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Primary thyroid smooth muscle tumour

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A 63-year-old male patient presented with a right neck mass that had been growing for 3 months. Over the past 20 days, the mass progressively increased in size and was accompanied by pain. The patient also experienced symptoms such as hoarseness and swallowing difficulties. Physical examination revealed a grade III enlargement of the right thyroid, with a palpable mass measuring approximately 4 cm in diameter. The mass was firm and had unclear borders. Laboratory tests, including thyroid function and calcitonin levels, were within normal range. Thyroid ultrasonography showed a mixed echogenic mass measuring 5.0 cm × 4.3 cm within the right lobe (Fig. 1). Fibreoptic laryngoscopy revealed right vocal cord paralysis. The preoperative diagnosis was thyroid neoplasm, and after obtaining consent from the patient and their family, the patient underwent surgical treatment. Intraoperative frozen section examination confirmed a malignant spindle cell tumour. Total thyroidectomy was subsequently performed. The postoperative pathology report (Fig. 2) indicated a spindle cell tumour with a tendency towards malignancy, accompanied by fibrosis, glassy transformation, calcification, and extensive necrosis. Immunohistochemical staining showed positive staining for smooth muscle actin (SMA) (Fig. 3) and vimentin, and negative staining for epithelial membrane antigen (EMA), thyroid globulin (TG), calcitonin (CT), thyroid transcription factor 1 (TTF-1), and paired box 8 (PAX-8). Ki-67 expression was 70%. These findings suggested a diagnosis of thyroid smooth muscle sarcoma. The patient was discharged on the 3rd day after surgery. They were treated with a combination of platinum-based chemotherapy and radiotherapy according to the cisplatin regimen. The therapeutic evaluation of the disease status was assessed as progressive disease (PD), and the family decided to discontinue treatment. The patient died 2 months postoperatively.

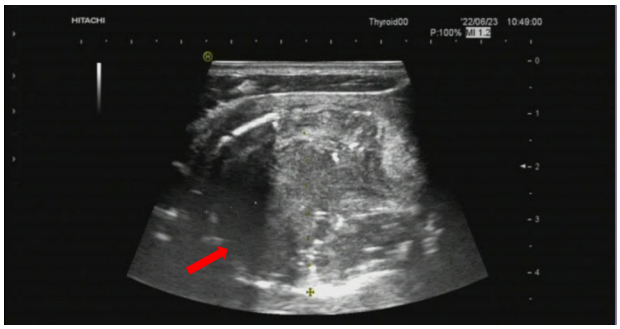

Figure 1. *A mixed echoic mass measuring approximately 5.0 cm × 4.3 cm × 3.5 cm, as indicated by the red arrow, was observed. Multiple punctate strong echoes were identified within the cystic wall and interior, accompanied by a comet tail artifact. No obvious blood flow signals were observed*

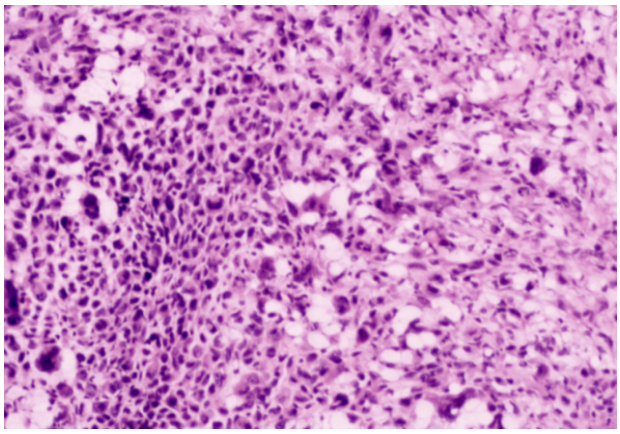

Figure 2. *HE10X10: spindle cell tumour with a malignant tendency, accompanied by fibrosis, hyaline degeneration, calcification, and extensive necrosis*

Primary leiomyosarcoma (LMS) of the thyroid gland is extremely rare, accounting for approximately 0.014% of

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all thyroid tumours [1]. It was first reported by Adachi in 1969 and is more commonly seen in individuals aged 65 years and above, with no gender difference. Current research suggests that it develops from the smooth muscle of the thyroid venous wall and is potentially associated with Epstein-Barr virus, previous history of malignancy, and prolonged radiation exposure. This disease progresses rapidly, and most patients present with the initial symptom of rapidly enlarging neck mass, accompanied by hoarseness, difficulty swallowing, and other symptoms, as described in this case.

