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Case report: a grievous tale of a rare primary thyroid leiomyosarcoma

Wan Nabila Wan Mansor[1*], Norhaslinda Abdul Gani[1], Khairul Azlan Shahril Abu Dahari[1], Aliza Ahmad[2] and Rohaizam Japar Jaafar[3]

Abstract

Background: Primary thyroid leiomysosarcoma is a rare and aggressive disease which represents 0.014% of primary thyroid cancers. It confers a diagnostic conundrum, due to difficulty to make a preoperative diagnosis of thyroid leiomyosarcoma and to differentiate it from anaplastic thyroid carcinoma.

Case presentation: We herein report a rare case of primary thyroid leiomyosarcoma. A 48-year-old male presented with history of anterior neck swelling for the past 2 months. Computed tomography (CT) neck showed hypodense nodule at the right thyroid lobe with calcification causing tracheal deviation, with intratracheal extension at 1st and 2nd tracheal rings and presence of lung metastases. The histopathology analysis yield high grade spindle cell neoplasm suggestive of sarcoma. Immunohistochemistry showed that neoplastic spindle cells were positive for *smooth muscle actin*, *vimentin*, *caldesmon* while no reactivity was reported for thyroglobulin, calcitonin, thyroid transcription factor-1, S-100, CD45, CD 5, and cytokeratins. Patient had undergone tracheostomy and chemotherapy but he succumbed to death 1 month after completion of chemotherapy.

Conclusion: Primary thyroid leiomyoarcoma is a very rare and aggressive disease. The treatment ranging from surgery, adjuvant radiotherapy and chemotherapy, with high recurrence rates.

Keywords: Thyroid, Leiomyosarcoma, Aggressive

Background

Thyroid can give rise to different types of malignancy, whereby primary cancer is the most common type [1]. World Health Organization (WHO)'s histological classification of thyroid and parathyroid tumors divides smooth muscle tumors into benign (leiomyoma) or malignant (leiomyosarcoma) [2]. Thyroid leiomysosarcoma is very rare and represents 0.014% of primary thyroid cancers. It confers a diagnostic conundrum, due to difficulty to make a preoperative diagnosis of thyroid leiomyosarcoma and differentiate it from anaplastic thyroid carcinoma [3]. The prognosis is poor in spite of aggressive surgery, adjuvant radiotherapy and chemotherapy, with high recurrence rates and an estimated 1-year survival of only 5–10% [4]. Few have reported on this case. In this report, one patient with primary thyroid leiomyosarcoma presented with review of literature.

Case presentation

A 48-year-old male presented with anterior neck swelling for the past 2 months, which rapidly increasing in size for 2 weeks (Fig. 1). He developed obstructive symptoms such as shortness of breath, noisy breathing and reduced effort tolerance. He experienced hoarseness with no aspiration symptoms. There was no symptoms of hyper- or hypothyroidism. He was initially intubated and ventilated for 3 days due to respiratory distress. Endoscopic laryngeal assessment revealed right vocal cord adductor palsy at lateral position. Subsequently, contrast-enhanced computed tomography

*Correspondence: wannabila.wanmansor@gmail.com

[1] Hospital Tuanku Jaafar Seremban, Seremban, Negeri Sembilan, Malaysia
Full list of author information is available at the end of the article

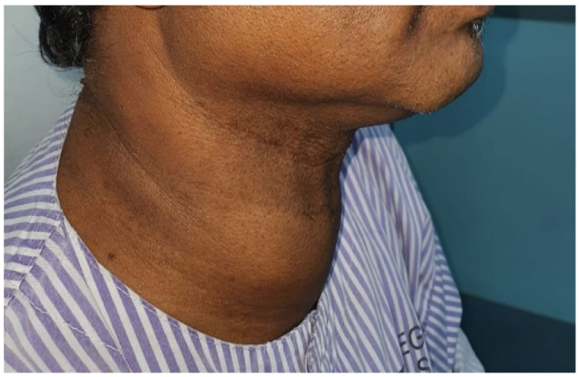

Fig. 1 Anterior neck swelling, appears to be thyroid origin

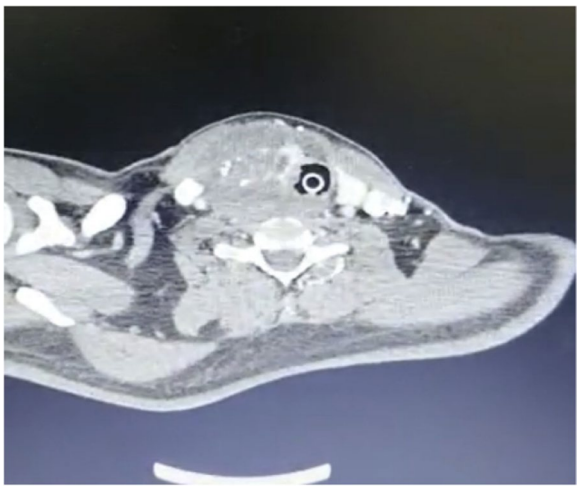

Fig. 2 Axial CT scan showed heterogenous mass with area of calcification arise from right thyroid lobe

(CECT) neck and thorax and ultrasound guided *tru-cut biopsy* of the right thyroid nodule were performed.

