Case Report

Rare Presentation of Medullary Carcinoma of Thyroid with Predominant Spindle Cell Pattern & abundant Calcification

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ABSTRACT

Medullary carcinoma thyroid (MTC) is a malignant tumour showing C-cell differentiation. It accounts for 10% of all thyroid malignancies. Many histological variants have been described in literatures. Among them most predominant one is the classical variant, comprising almost one third of all. Predominantly spindle cell pattern is seen very rarely. MTCs are mostly seen in middle aged patient. It is rarely seen in second decade except familial type. Here we present a case of MTC with predominant spindle cell morphology & abundant calcification in a 19 years old female patient.

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Introduction

Thyroid cancers account for 1% of all cancer cases, whereas MTS comprises of 10% of all thyroid malignancies.[1,2] It occurs in all age groups, mostly seen in middle aged patient with a female predominance. It is seen less commonly in second decade of life. Though MTC is a rare tumour, it shows wide varieties of morphological patterns.[3] Most common pattern encountered is the classical pattern where it shows sheets of round to polygonal cells with neuroendocrine features, separated by hyalinised fibrous stroma sometimes containing amyloid. Pure spindle cell pattern occurs less commonly.[4] Calcification in MTCs is also seen rarely.[4] Here we report a case of MTC in a younger female patient in which we showed predominantly spindle cell pattern along with ample amount of calcification both cytologically & histologically.

Case Report

A 19 years old female patient presented with gradually increasing, tender, midline neck swelling of 2 years duration, which was initially small in size. On clinical examination, patient was conscious & oriented. Swelling was diffuse & firm, moving with deglutition. Family history was negative. Biochemical examination showed mild raised calcitonin level (8.9 mg/dl) & normal serum parathormone level. Thyroid function test was within normal range. Computed tomography of neck & FNAC were advised. Imaging revealed a thyroid mass. Cytosmears showed predominantly spindle cell population arranged in groups as well as singly scattered, mostly monomorphic spindle to oval nuclei showed granular type of chromatin (Fig 1, 2). Focally amyloid material was also noted. On cytology medullary carcinoma of thyroid was suggested. Near total thyroidectomy was performed & the specimen was sent for histopathological examination. Grossly thyroid gland was slightly enlarged (left lobe- 6x4x3.5 cm, right lobe- 6.5x4x3.5 cm & isthmus- 2x1.5x1 cm). Externally capsule was intact. Cut sections of both lobes showed a bilateral, multifocal, circumscribed gray white tumour measuring 3.5x2x2 cm in left lobe & 3.5x2.5x2 cm in right lobe (Fig 3). On histological examination, tumour showed predominantly monomorphic spindle cell pattern, arranged in small nests, sheets as well as in short fascicles, separated by thin fibrovascular septa. Tumour cells showed oval to spindle nuclei with granular chromatin & moderate amount of granular eosinophilic cytoplasm (Fig 4, 5). Abundant eosinophilic amyloid material & large areas of calcification were also identified (Fig 6 & 7). Rest of the thyroid parenchyma showed adenomatous changes. C-cell hyperplasia, vascular invasion were not seen. Tumour cells were strongly positive for calcitonin & carcinoembryonic antigen (Fig 8, 9). Final diagnosis of medullary carcinoma of thyroid with abundant calcification was given. After a follow up of 5 months, patient is doing well & completely asymptomatic.

Discussion

Medullary carcinoma of thyroid was first described by Hazard et al.[5] It accounts for 10% of all thyroid malignancies.[1,6] Around 20% of MTCs are associated with autosomal dominant inherited MEN (multiple endocrine neoplasia) syndromes (specially MEN 2A & 2B), while rest 80% cases are sporadic. MTCs affect patients of wide age ranges. Familial MTCs occur at an earlier age, while sporadic MTCs at a later stage. Patients usually present with a painless thyroid mass, which is mostly bilateral or multicentric in familial cases & unilateral in sporadic cases.[7] Our patient was 19 years old, presented with gradually

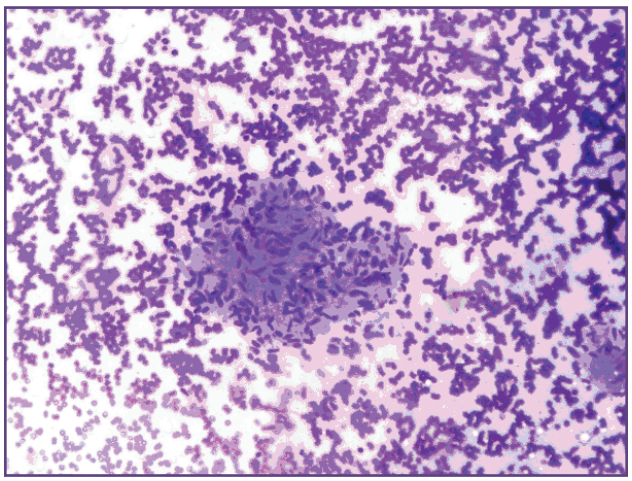

Fig. 1: Cytosmear shows predominantly spindle cell population arranged in groups as well as singly scattered (Giemsa, 40X).

