

## Low-Grade Fibromyxoid Sarcoma of the Thyroid: A Case Report

### Dear Editor,

Low-grade fibromyxoid sarcoma (LGFMS) is a rare form of neoplasm that usually occurs in the deep soft tissue of the lower limbs with a tendency towards local recurrence and distant metastasis. LGFMS affects mainly the young and middle-aged, with both gender having an equal chance of being affected. To the best of our knowledge, there is only 1 case of thyroid LGFMS reported in the literature, limited to English language. Here we describe another case of thyroid LGFMS with giant rosettes confirmed by pathology.

### Case Report

A 65-year-old man presented with a 6-month history of a rapidly increasing painless mass in the neck. The patient did not complain of any dysphagia or dyspnoea. He underwent bilateral subtotal thyroidectomy 15 years ago and unilateral subtotal thyroidectomy (right lobe) 18 months ago at another hospital. Pathologic findings revealed the presence of a goiter. The patient did not have a history of radiotherapy or chemotherapy. Physical examination revealed a 4 × 3 cm mass on the left lobe of the thyroid. The mass was firm in consistency, non-tender, with a smooth surface and was movable during swallowing. No cervical lymph node was palpable. Thyroid studies revealed a normal triiodothyronine (FT3), thyroxine (FT4), thyroid stimulating hormone (TSH), thyroid peroxidase antibodies (TPOAb) and thyroglobulin antibodies (TGAb). A neck ultrasound confirmed the presence of a 33.8 × 24.6 mm thyroid lesion with a regular margin in the left lobe of the thyroid. A unilateral subtotal thyroidectomy (left lobe) was performed. Intraoperative frozen section examination suggested malignant change of nodular goiter in the left lobe of the thyroid. Therefore, a total thyroidectomy with a prophylactic central compartment dissection was performed.

Grossly, the resected specimen measured about 3 cm in the greatest dimension. It was well demarcated and greyish-tan on the cut surface, but not very solid in the texture. Microscopically, the tumour showed an admixture of hypocellular myxoid and hypercellular spindle cell areas in a collagenous stroma with arcades of blood vessels. Short fascicular and whorled growth patterns were seen, with the latter often most apparent at the transition from collagenous to myxoid areas (Fig. 1). The central area of the giant rosettes was formed by collagen fibres. The surrounding cells were more crowded, round or ovoid, and

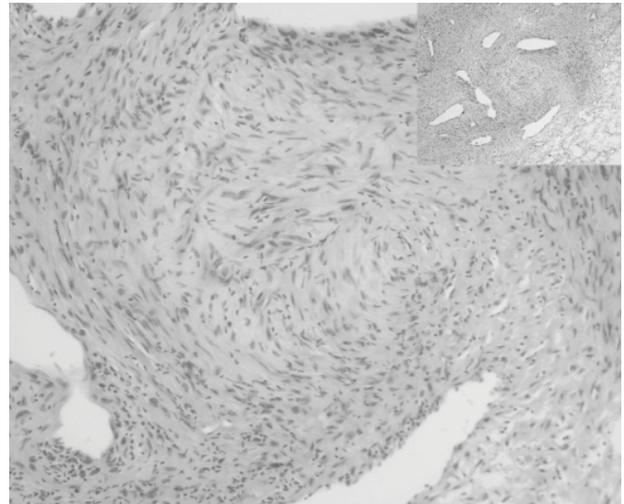


Fig. 1. The tumour showed an admixture of hypocellular myxoid and hypercellular spindle cell areas in a collagenous stroma with arcades of blood vessels. The tumour cells are bland spindled cells with a fibroblast pattern that are arranged in a short fascicular or whorled growth pattern (Haematoxylin and Eosin, × 200; inset, Haematoxylin and Eosin, × 100).

