ISSN- O: 2458 - 868X, ISSN–P: 2458 – 8687
National Library of Medicine - ID: 101731606

International Journal of Medical Science and Innovative Research (IJMSIR)
IJMSIR : A Medical Publication Hub
Available Online at: www.ijmsir.com
Volume – 7, Issue – 5, October – 2022 , Page No. : 127 – 131

Medullary carcinoma of thyroid – spindle cell and paraganglioma like variant: A rare entity

[1]Dr. Neoky Suiam, Resident, Department of Pathology, Dr. D Y Patil School of Medicine, Navi Mumbai, India.

[2]Dr. Surekha Bhalekar, M.D. Pathology, Professor, Department of Pathology, Dr. D Y Patil School of Medicine, Navi Mumbai, India.

[3]Dr. Sharad Bhalekar, MS ENT, Consultant ENT, Head and neck Surgeon at Sharda ENT hospital, Nerul west, Navi Mumbai.

[4]Dr. Hemant Bhalekar, M.D. Pathology, Consultant pathologist at Dr.Bhalekar's pathology laboratory, New Panvel, Navi Mumbai, India.

Corresponding Author: Dr. Neoky Suiam, Resident, Department of Pathology, Dr. D Y Patil School of Medicine, Navi Mumbai, India.

Citation this Article: Dr. Neoky Suiam, Dr. Surekha Bhalekar, Sharad Bhalekar, Hemant Bhalekar, "Medullary carcinoma of thyroid – spindle cell and paraganglioma like variant: A rare entity", IJMSIR- October - 2022, Vol – 7, Issue - 5, P. No. 127 – 131.

Type of Publication: Case Report

Conflicts of Interest: Nil

Abstract

A 48-year-old male presented with a midline neck swelling along with pain and difficulty in swallowing since 4 months. FNAC smears revealed adequate cellularity against a plasmaceous background. The tumour cells were spindle shaped and arranged in fascicles as well as singly scattered. Based on cytomorphology, a provisional cyto-diagnosis of benign spindle cell lesion was made. On histopathologic examination, a malignant tumour composed of tumour cells arranged in nests, separated by thin fibrovascular bundles resembling Zell-ballen pattern. Individual tumour cells are medium sized, round to polygonal to spindle shaped with stippled chromatin giving salt and pepper chromatin and granular amphophilic cytoplasm. Focal areas of hyalinized collagen and amyloid also noted. Immunohistochemistry study was done for calcitonin and synaptophysin both of which show strong and diffuse cytoplasmic positivity of tumour cells. Based on histomorphology and IHC a diagnosis of Medullary thyroid carcinoma- spindle cell variant was made. This tumour can mimic benign spindle cell lesions on cytology. Hence the histopathology is considered as gold standard for diagnosis of spindle cell variant of MTC.

Keywords: Fine Needle Aspiration Cytology, Medullary Thyroid Carcinoma, Spindle Cell, Thyroid

Introduction

Medullary thyroid carcinoma (MTC) is a malignant neuroendocrine tumor derived from C cells and accounts for about 10% of all thyroid malignancies [1-5]. Both sporadic and familial forms occur and majority of the cases are sporadic (70%). MCT is found in middle-aged persons with a slight female predominance. There are about 14 histological variants of MTC, and among them, spindle cell is one of the rarest variants. Most common is the classical pattern displaying sheets of round to

polygonal cells with neuroendocrine features, separated by hyalinised fibrous stroma and sometimes containing amyloid [2]

Here, we have a case where on fine needle aspiration cytology from midline neck swelling (thyroid) revealed clusters as well as singly scattered benign spindle cells. Based on cytomorphology, a diagnosis of benign spindle cell lesion was made.

But on histopathology examination a diagnosis of Medullary thyroid carcinoma – spindle cell variant was made. This tumour can mimic benign spindle cell lesions like schwannoma on FNAC [2].

Hence, a very careful assessment is required on FNAC.

Case report

A 48-year-old male presented with a midline neck swelling along with pain and difficulty in swallowing since 4 months.

Ultrasonography revealed a heterogenous soft tissue mass (74 x 68 x 47 mm) in the right supraclavicular region suggestive of mesenchymal tumour of neurogenic origin.

FNAC from the neck swelling was done. The air-dried smears were stained by May-Grunwald Giemsa (MGG) stain and alcohol-fixed smears were stained by Papanicolaou (Pap) stain.

