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Thyroid Leiomyosarcoma: A Rare Case Report

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ABSTRACT

Background
Primary thyroid leiomyosarcoma is a very rare tumor. The incidence is less than 0.01% of all primary thyroid tumors. Thyroid leiomyosarcoma are tumours showing smooth muscle differentiation, originating from thyroid gland vascular smooth muscle.

Case Description
Female, 47 years old, with lump on the left neck since 1 year ago, getting bigger in the last 3 months, no shortness of breath, no history of previous surgery. Physical examination revealed a mass was 13x6 cm in size, solid, firm. Cytological features from fine needle aspiration biopsy (FNAB) showed cellular spindle cells with atypical, pleomorphic, hyperchromatic nuclei and several normal thyroid follicles among them, sugesting malignant thyroid tumor (Bethesda VI). Total thyroidectomy was performed, followed by histopathological examination. Microscopically showed proliferation of atypical, pleomorphic, hyperchromatic spindle cells with eosinophilic cytoplasm. The spindle cells arranged in fascicular and interlacing pattern. Several follicular thyroid are entrapped within this atypical spindle cells. Based on these finding, this case was diagnosed as anaplastic thyroid carcinoma. To confirm the diagnosis, immunohistochemical examination with cytokeratin, desmin and Ki-67 were performed and the result showed negative for cytokeratin, positive for desmin and Ki-67 was highly positive. Based on these results, a diagnosis of thyroid leiomyosarcoma was made.

Discussion and Conclusion
Differential diagnosis with other primary mesenchymal thyroid tumors should be considered in the cases with atypical spindle cells microscopic appearance. The diagnosis of thyroid leiomyosarcoma based on cytology and histopathology is difficult to establish, therefore immunohistochemical staining is required to establish the accurate final diagnosis.

Keywords: Thyroid, Leiomyosarcoma, Immunohistochemistry

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INTRODUCTION

Malignancies of the thyroid gland is one of the most common neoplasms and mainly affects women[1]. Rare types include anaplastic carcinomas, sarcomas and lymphomas. Sarcomas are a very rare group of tumors. The types of sarcomas observed in the thyroid gland are liposarcoma, leiomyosarcoma, and angiosarcoma[2]. Primary thyroid leiomyosarcoma is a very rare tumor. The incidence is less than 0.014% of all primary thyroid tumors[3]. Thyroid leiomyosarcoma are tumours showing smooth muscle differentiation, originating from thyroid gland vascular smooth muscle[4].

CASE DESCRIPTION

Female, 47 years old, with lump on the left neck since 1 year ago, getting bigger in the last 3 months, no shortness of breath, no history of previous surgery. Physical Examination revealed a mass measuring 13x6 cm, gray-brown in color, solid and hard in consistency, solid white in cross section. FNAB smear showed elongated spindle shaped cells with atypical nuclei and several normal thyroid follicles in between (Figure 1), suggesting a malignant thyroid tumor (Bethesda VI).

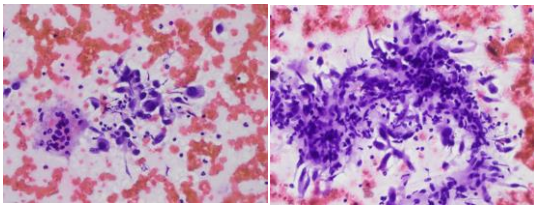

Figure 1: FNAB Cytological Smear showed elongated spindle shaped cells with atypical nuclei and several normal thyroid follicles.

Total thyroidectomy surgery has been performed and gross examination showed mass measure 13x11x11,5 cm, on cut section: solid, light tan and fleshy with zones of necrosis. Microscopically showed solid tumor massed (Figure 2A) with proliferation of elongated spindle-shaped cells arranged in fascicular and interlacing pattern with eosinophilic cytoplasm and hyperchromatic nuclei (Figure 2B). Several follicular thyroid are entrapped within this atypical spindle cells (Figure 2C). Diagnosed as anaplastic thyroid carcinoma. For a definite diagnosis, cytokeratin, vimentin and desmin immunohistochemistry are required.

