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Rare Presentation: Primary Leiomyosarcoma of the Thyroid Gland

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Abstract: Leiomyosarcoma, an uncommon tumor, becomes even rarer when it emerges within the thyroid gland. Diagnosing it accurately is a challenging task due to its atypical location, diverse clinical and radiological presentations, its rarity, and overlapping differential diagnoses. However, immunohistochemistry studies can offer a precise identification. In this article, we present a compelling case of recurrent primary leiomyosarcoma of the thyroid gland in a 23-year-old man, revealed by inspiratory dyspnea caused by a voluminous cervical mass and ultimately confirmed as leiomyosarcoma through histopathological examination.

Keywords: Endocrinology, Thyroid gland, leiomyosarcoma, primary recurrent leiomyosarcoma.

Case Report

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INTRODUCTION

The occurrence of primary leiomyosarcoma within the thyroid gland is exceedingly rare, constituting only 0.014% of thyroid cancers [1, 2]. Originating from tissues rich in smooth muscle, such as the retroperitoneum, pelvis, digestive tract, and skin, this aggressive tumor is associated with poor prognoses [3, 4]. Surgical excision remains the gold standard treatment, with immunohistochemistry techniques aiding in its histological diagnosis [2].

CASE REPORT

A 23 years old man, was admitted to the ENT emergency department due to sudden inspiratory dyspnea that had developed over three days, without odynophagia nor dysphagia or dysphonia, the patient was unconscious with a Glasgow scale of 12/15 and signs of hypercapnia, cyanosis of the extremities, and paradoxical breathing with a SpO2 of 70%, BP of 80/40 mmHg and no palpable pulse. The patient present a midline anterior infrahyoid cervical mass, the latter is hard plunging, immobile with a thyroidectomy scar and the trachea was not palpable. Endoscopic examination revealed a normal larynx with doubt about subglottic/tracheal compression. CT scan performed 1 day prior to the emergency admission showed a voluminous anterior cervical heterogeneous mass at the expense of the thyroid gland with a triple component, tissue, calcium, and liquid related to necrosis. This mass extends from C5 to T3 and is responsible for an encompassment with a deviation of the trachea towards the left side with repression of the esophagus “Figure 1”.

After conditioning the patient, an emergency tracheotomy was performed by our ENT and neck and head surgery team, which lead to a spectacular improvement of the patient's condition with SpO2 of 97%, BP 120/80 mmHg, pulse of 70 bpm, without signs of dyspnea or signs of respiratory struggle. Afterward, the anamnesis revealed that the patient has no prior pathological history other than a thyroidectomy procedure 3 months earlier for a hard, immobile, rapidly growing thyroid mass. The histopathological examination revealed a total thyroidectomy specimen with a left lobe showcasing a well-defined whitish nodule with hemorrhagic and fibrous remnants and tumor proliferation. The tumorous cells which demonstrated a very high cell density were either round or spindle-shaped and they stain positively for anti-KI67, anti-SMA, and anti-Desmin; and negatively for anti-CD20, anti-CD3 anti-Pancytokeratin, anti-thyroglobulin anti-calcitonin anti-TTF1; which lead to the diagnosis of the thyroid leomyosarcoma “Figure 2”. The primary part of the diagnosis was confirmed with a negative extension assessment made by a body CT scan and abdominal ultrasound. The patient thereupon was referred to the Oncology department for further treatment. In the meantime, he presented signs of recurrence of the tumor

made by the rapidly growing midline anterior infrahyoid cervical mass with the same initial characteristics; which lead eventually to the dyspnea chart. Note that histopathological examination of the biopsy of the recurrent mass has the same conclusion as the thyroidectomy specimen; leomyosacroma of the thyroid gland. After discharging the patient from the ENT and Head and Neck surgery department, he was then submitted to the oncology department. Despite treatment efforts, the patient's condition worsened, and he succumbed to the disease, 2 weeks after the tracheotomy was performed.

