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Myasthenia gravis and retrosternal accessory Struma nodosa solved with cervico manubriotomy

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Abstract

The patient in this case was a 52-year-old woman with history of subtotal left thyroidectomy (struma nodosa colloida cystica) four years ago. She was euthyretic, with complaints of muscle weakness fatigue and slight respiratory difficulties. A few months before the operation she contracted COVID 19 (December 2020), with general respiratory problems. She had continued fatigue following her infection. In her CT thorax, we observed a tumor mass in the anterior superior mediastinum (5 x 6 x 5 cm), suggesting a thymic tumor. The neurologist did not report any neurological symptoms. It was treated as myasthenia gravis, and the patient was prescribed mestinon (60 mg x 4/24 hours) and supplemental calcium for a period of 2 and a half months. The surgical excision of the formation was made of through cervicomaniotomy. The histopathological result for the tumor was: cystic colloid

nodose goiter of the ectopic retrosternal thyroid gland. After the operation, the patient's condition was stable. The mestinon was continued with reduced doses up to 8 months after the operation, along with Letrox (75 mg/day) which she still takes. She continues to have fatigue and muscle weakness of the lower extremities, hypotrophy and hyporeflexy (unspecified polyneuropathy) and is undergoing treatment from a neurologist, a physiatrist and an endocrinologist.

Key words: Myasthenia gravis, Ectopic nodose struma; cervicomaniotomy

Introduction

Myasthenia gravis (MG) is an autoimmune disease characterized by fluctuating weakness of striated muscles, which can be severe enough to cause death¹.

MG is caused by antibodies against postsynaptic neuromuscular junction components². The disease can frequently coexists with comorbidities such as thymic tumors, but also with thyroid diseases, dislipidemias, diabetes mellitus, arterial hypertension and other autoimmune diseases.¹

Every patient with MG should receive a chest CT to rule out the presence of thymoma. In addition diseases must be ruled out before confirming the diagnosis³. Treatment for MG includes a limited range of interventions and there is no standardized scheme, so each case should be treated individually, according to stated recommendations⁴.

Case Report

The presence of ectopic (accessory) goiter in mediastinum is not commonly observed. It is associated primarily with symptoms of compression. Here we report the case of a 52-year-old woman with a history of subtotal left thyroidectomy (colloid cystic nodose goiter) four years ago. Was euthyreoid and her symptoms were mild difficulty breathing, facial swelling, and muscle fatigue. Clinically, there were also discrete signs of exophthalmus. A few months before she contracted COVID19 (December 2020), with general respiratory problems and ongoing fatigue after recovery, i.e. post-COVID symptoms.

The neurologist did not observe neurological deficits, and Acetyl choline receptor antibodies were negative. Her condition was treated as Myasthenia gravis and she was prescribed mestinon tabl. (60 mg x 4 / 24 hours) and supplemental calcium for a period of 2 and a half months.

The neck ultrasound showed that the contours of the thyroid gland were within a normal range. The CT scan of the thorax showed expansive heterogen formation (47x 67 x 50 mm) with regular contours but nodular in anterior superior mediastinum. Comprimed *Vena*