CECT showed hypodense nodule at the right thyroid lobe with calcification measuring 7 cm × 4.8 cm × 8.2 cm, causing tracheal deviation, with intratracheal extension at 1st and 2nd tracheal rings and no clear plane with prevertebral muscles (Fig. 2). The scan showed presence of lung metastases.

The histopathology analysis yield high grade spindle cell neoplasm suggestive of sarcoma (Fig. 3). Immunohistochemistry showed that neoplastic spindle cells were positive for *smooth muscle actin* (Fig. 4), *vimentin, caldesmon* (Fig. 5) while no reactivity was reported for *thyroglobulin*, *calcitonin*, *thyroid transcription factor-1*, *S-100*, *CD45, CD 5*, and *cytokeratins*.

Subsequently, he had undergone tracheostomy. Intraoperative findings revealed the tumor arising from the

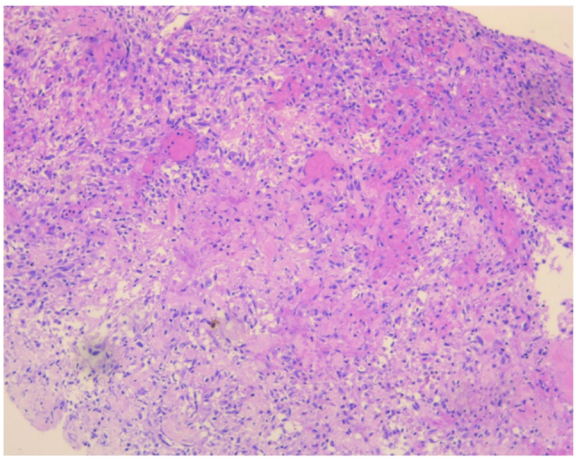

Fig. 3 Moderately pleomorphic spindle cells arranged in storiform and fascicles with area of necrosis (*haematoxylin eosin* (H&E) × 20)

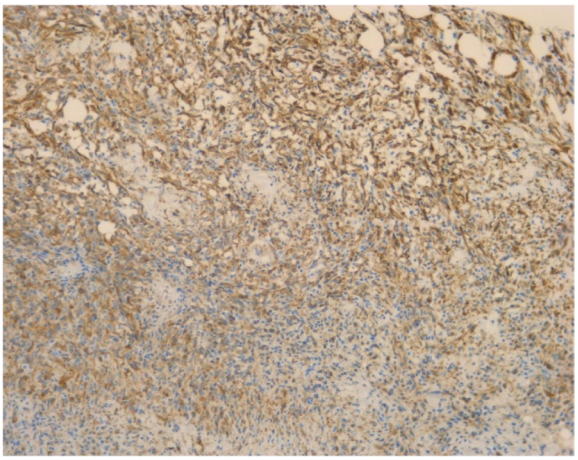

Fig. 4 Diffuse and strong membranous and cytoplasmic expression for *Smooth muscle actin* (SMA) × 20

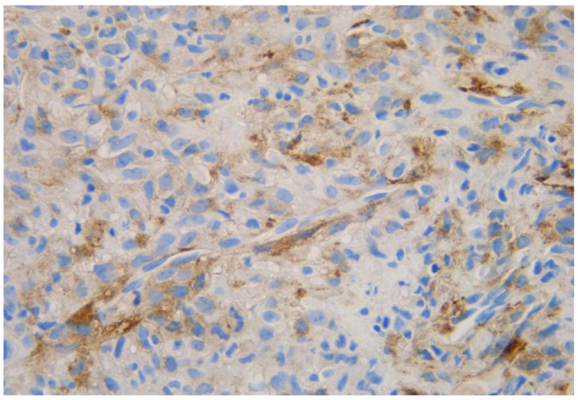

Fig. 5 Presence of focal expression of H-caldesmon × 40

right thyroid lobe, adhered to the anterior wall of the trachea, and infiltrated intraluminally. The mass was firm to hard. He had received palliative chemotherapy Doxorubicin. However, CECT scan post completion of chemotherapy yield devastating advanced stage of thyroid leiomyosarcoma whereby the tumor encased the right common carotid artery with multiple enhancing brain lesion at the right parietal and frontal lobes, likely to be brain metastases. Unfortunately, he succumbed to death 1 month after completion of chemotherapy.