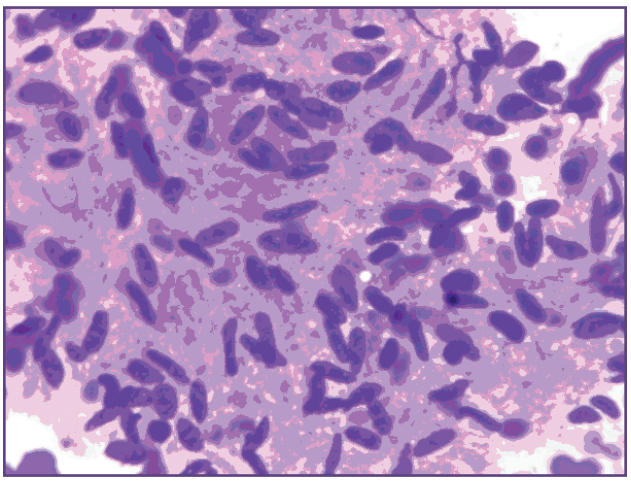

Fig 2: Cytosmear shows monomorphic spindle to oval nuclei with granular type of chromatin (Giemsa, 400X).

Fig. 3: Cut sections of both lobes showed a bilateral, circumscribed gray white tumour.

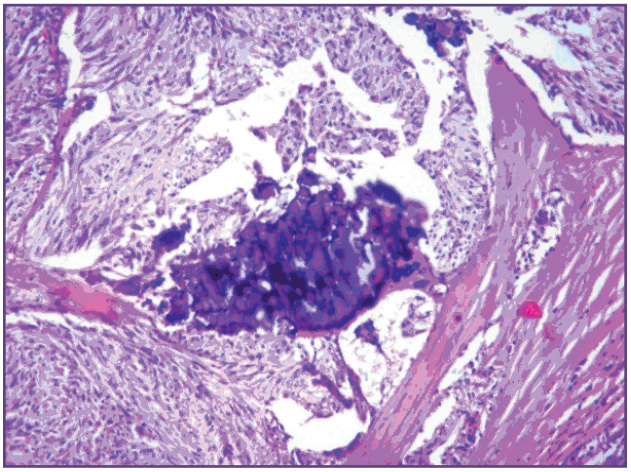

Fig 6: Microsection showing large areas of calcification (H&E, 100X).

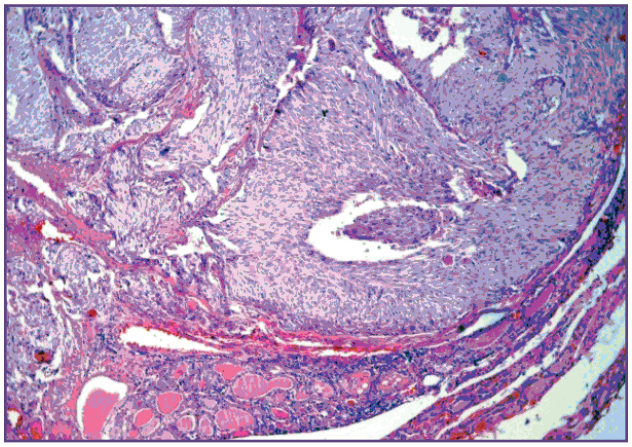

Fig. 4: Tumour cells showed oval to spindle nuclei with granular chromatin & moderate amount of granular eosinophilic cytoplasm (H&E, 100X).

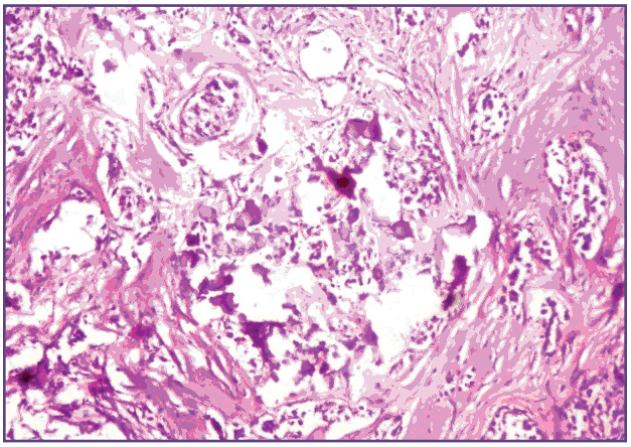

Fig 7: Abundant eosinophilic amyloid material & areas of calcification (H&E, 200X).