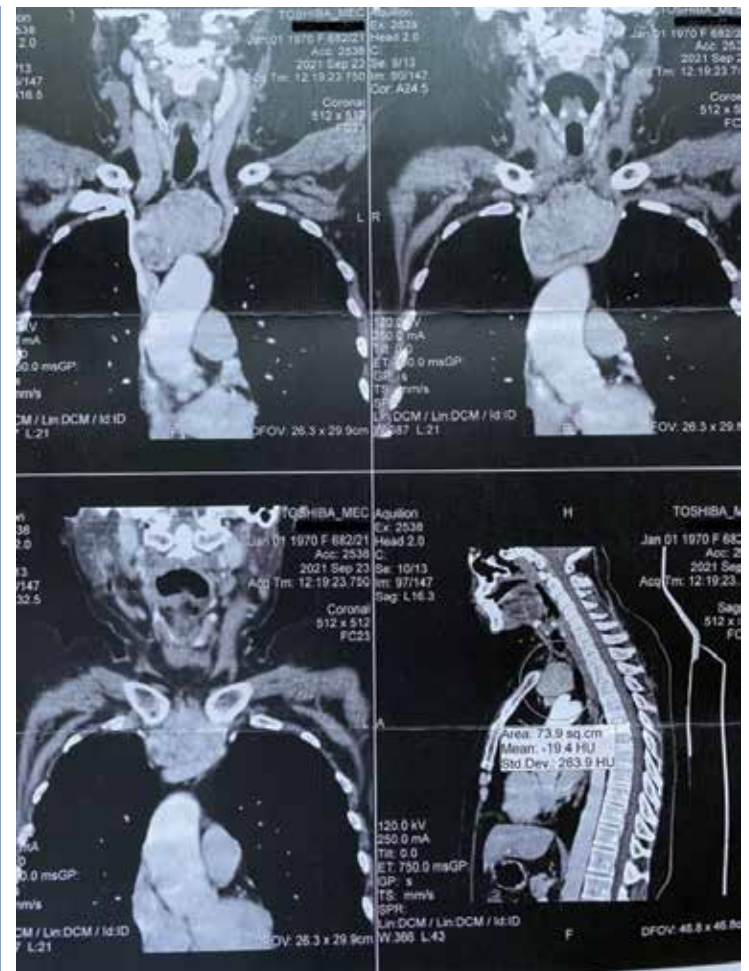


Figure 1: CT scan of neck /thorax
Figure 2: Scintigraphy of thyroid gland

Cava Superior but functional. (Figure 1)

The scintigraphy of the thyroid gland showed a normal right lobe. Technetium 99 m pertechnetate accumulated in the level of the retrosternal region with benign characteristics. (Figure 2)

The surgical excision was indicated. Next, we performed cervico manubriotomy, and the excision of the

Hemogramme	Electrolytes	Thyroid hormones	Receptors/ Antibodies
RBC: 4.82x 10 ⁶ ;	Na: 138mmol/L ;	TSH: 0.84 mIU/L	Acetyl choline receptor anticorps (AchRa): 0.1 nmol/L -negative (Refer.val. 0.1 nmol/L)
WBC: 11.2x 10 ⁹ μL;	Ca: 2.22mmol/L	T4 : 47 nmol/L	Muscle-specific tyrosine-kinase-antibody-(MuSK-MG): 0.015-negative (Refer.val.<0.015 nmol/L);
Hb: 144g/L ;	K: 4.3 mmol/L	T3: 2.01 nmol/L	
Htc: 39.9%;			
Plt: 191x10 ³ μL;			

Table 1: Hematologic, electrolytic, hormonal and receptor/antibody findings



Figure 3 and Figure 4: Lines for incision of cervicomantubriotomy and excision of tumour

tumour mass (Figures 3 and 4). This was done under general anesthesia with armored intratracheal intubation.

Furthermore, histopathologic test results (Figures 5 and 6), revealed a polynodose tissular structure. The tissue mass had a rose color and a volume of 7.3 x 5 x 3 cm which was determined macroscopically. Microscopically are seen strumogene changings with cystic foci and calcified zones. Histopathologic diagnosis were: *Struma nodosa colloides cystica gl. thyroideae*.

After the operation, the patient's condition was stable. Her post surgery treatment was as follows: continued treatment with mestinon in reduced doses for 8 months after surgery, and Letrox (75 mg/day), which she still receives. She currently complains of fatigue and

muscle weakness (hypotrophy) of the lower extremities and hyporeflexy (non-specific polineuropathy). In addition to prescribed medication, the patient continues to receive care from neurologist, physical therapist, and endocrinologist.

Discussion

The accessory or ectopic thyroid is a tissue, located anterolaterally between the second and fourth tracheal cartilages, in addition to thyroid tissue.