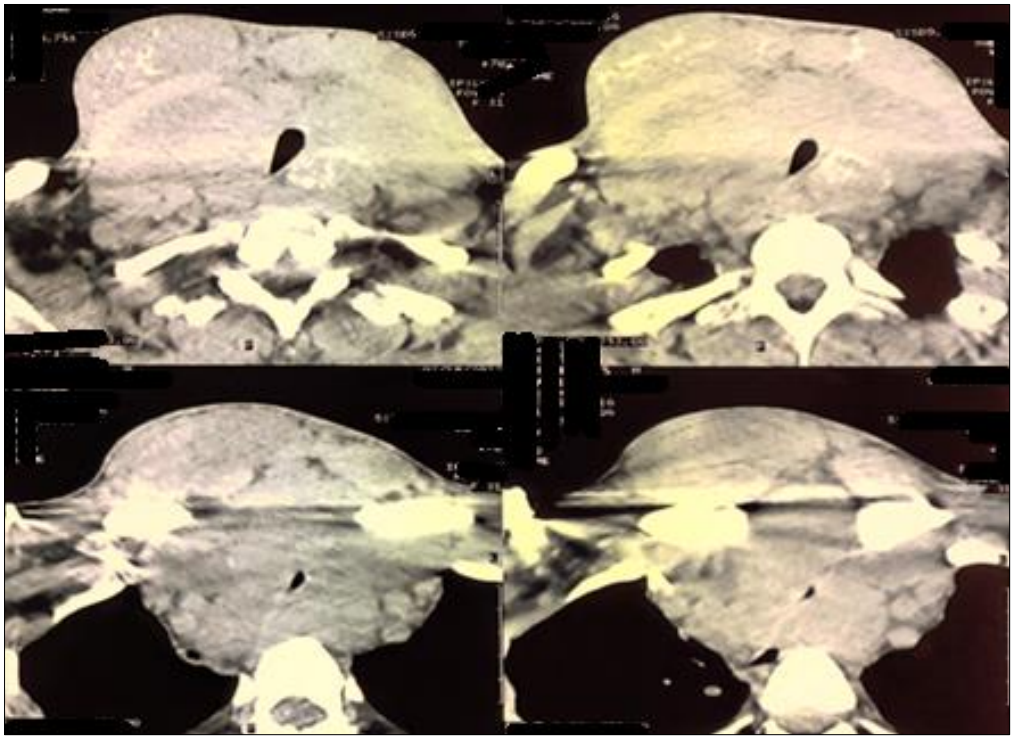

Figure 1: axial CT scan showcasing the voluminous heterogeneous thyroid mass with encompassment with deviation of the trachea towards the left side with repression of the esophagus.

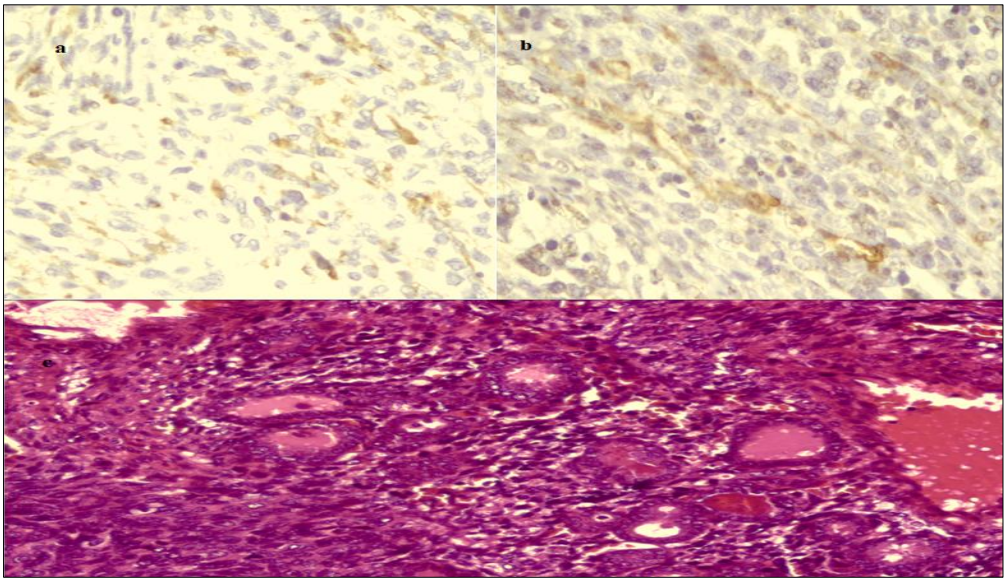

Figure 2: Anatomopathology images: (a): thyroid leimyosarcoma with spindle tumor cells and thyroid vesicles. (b): thyroid leiomyosarcoma with positive Ki67 stain, (c): thyroid leiomyosarcoma with a positive desmin staining