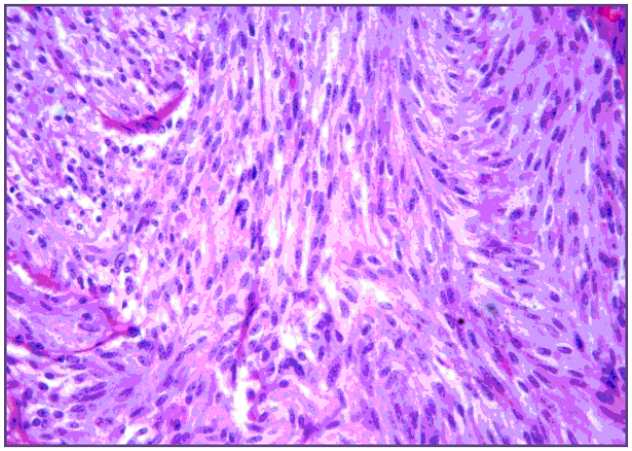

Fig 5: Tumour cells showed oval to spindle nuclei with granular chromatin & moderate amount of granular eosinophilic cytoplasm (H&E, 400X).

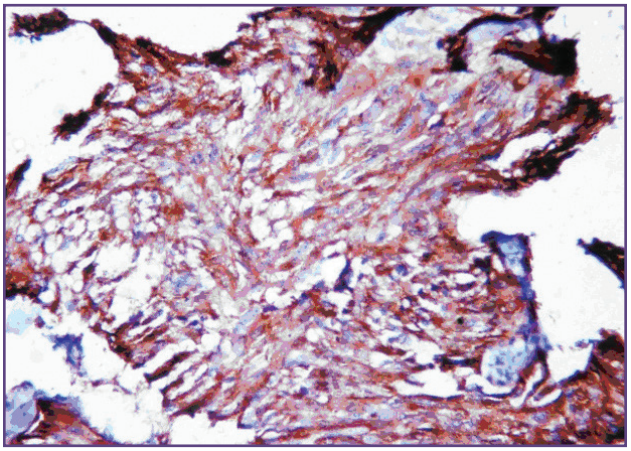

Fig 8: Tumour cells were strongly positive for calcitonin (DAB, 400X).

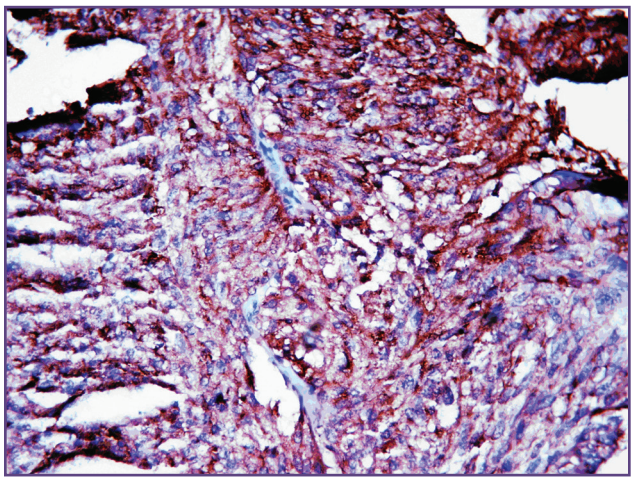

Fig 9: Tumour cells were strongly positive for carcinoembryonic antigen (DAB, 400X).

increasing tender thyroid mass which was multicentric & showed bilateral lobes involvement. About 50% of the MTC patients present with cervical lymphadenopathy at the time of presentation.[7,8] In present case the patient did not show any lymph nodal involvement. Familial cases are associated with extrathyroidal findings like hyperparathyroidism, symptoms of pheochromocytoma, pituitary, pancreatic dysfunctions & mucosal neuromas.[8,9] There were no such findings in our patient.

In spite of rarity various morphological patterns described in literatures are classical, pseudopapillary, small cell, microglandular, cribriform, follicular, rosette like, oncocytic, osteosarcoma like, cystic, whorled & encapsulated. Among these, classical pattern accounts for almost 1/3rd of all. It shows sheets of cells having round to oval nuclei with salt & pepper like chromatin, separated by hyalinised fibrous septa of variable thickness containing amyloid.[4,10] MTC with predominant spindle cell morphology is seen less commonly. However studies showed that these morphological patterns did not have any significance differences in tumour prognosis.[4]

Histological criteria for diagnosis of MTCs depend on growth pattern, cytological features of a neuroendocrine tumor & amyloid deposition. Immunohistochemistry is used for confirmation.[4] Most of the literatures mentioned that MTCs are more common in women as compared to men, mostly occurring at middle age.[8,9,11] One study from India showed male predominance with a male to female ratio of 1:0.45.[4] Sporadic tumour presentation in second decade of life is very rare. Our 19 years old female patient presented with neck swelling of 2 years duration. MTCs are usually well defined tumor, often showing lymph node involvement, vascular invasion & extrathyroidal extension.[4,7] In our case, the tumour was well circumscribed & did not show any lymph nodal involvement, vascular invasion or any other extrathyroidal extension. Desai et al mentioned that around 19% of MTCs showed presence of calcification.[4] Another study also mentioned about calcification, but none of these studies have described about the relative amount of calcification. We found abnormally large amount of calcification in our case which we think an unusual presentation.

Conclusion

In conclusion, predominant spindle cell pattern with abundant calcification in medullary thyroid carcinoma in a younger asymptomatic patient is a rare presentation.

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Competing Interests

None declared.

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