The terms ectopic/accessory thyroid have been used interchangeably in literature. The occurrence of ectopic mediastinal goiters is rare. Imaging plays a key role in preoperative evaluation of ectopic thyroid⁵. Kumar et al.⁶ presented a case with symptoms of myasthenia

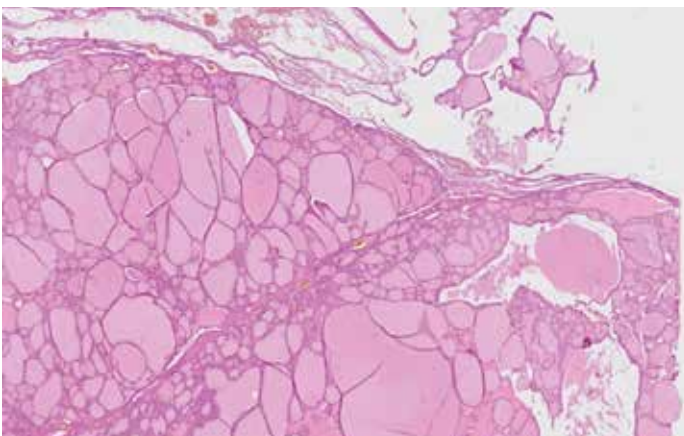


Figure 5: Strumogene changings with cystic foci

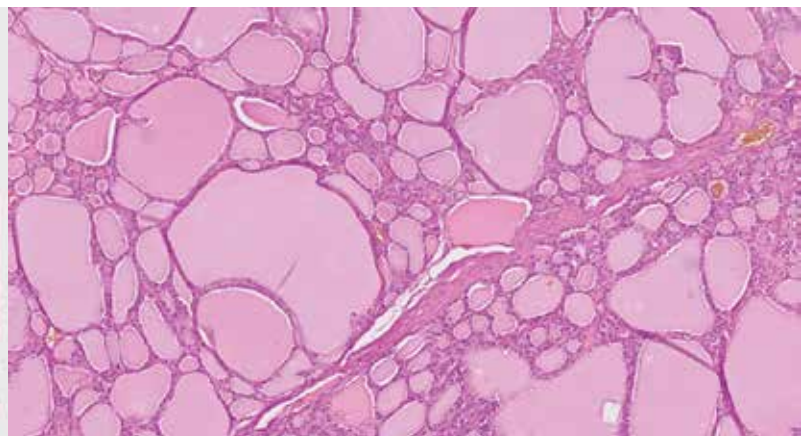


Figure 6: Calcified zones

gravis, where the patient was found to have an anterior mediastinal mass (colloid goiter), in addition to Crohn's disease. The authors also noted that 7% to 10% of adults have an ectopic asymptomatic thyroid tissue.

In a group of 304 myasthenic patients, 15 cases with thyropathies were reported: nine with hyperthyroidism, one with hypothyroidism and five with non-toxic goiters. Findings suggest that there is no clinical correlation between both myasthenia symptomatology and thyroid dysfunction⁷. Another work examined the records of 520628 cases of thyroid diseases and 7965 cases of myasthenia gravis. Diffuse toxic goiter had the highest association rate with MG, followed by nontoxic nodular goiter, simple goiter, chronic lymphocytic thyroiditis, thyroid cancer, and toxic nodular goiter.⁸ It must be underlined also that around 10-20% of myasthenia gravis (MG) patients do not have acetylcholine receptor (AChR) antibodies (seronegative), of whom some have antibodies to a membrane-linked muscle specific kinase (MuSK).⁹

Conclusion

This report examines and outlines the coincidence or possible comorbidity of MG with other thyroid illnesses such as, in this case, cystic colloid goiter. Therefore, patients with MG should be screened for thyroid diseases, diabetes, hypertension, dyslipidemia, autoimmune diseases and thymus tumors. Continuing post-surgical care and frequent follow-ups are also essential for long-term recovery.

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