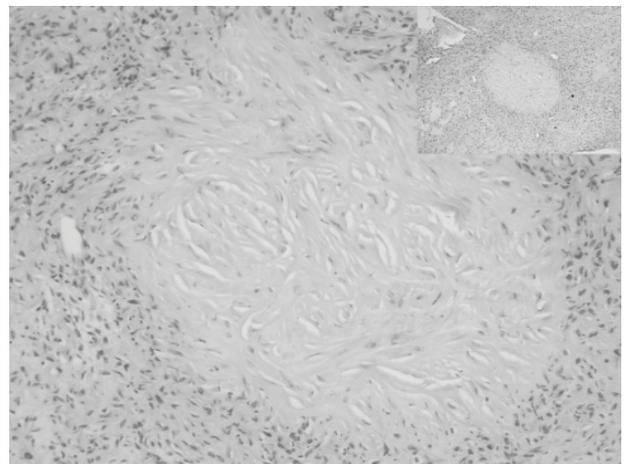


Fig. 2. The central area of the giant rosettes are formed by collagen fibers. The surrounding cells are more crowded, round or ovoid, and blend imperceptibly with the surrounding spindle-shaped cells (Haematoxylin and Eosin, × 200; inset, Haematoxylin and Eosin, × 100).

blend imperceptibly with the surrounding spindle-shaped cells (Fig. 2). Immunohistochemical stains were positive for vimentin, P53, bcl-2 and smooth muscle actin and negative for CD99, cytokeratin (CK)-19, galectin-3, and calcitonin. The thyroid cells within the tumour were positive for CK suggesting that the tumour originated from the thyroid. The

arcades of blood vessels were positive for CD34, but the tumour cells were negative. Based on these results, the final histopathological diagnosis was LGFMS. Postoperatively, the patient received radiation therapy with 56 Gy in 28 portions for 28 days. The patient complained of lower back pain at 14 months after the operation. Conventional and contrast-enhanced magnetic resonance imaging (MRI) scan showed that the tumour had metastasised to the thoracic vertebra. After the metastatic lesions were completely resected, the pathologic findings were LGFMS. The patient recovered with no remission for the 11 months after surgery.

## Discussion

LGFMS is a rare form of neoplasm that usually occurs in the deep soft tissue of the lower limbs with a tendency towards local recurrence and distant metastasis. It affects mainly the young and middle-aged, with both men and women having equal chances of being affected. LGFMS of the thyroid is extremely rare. In fact, only 1 case has been reported previously in literature, limited to English language. Merchant<sup>1</sup> reported on a 57-year-old male with LGFMS located in the left thyroid lobe. A surgical resection of the mass and complete thyroidectomy were performed after ultrasound-guided fine needle aspirations. No chemotherapy or radiotherapy was administered at that point. At 13 months postoperatively, the patient had local recurrence of the tumour which was excised. The patient also received intensity-modulated radiation therapy and was free of clinical/imaging evidence of recurrent/metastatic disease within 8 months.

The diagnosis of LGFMS of the thyroid is usually not difficult if the tumour has been removed completely and all the characteristic morphologic and immune-phenotypic features are present. Cytogenetic and/or molecular genetic analyses could be used as an ancillary diagnostic tool for LGFMS. Specifically, FUS/CREB3L2, which is specific for LGFMS, could be used as a diagnostic marker for difficult cases with unusual histologic features.<sup>2-5</sup> However, it is difficult to make a preoperative diagnosis of LGFMS, because imaging examination and fine needle aspiration biopsy are non-specific.

The choice of treatment of LGFMS of the thyroid is wide surgical resection. The roles of radiotherapy and chemotherapy remain unspecified, because of the rarity of LGFMS of the thyroid. Long-term follow-up is recommended even after a successful resection, because LGFMS is known to be associated with local recurrence and distant metastasis. The patient reported by Merchant<sup>1</sup> had local recurrence and our patient had distant metastasis after surgery.

## Conclusion

LGFMS of the thyroid is a rare form of tumour. Early recognition and complete eradication of the tumour may offer a better prognosis. The choice of treatment for LGFMS of the thyroid is wide surgical resection. Although traditional adjuvant radiotherapy and chemotherapy have an uncertain curative effect, newer chemotherapeutic and possibly biologic agents should be tested in order to find an effective way to control the disease. Long-term follow-up is recommended because of the relatively high rate of recurrence and distant metastasis.

## REFERENCES

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