DISCUSSION

Primary thyroid leiomyosarcoma is an exceptionally uncommon malignancy, constituting just 0.014% of thyroid cancers [1, 2]. Reported cases are sparse in the literature, often affecting elderly females, with an average age of 61.4 years [3-5], though Tulbah *et al*., [6], reported a case of a leiomyosarcoma of the

thyroid in a 6-year-old child with congenital immunodeficiency.

Though the pathogenesis of this tumor remains uncertain, many authors incriminate the transformation of the smooth muscle-walled vessels in the thyroid gland capsule [4-7]. Others claim that it can result of metaplasia of the smooth muscle in a thyroid anaplastic carcinoma [8], or a malignant transformation of the smooth muscles, after an Epstein-Barr virus infection [6]. Overall, immune-deficient individuals appear to have a higher susceptibility [2-4].

Despite the lack of a pathognomonic clinical presentation, primary thyroid leiomyosarcoma typically manifests as a rapidly developing, painless neck tumor. Other symptoms, including weight loss, odynophagia, dysphagia, hoarseness, dysphonia, dyspnea, pain in the arm, and occurring cough have also been mentioned in the literature [9, 10], and to our knowledge, the majority of the cases reported in the literature are in euthyroidism, confined to one lobe [11, 12]. There is no preferred metastatic site, however, the most frequently mentioned site is the lung [12-13].

Radiologically, there isn't much of a difference between leiomyosarcoma and other thyroid tumors. The findings on ultrasonography revealed a smooth or irregular hypoechoic mass with cysts and solid components with or without calcifications [8, 9]. In contrast, a CT scan revealed a low-density mass with necrosis and the presence or absence of calcifications [14, 15]. MRI revealed isointense T1, with a fair signal increase in gadolinium T1 and a medium signal increase in T2 [1-5]. Fine needle aspiration cytology can detect suspicious malignant spindle-shaped cells, leading to the identification of a unique thyroid tumor, but it cannot differentiate leiomyosarcoma from anaplastic carcinomas [15, 16].

The main differential diagnoses are anaplastic thyroid carcinoma, spindle cell tumor with thymus-like differentiation, spindle cell variant of medullary thyroid carcinoma [17, 18].

Primary leiomyosarcoma of the thyroid is often composed of pleomorphic spindle cells with prominent mitotic activity and is extensively necrotic [13-19]. Vimentin [11-15], smooth muscle actin [7, 8], H-Caldesmon [15-19], desmin [8, 9], and HHF35 [2-13], stain positively, but calcitonin, cytokeratin, chromogranin, and thyroglobulin [2-15] do not.

The conventional therapy is still radical tumor excision [1-6], with surgeries ranging from thyroid lobectomy to complete thyroidectomy with therapeutic modified radical cervical lymph node dissection [7-20]. In other circumstances, palliative care is the sole option for preventing esophageal or airway blockage [15]. Immunotherapy, chemotherapy, and radiation are not effective in treating this particular tumor [21, 22].

In spite of the radical treatment, patients suffering from primary thyroid leiomyosarcoma, have somber prognosis; approximatively 50% of the patients died within 1 year of the diagnosis [1-10].

CONCLUSION

Due to the rarity of primary leiomyosarcoma of the thyroid gland; the treatment management is controversial; though radical surgery is the typical route, the roles of chemotherapy radiation or even immunotherapy remains to be established. The goal is proper, early diagnoses which can be obtained by regrouping the effort of multidiscipline participants in the hope of a better prognosis.

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