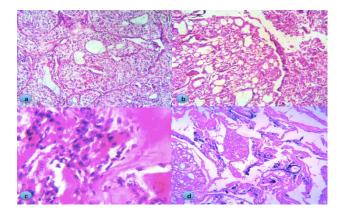


Figure 3: H and E stain (a) insular pattern; (b) glandular pattern; (c) rhabdoid cells and (d)azzopardi phenomenon.

The tumor cells stained strongly positive for thyroglobulin [figure 4] but were negative for Thyroid transcription factor-1 (TTF-1), S-100, pancytokeratin, p53 and calcitonin. Ki-67 was low and positive in only about 20% of the tumor cells. 85% of the tumor cells depicted Bcl-2 expression [figure 5]. We noticed few tumor cells with abundant cytoplasm that stained positive for Vimentin depicting a possibility of 'Rhabdoid' differentiation.

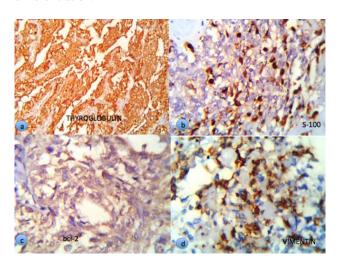


Figure 4: Immunohistochemistry (a) strong positivity for thyroglobulin; (b) Focal positivity for s-100; (c) scattered positivity for Bcl-2; (d) vimentin positivity in rhabdoid cells.

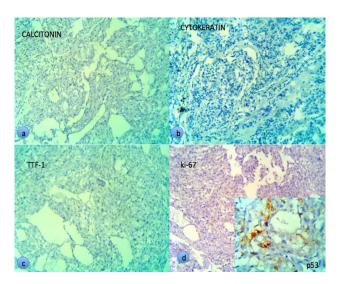


Figure 5: Negative staining (a) calcitonin; (b) cytokeratin; (c) ttf-1; d) ki-67, inset- negative p53.

Thus a final diagnosis of poorly differentiated insular thyroid carcinoma with focal rhabdoid differentiation was made. The patient remains asymptomatic and disease free after 6 months of follow up.

DISCUSSION

Neoplasms derived from the follicular epithelium range from anaplastic to poorly differentiated to well differentiated follicular and papillary carcinoma. Prognosis generally is better for the well differentiated than the poorly differentiated tumors.

Azzopardi phenomenon was first described in lung tumors and further was also documented in various brain tumors. To our knowledge, none has been described in context to poorly differentiated carcinoma of thyroid. It is a phenomenon of DNA in the vessel wall presumably the result of liberation of nucleic acids in large amounts from degenerating neoplastic cells.²

In a case report by Albores Saavedra the presence of rhabdoid differentiation of tumor cells with vimentin cytoplasmic positivity was also seen in our case. ³ Kirdak reported low ki-67 proliferative index which was similar to our findings. ⁴ Although Matias had p53 expression in poorly differentiated and anaplastic areas, the tumor cells did not expressed p53 in our experience. ⁵ Novosel had similar immunohistochemical findings of thyroglobulin, calitoin, and bcl-2. However, focally TTF-1 and cytokeratin positivity was absent in our case. ⁶

Rarely these tumor cells can secrete both thyroglobulin and carcinoembryonic antigen (CEA). While most cases had diffuse cytoplasmic positivity with TG, Begin et al experienced dot-like / prominent inclusion pattern (often paranuclear) which ultrastructurally corresponded to an abortive / rudimentary follicular lumina with abundant periluminal dense bodies, and with varying amounts of colloid and microvilli, both at the intercellular and intracellular levels and secondly, large inculsions reflected altered production and/or storage of thyroglobulin substrate. 8

Cystic component was also seen in poorly differentiated thyroid carcinoma reported by Wu. Thus poorly differentiated thyroid carcinoma should be kept in the differentials for a cystic lesion of thyroid on ultrasonography. Pericapsular and peritumoral papillary endothelial hyperplasia have also been reported. 10

In a case report published by Fadare, the thyroid carcinoma apart from insular pattern of growth, also showed glandular features with clear cell changes. Approximately 30% of the tumor was composed of diffuse glandular formations lined by elongated, simple columnar cells, periodic acid-schiff positive cytoplasm, empty lumens and no myoepithelia or basement membranes. Immunohistochemically, the glandular elements displayed diffuse and strong positivity for TTF-1, bcl-2, sparse positivity for TG and ki-67 and diffuse but weak positivity for p53. Calcitonin was negative throughout the tumor. ¹¹

The tumor cells have also shown to diffusely immunoreact with anti-neuron-specific enolase (NSE), S-100 protein and Leu-7, however did not exhibit nuerosecretory granules ultrastructurally, raising a possibility of aberrant differentiation, for example neural. 12

CONCLUSION

Depth of knowledge about the spectrum of differentiation of thyroid tumors along with extensive immunochemical labeling helps in evaluating and providing extensive insight into the realm of diagnosing such tumors along with emphasis on aberrant expressions seen in these tumors. Thus, thorough evaluation of such cases will help in better patient management and improve healthcare standards.

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