There are no specific imaging features for this disease. Under ultrasound, most cases appear as cystic-solid masses with irregular borders, showing heterogeneous hypoechoic patterns and occasional calcifications. Computed tomography (CT) imaging usually reveals large areas of necrosis [2]. Magnetic resonance imaging (MRI) is only performed in a small number of cases, showing isointense signals on T1 weighted image (T1WI) and heterogeneous signals on T2WI, with significant enhancement on contrast-enhanced T1WI images [3]. In patients with rapidly enlarging neck masses, when imaging examinations indicate mixed echoic masses without blood supply and accompanied by punctate calcifications, a high suspicion for this disease should be maintained.

Most of these patients have normal thyroid function [4], making it extremely difficult to diagnose this disease based solely on laboratory and imaging examinations. Differential diagnoses often include the following diseases: (1) primary thyroid lymphoma: commonly observed in middle-aged to elderly females, with rare calcifications seen on ultrasonography. CT shows diffuse enlargement of the thyroid gland with homogeneous enhancement, while MRI shows isointense signals on T1WI, slightly high signals on T2WI, and high signals on diffusion-weighted imaging (DWI); (2) undifferentiated thyroid carcinoma: commonly seen in elderly females, with large solid hypoechoic nodules and accompanying cystic changes seen on ultrasound. CT shows irregular lobulated, poorly defined low-density masses with necrosis, cystic changes, and calcifications.

Histopathological examination reveals atypical spindle-shaped cells, making diagnosis challenging. Therefore, immunohistochemistry is crucial for a definitive diagnosis. CT negativity rules out medullary thyroid carcinoma, while TG and TTF-1 negativity in undifferentiated carcinoma, similar to primary thyroid LMS, require the evaluation of SMA, desmin (Des), vimentin, and PAX8 expression to determine the origin of the tumour [5]. EMA negativity excludes epithelial sarcomas and rhabdomyosarcomas.

Treatment options for primary thyroid LMS include surgical resection, chemotherapy, and radiation therapy, with most cases undergoing surgery. The efficacy of radiation and chemotherapy is still under research, and in this case, the patient had a very poor response to postoperative chemotherapy combined with radiation therapy. The one-year survival rate is only 5–10% [1], making early detection of this disease crucial.

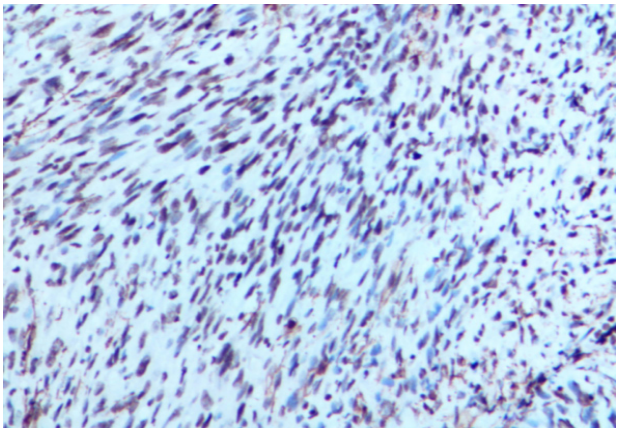

Figure 3. *Staining for smooth muscle actin (SMA)*

In conclusion, primary thyroid LMS is a rare and highly invasive malignant tumour that requires immunohistochemistry for accurate diagnosis. Currently, surgical resection is the primary treatment, with a very poor prognosis.

Conflict of interests

The authors have no conflicts of interest to disclose.

Statement of ethics

The research was conducted ethically in accordance with the World Medical Association Declaration of Helsinki. Data were collected retrospectively.

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Authors' contributions:

J.J. — article writing; F.Y. — topic selection; Y.Z. — data collection; F.-J.S. — data collection; B.Q.W — surgeon, manuscript revision.

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