

A Giant Primary Thyroid Fibrosarcoma in an Octogenarian

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Rezumat

Fibrosarcom tiroidian primitiv gigant la un octogenar

Prezentăm cazul unui pacient în vârstă de 89 de ani, care suferea de dispnee severă și disfagie moderată datorate compresiunii traheale și esofagiene printr-o gușă gigantă. Pacientul era eutiroidian. S-a practicat cu succes o tiroidectomie totală. Examenul histopatologic a relevat fibrosarcom G1, o tumoră extrem de rară a tiroidei.

Cuvinte cheie: gușă gigantă, tiroidectomie totală, fibrosarcom tiroidian

Abstract

We present an 89-year-old patient who was suffering from severe dyspnoea and mild dysphagia due to tracheal and esophagus compression by a giant goitre. The patient was euthyreotic. A total thyroidectomy was successfully performed. The pathology examination revealed fibrosarcoma G1, which is an extremely rare tumor of the thyroid.

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Key words: giant goitre, total thyroidectomy, thyroid fibrosarcoma

Introduction

The most frequent pathology of the thyroid is a nontoxic multinodular colloid goitre. Diagnostic procedures have improved due to better access to ultrasound, computed tomography, thyroid scintigraphy and fine-needle aspiration (FNA) biopsy. Thyroid carcinomas occur in less than 1% of patients with goitres. Lymphoma, fibrosarcoma and haemangioendothelioma lesions are extremely rare.

Case report

In this paper we describe the case of an 89-year-old woman who was admitted to our Casualty Department complaining of dyspnoea and mild dysphagia due to a giant goitre (Fig. 1). Her medical history was unremarkable. The patient had earlier been hospitalized on the General Internal Medical Ward because of the exacerbation of COPD. Steroids, bronchodilators, mucolytics and broad-spectrum antibiotics were administered with success. On the fourth day the patient was discharged from hospital and the recommended treatment was continued. Prior to hospitalization the patient had refused her consent to surgery. On admission the general condition of the patient was satisfactory, although she complained of difficulties with breathing and swallowing. On auscultation the breathing sounds were normal with isolated wheezes and coarse rales. Inspiratory stridor was easily observed. The ECG tracing demonstrated a regular sinus rhythm. The pulse rate was



Figure 1. The size of the giant multi-nodular goitre in an 89-year-old woman

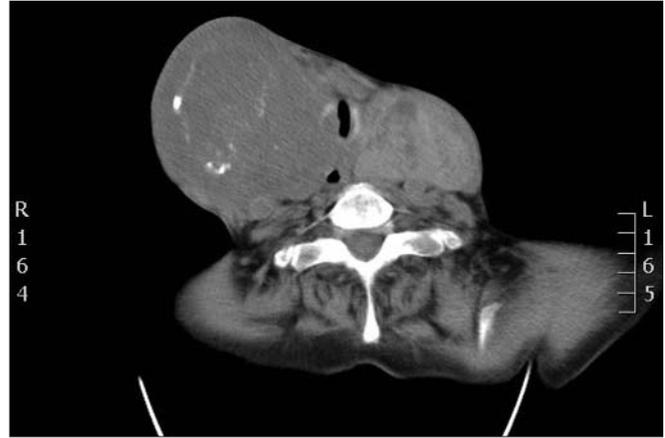


Figure 2. A computed tomography (CT) scan at the level of C7 showed the left lobe measuring 6x7cm in cross section, and 13 cm vertically in diameter; and the right lobe measuring 10 x 9.5 cm in cross section, and of 10 cm vertically in diameter. The thyroid had a heterogeneous density with areas of necrosis and calcifications