FNAC smears revealed adequate cellularity against a plasmaceous background. The tumour cells were spindle shaped and arranged in fascicles as well as singly scattered. Based on cytomorphology, a provisional cytodiagnosis of benign spindle cell lesion was made. A histopathologic examination was advised for confirmation.

A right hemithyroidectomy was done. Gross (Figure 2): External surface of the thyroid gland shows a well demarcated tumour with intact thyroid capsule measuring 8 x 6.5 x 6 cm. Cut section shows a solid, firm, encapsulated tumour, relatively well- circumscribed, grey to yellowish in colour with areas of hemorrhage and cystic areas.

On histopathologic examination, a malignant tumour composed of tumour cells arranged in nests, separated by thin fibrovascular bundles resembling Zell-ballen pattern. Individual tumour cells are medium sized, round to polygonal to spindle shaped with stippled chromatin giving salt and pepper chromatin and granular amphophilic cytoplasm. Focal areas of hyalinized collagen and amyloid also noted. Immunohistochemistry study was done for calcitonin and synaptophysin both of which show strong and diffuse cytoplasmic positivity of tumour cells. Based on histomorphology and IHC a diagnosis of Medullary thyroid carcinoma- spindle cell variant was made.

Discussion

Spindle cell variant of MTC is extremely rare and only 5 cases have been reported in thus far [1].

MTC is a neuroendocrine tumor, accounting for 10% of all thyroid malignant tumors [1-5]. The patients with spindle cell variant of MTC generally have no specific clinical symptoms, only presenting with a painless thyroid mass, sometimes presenting with intractable diarrhea, facial flushing or other neuroendocrine symptoms and these symptoms are consistent with that of MTC. Also, serum calcitonin (sCT) is the most sensitive and specific marker for preoperative diagnosis of MTC, and serum CEA is synchronous with sCT in some MTC patients, which is a good reference value for diagnosis and prognosis of the disease. When the tumor causes compression or invasion to the surrounding tissues, the above mentioned clinical symptoms may appear, and lymph node metastasis can occur in the early stage [1]. Familial cases are associated with extrathyroidal findings like hyperparathyroidism, symptoms of

pheochromocytoma, pituitary, pancreatic dysfunctions & mucosal neuromas [2].

Both sporadic and familial forms can occur and majority of the cases are sporadic (70%) [3]. This tumor is also associated with other endocrine disorders such as pheochromocytoma, parathyroid hyperplasia and as part of multiple endocrine neoplasia syndrome (MEN) [3]. The histological diagnosis of MTC depends on its cytological feature, growth pattern, amyloid deposition, as well as its immunophenotype [3]. MTC with predominant spindle cell morphology is extremely rare. The spindle cell variant of MTC consists of irregularly arranged bundles or interlaced spindle cells in which is separated by fibrous stroma or amyloid although amyloid deposition is not a necessary criteria for the diagnosis of MTC as amyloid may not always be present in atypical cases. The tumor cells are short or long with spindled nuclei, abundant cytoplasm, inconspicuous nucleoli and rare mitoses, with clear demarcation from surrounding tissues [1]. Immunohistochemically, the most sensitive markers for the tumour are Calcitonin, although it is not specific because it also can be found in endocrine tumors of non-thyroid origin, such as islet cell tumors and intestinal neuroendocrine carcinomas. In addition, MTC cells also are immunoreactive for cytokeratins, neuron-specific enolase, synaptophysin, CEA, and chromogranin A, whereas they are not stained by Thyroglobulin [1]. In our case, the tumour showed spindle cell morphology, with positive expressions of calcitonin, synaptophysin and amyloid deposition, confirming the diagnosis as spindle cell variant MTC.

Spindle cell variant of MTC can mimic some spindle cell mesenchymal tumours occurring in the thyroid gland, such as leiomyoma, peripheral schwannoma, and spindle cell melanoma. These mesenchymal tumours are negative for neuroendocrine markers. Therefore, the morphology in conjunction with immunohistochemistry is sufficient to make a diagnosis of spindle cell variant of MTC [1].

Conclusion

In conclusion, spindle cell variant MTC is an extremely rare histological variant and a mimicker of other mesenchymal tumours in the thyroid gland [1]. Therefore, a careful assessment in FNAC, histological morphology examination and immunohistochemical staining with Calcitonin , chromogranin A, synaptophysin, CD56, and TTF-1 is required for accurate diagnosis.

Legend Figure

Fig 1: PAP, 40x: FNAC smears revealed tumour cells which were spindle shaped and arranged in fascicles as well as singly scattered.