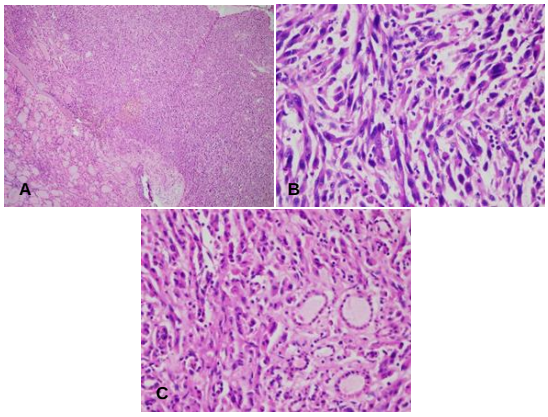

Figure 2. A. Showed solid tumor massed (HE 4 times). B. Showed proliferation of elongated spindle-shaped cells arranged in fascicular and interlacing pattern (HE, 40 times). C. Showed follicular thyroid are entrapped within this atypical spindle cells.

Immunohistochemical examination revealed cytokeratin: negative (Figure 5), vimentin: positive (Figure 6), desmin: positive (Figure 7) and Ki-67: highly positive (Figure 8).

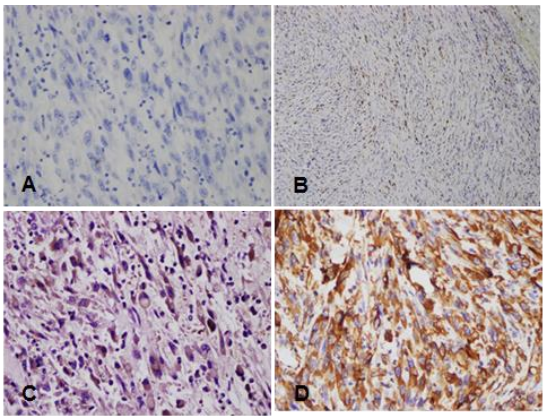

Figure 3. A. Negative stain in Cytokeratin imunohystochemistry. B. Positive stain in Vimentin imunohystochemistry. C. Positive stain in Desmin imunohystochemistry. D. Positive stain in Desmin imunohystochemistry.

DISCUSSION

Primary thyroid leiomyosarcoma is the second most common primary thyroid sarcoma after angiosarcoma[4]. Primary thyroid LMS most commonly presents as a painless, rapidly-growing neck mass. Primary thyroid LMS most often presents as a rapidly growing, painless neck mass. Additional symptoms may include hoarseness, dysphagia, dyspnoea and weight loss. The etiology of this tumor is not clear; however, it is thought to develop from the smooth muscle of the capsular veins in the thyroid gland[1].

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This tumor seems to have a slight predilection for female patients and the mean age at diagnosis, including our case, is 63.4 years, with a range between 32 and 90 years. additionally, a case of epstein-Barr virus-associated primary thyroid LMS was reported in a 6-year-old child patient with congenital immunodeficiency[5].

An accurate diagnosis is difficult, The majority of patients are euthyroid[3]. Ultrasound can show an ill-defined or well-defined hypo-echogenic mass with calcified or cystic components[6]. Thyroid isotope scanning can reveal a cold nodule or hyperplasia with increased and decreased uptake of radioactive iodine[3]. CT scan usually shows a low density mass with calcification and necrosis and moreover, can reveal a direct tumoral invasion of the adjacent structures[7].

On tissue sample, partrculary FNAB. The main differential diagnosis includes anaplastic thyroid carcinoma, spindle cell variant of medullary carcinoma, solitary fibrous tumor. The final diagnosis requires immunohistochemical investigations.
Immunohistochemistry plays a key role in the diagnosis[5]. On Immunohistochemistry, positivity for smooth muscle actin, vimentin, desmin and caldesmon may diagnose thyroid leiomyosarcoma, while these tumour cells don't react to keratin, thyroglobulin, chromogranin, and calcitonin. This tumor has poor prognosis and without any established treatment[1].

In our experience, although radical surgery was performed, a local recurrence was detected after only 2 months of follow-up. To date, chemotherapy and radiotherapy has not shown any substantial therapeutic efficacy[5].

CONCLUSIONS

Differential diagnosis with other primary mesenchymal thyroid tumours should be considered in the cases with atypical spindle cells microscopic appearance. The diagnosis of thyroid leiomyosarcoma based on cytology and histopathology is difficult to establish, therefore immunohistochemical staining is required to establish the accurate final diagnosis.

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