60/min and the BP was 130/65. Her lab tests, including TSH, fT3 and fT4, as well as blood gases were within normal limits. The arterial oxygen saturation (SaO₂) was 95%. An X-ray of the thorax disclosed discreet signs of emphysema and an enlargement of the superior vena cava. An ultrasound examination of the neck revealed that both lobes of the thyroid were enlarged and were of heterogeneous echogenicity, with multiple nodules and calcifications. CT scans confirmed that the substernal left lobe measured 6 x 7cm in cross section, and 13 cm vertically in diameter and that the right lobe measured 10 x 9.5cm in cross section, and 10cm vertically in diameter. Both lobes were of very heterogeneous density with areas of necrosis and calcifications (Fig. 2). The cystic lesion in the median part of the right lobe surrounded the larynx. The trachea was compressed and narrowed, being 0.5 x 1.8cm in cross section. The cervical vessels were dislocated. An ENT examination revealed a right vocal cord palsy and normal functioning of the left vocal cord. Surgery was recommended and informed consent was obtained. Under general anaesthesia, a traditional low transverse Kocher's incision and a total thyroidectomy were performed on February 28, 2011 (Fig. 3). The gland was exposed and dissected. First, the left substernal thyroid lobe, approaching the aortic arch, was excised. Then, the right lobe, constricting both the right carotid artery and the trachea, separating the esophagus from the trachea, was resected. The trachea was normal with no features of tracheomalacia. A fibrous mesh dressing measuring 10x10cm was applied for better hemostasis, and a drainage tube was inserted. On the first postoperative day the patient felt worse. She was dyspnoeic and her arterial oxygen saturation (SaO₂) decreased to 89-90%. An urgent ENT examination revealed only the oedema of both vocal cords, and mucus accumulation in the trachea, slightly constricting its lumen. The patient was conscious, but not well-oriented as to time, place and person. The patient then developed pronounced dyspnoea (SaO₂ was 77%). Respiratory insufficiency was diagnosed and the patient was immediately

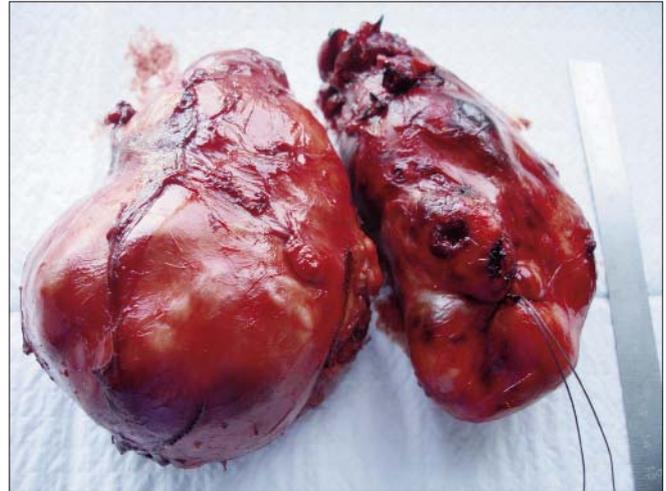


Figure 3. The combined weight of both excised thyroid lobes was over 200 g

transferred to the Intensive Care Unit. A tracheotomy was performed. She was administered invasive mechanical ventilation at first and assisted respiration afterwards. Eventually her spontaneous respiration returned. The patient spent seven days in the ICU, and finally made a gradual recovery. Our attempts to introduce oral nutrition were ineffective as the patient vomited several times. A mixed (parenteral and via naso-gastric catheter) nutrition was commenced. As the patient developed atrial fibrillation and arterial hypertension, she was put on routine hypotensive and beta-blocker therapy and her haemodynamic status improved. The gradual introduction of thyroid hormones was commenced, up to doses of 100 mcg daily while monitoring their concentration in the blood serum. The patient, coping well with the tracheostomy tube, was transferred back to the Surgery Department. Tracheobronchial toilet and reduced oxygen therapy was continued until finally the tracheostomy tube was removed. On the 14th postoperative day the patient