Discussion

Papillary and follicular variants are the most common type of thyroid neoplasms, followed by medullary cancers [2]. Primary leiomyosarcoma (LMS) of the thyroid gland is a very rare tumor which accounts for 0.014%. They usually develop in older people (mean age 66 years) with no gender predisposition [4, 5]. Primary LMS may originate from smooth muscle cells of the capsule vessels. Metaplasia from a previously existing thyroid anaplastic carcinoma should take into consideration as well [2]. The initial symptom of all adult cases in the literature was mainly a rapidly growing mass in the neck, which causes breathing obstruction and dysphagia [3].

Primary LMS has no characteristic imaging features that might be useful for diagnostic purposes [5]. However, in the tumors observed in the ultrasonography (US) had smooth borders or were irregularly hypoechoic without halo, had cystic parts, were solid, and in some cases were calcified masses [3]. In the CTs, the masses involved large necrosis areas, with or without calcification [1, 3]. Ultrasound-guided *tru-cut* biopsy in our case was able to yield sufficient cells sample for further analysis.

Spindle cell tumor with thymus-like differentiation (SETTLE), anaplastic thyroid carcinoma and medullary thyroid carcinoma are thyroid epithelial tumors that can imitate sarcomas with a spindle cell pattern [5]. Thus, it can be difficult to exclude a diagnosis of undifferentiated thyroid carcinoma. Histologically, these tumors composed of an admixture of spindle cells, pleomorphic giant cells, and epithelioid cells. The percentage and distribution of these cell types varies widely from case to case. Tumors composed predominantly of spindle cells often have a sarcomatoid appearance. The individual neoplastic cells are elongated, with abundant acidophilic fibrillary cytoplasm; the nucleus is generally centrally located [2, 4, 5].

Immunohistochemically, LMS show reactivity for *vimentin*, *smooth muscle actin*, *muscle-specific actin*, *smooth muscle myosin*, *desmin*, *H-caldesmon*, and basal lamina components, including *laminin* and *type IV collagen*. *H-caldesmon* is a muscle marker used to discriminate between smooth muscle cells and myofibroblasts. On immunohistochemistry, positivity for *smooth muscle actin*, *vimentin*, *desmin*, and *caldesmon* may diagnose thyroid LMS, while these tumour cells show no reactivity to *keratin*, *thyroglobulin*, *chromogranin*, and *calcitonin* [2, 4, 5].

The dilemma in managing this case in terms of diagnosis and treatment enthralled surgeons. In contrast to thyroid carcinoma, there is no general recommendations regarding therapy and follow-up of LMS. Treatment strategies for LMS ranging from surgery, with or without additional chemotherapy and local radiation to palliative care. In this case, due to advanced LMS, it is not amenable for surgery, thus patient underwent chemotherapy, but the tumor showed poor response to chemotherapy. The general prognosis of thyroid leiomyosarcoma is poor. The disease is mostly fatal and survival rates are reported to be 5–10% in the first year [1–3]. We have reviewed several articles that mentioned their experiences in managing primary thyroid leiomyosarcoma (Table 1).

Table 1 Summary of 3 reports that discussed the management and outcomes of *Primary thyroid leiomyosarcoma*

Author (date)	*n*	Management	Outcomes
Surov et al. (2015) [1]	16	A total of 142 diagnosed with thyroid sarcoma, 16 patients diagnosed with *leiomyosarcoma*. 75 patients undergone surgery only 53 patients undergone surgery with chemotherapy with/without radiotherapy	Observation time 0.5 to 120 months; 45 patients were alive and 73 patients have died.
Conzo et al. (2014) [2]	2	Patient was diagnosed with thyroid leiomyosarcoma with lung metastases and undergone total thyroidectomy.	Patient's general condition deteriorated and succumbed to death 40 days after surgery.
Şahin et al. (2016) [3]	1	Patient 1 diagnosed with leiomyosarcoma of the thyroid gland with lung metastases. Patient undergone palliative radiotherapy. Patient 2 diagnosed with leiomyosarcoma of the thyroid gland with lung metastases with infiltration of the common carotid artery and trachea. Patient was not fit for chemoradiation.	Patient 1 have died 3 months after diagnosis due to extensive pulmonary metastases.Patient 2 died due to deteriorated general condition 45 days after diagnosis.

Conclusion

The encounter with primary thyroid leiomyosarcoma is a rare occurrence. It is a fatal disease which possessed a diagnostic conundrum as it may mimics anaplastic thyroid carcinoma. The management ranged from surgery, chemotherapy, and radiotherapy.

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Authors' contributions
WN made the conception and design of the work and writing. RJ and AA made the acquisition, analysis, and interpretation of data. KA performed the editing of the manuscript. NA performed editing of the manuscript. All authors read and approved the final manuscript.

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Availability of data and materials
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Declarations

Ethics approval and consent to participate
Not applicable.

Consent for publication
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Competing interests
The authors declare that they have no competing interests.

Author details
[1]Hospital Tuanku Jaafar Seremban, Seremban, Negeri Sembilan, Malaysia. [2]Universiti Sains Islam Malaysia, Nilai, Malaysia. [3]Hospital Kuala Lumpur, Kuala Lumpur, Malaysia.

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