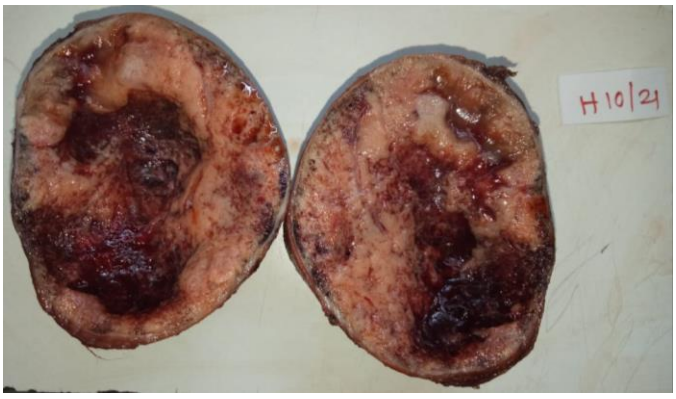

Fig 2: Excised lesion on gross.

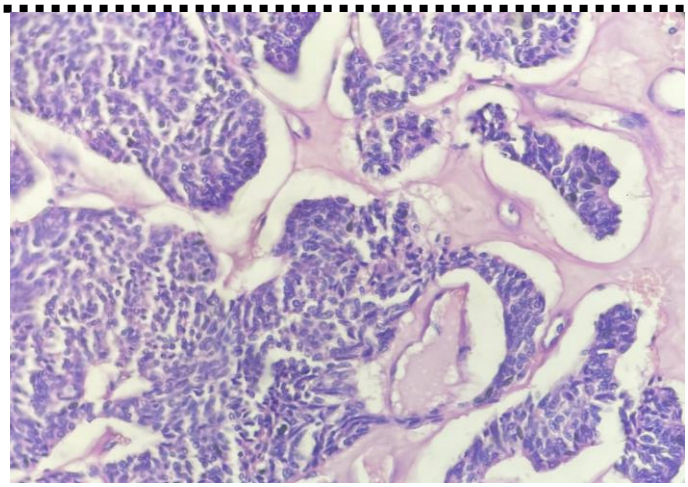

Fig 3: H&E, 40x: Individual tumour cells are medium sized, round to polygonal to spindle shaped with stippled chromatin giving salt and pepper chromatin and granular amphophilic cytoplasm. Focal areas of hyalinized collagen and amyloid also noted.

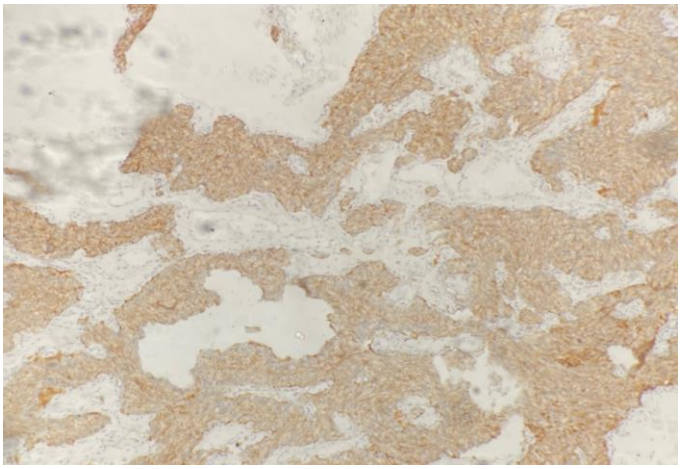

Fig 4: IHC,4x: IHC for Synaptophysin

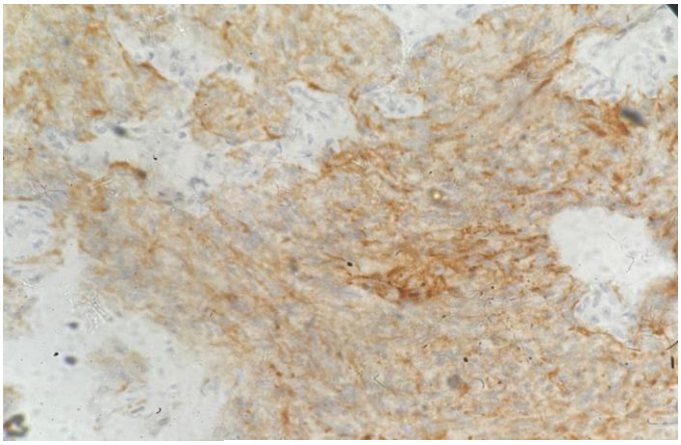

Fig 5: IHC,40x: Immunohistochemistry synaptophysin show strong and diffuse cytoplasmic positivity of tumour cells.

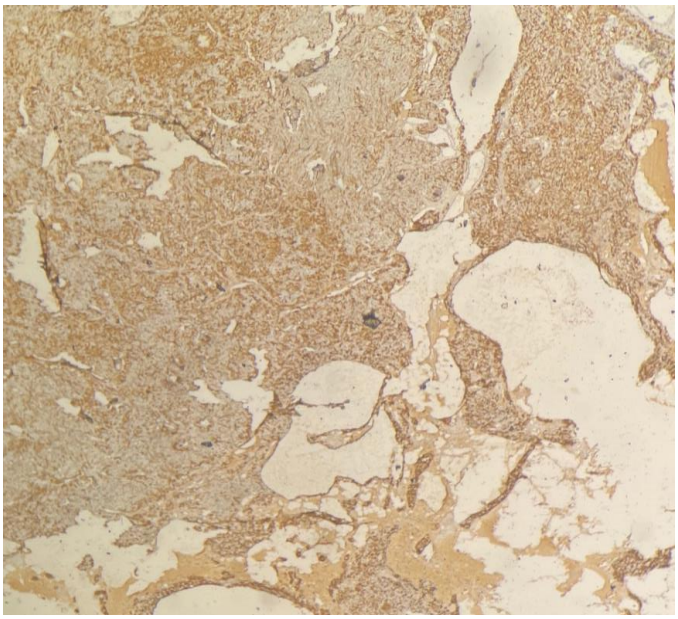

Fig 6: IHC, 4x: Calcitonin

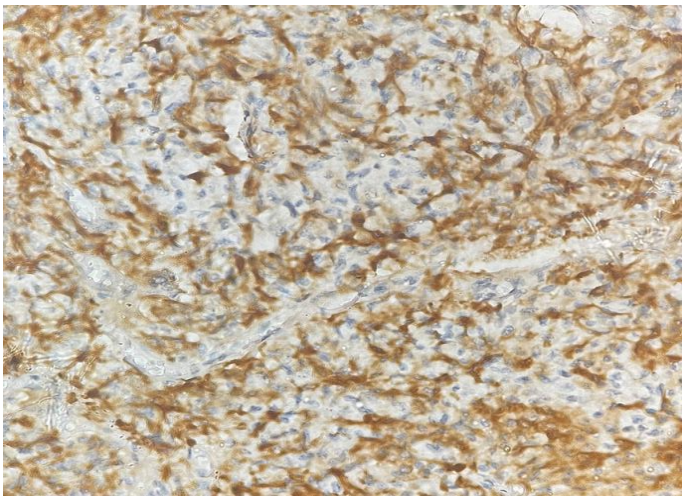

Fig 7: IHC, 40x: Immunohistochemistry calcitonin show strong and diffuse cytoplasmic positivity of tumour cells.

References

1. Wang, Y.X., Yang, S.J. Spindle cell variant of medullary thyroid carcinoma: a clinicopathologic study of four cases. *Diagn Pathol* **16**, 112 (2021).
2. Das A, Nargotra N, Singh S, Deepak RKTyagi I. Rare presentation of medullary carcinoma of thyroid with predominant spindle cell pattern & abundant

calcification. Annals of Pathology and Laboratory Medicine. 2016;03:C130–C134.

3. Mondal SK, Nag DR, Bandyopadhay R. Spindle cell variant of Medullary Carcinoma of thyroid with nodal metastasis: Cytodiagnosis and diagnostic dilemma. Thyroid Res Pract 2012;9:60.
4. Sophie Leboulleux, Eric Baudin, Jean-Paul Travagli and Martin Schlumberger Institut Gustave Roussy, Villejuif, France (Received 31 October 2003; returned for revision 15 December 2003; finally revised 24 December 2003; accepted 3 March 2004)
5. Verma A, Kane S, Vinarkar S, D'Cruz AK. Small cell medullary thyroid carcinoma: A diagnostic dilemma. Indian J Pathol Microbiol 2017;60:562-4
6. Laforga JB, Aranda FI. Pseudoangiosarcomatous features in medullary thyroid carcinoma spindle-cell variant. Report of a case studied by FNA and immunohistochemistry. Diagn Cytopathol. 2007 Jul;35(7):424-8. doi: 10.1002/dc.20656. PMID: 17580345.