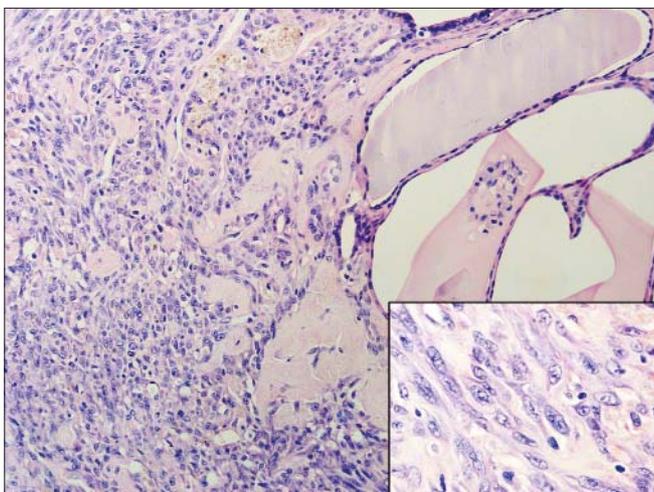


Figure 4. Thyroid fibrosarcoma. There are some follicles infiltrated by spindle cell neoplasm with minimal nuclear atypia (hematoxylin and eosin stain, magnification 100x and 400x)

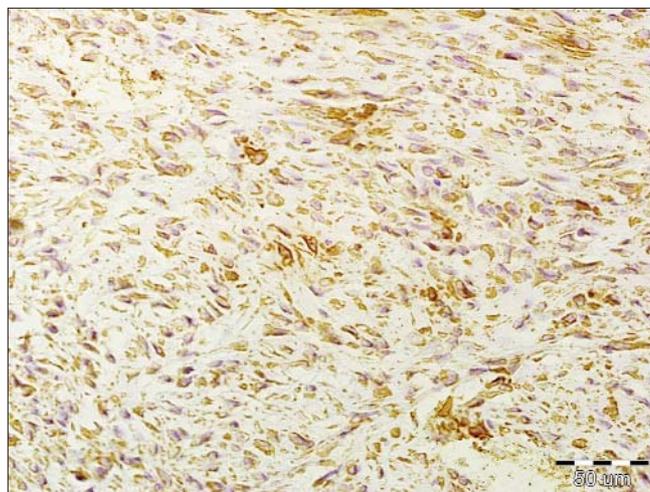


Figure 5. Immunostaining for vimentin in the tumor cells (magnification 200x)

was discharged from hospital. On a follow-up visit the patient manifested only slight difficulties in swallowing. The pathology examination of formalin-fixed paraffin-embedded (FFPE) tissue revealed malignant mesenchymal spindle-shaped tumor cells. A diagnosis of fibrosarcoma G1 seems to have been appropriate (Fig. 4). Immunohistochemically, the tumor cells were vimentin positive as well as negative for cytokeratin 19 and calcitonin. The proliferating index measured by the expression of Ki 67 protein was 1% (Fig. 5). The consultant oncologist did not recommend adjuvant chemotherapy or radiotherapy. The patient died suddenly several months after the operation.

Discussion

Soft tissue sarcomas are malignant mesenchymal tumors (1,2). A primary sarcoma may develop in any organ due to the ubiquity of mesenchyma in the human body. Fibrosarcomas affect organs containing fibrous tissue, most often the lower and upper limbs as well as the trunk. The thyroid is extremely rarely affected by fibrosarcoma. Thyroid sarcomas constitute less than 1% of all thyroid tumors (3). Primary fibrosarcoma of the thyroid gland has very rarely been presented in literature (1,2,4). It was reported for the first time by Alibert in 1879 (5). In our case the clinical and

radiological symptoms were not characteristic of a malignant tumor. The patient was euthyretic, so the tentative diagnosis was questionable. The indication for the surgery was the stridor due to tracheal compression by a giant goitre.

Conclusion

Primary thyroid fibrosarcoma is an extremely rare tumor. Its prognosis is poor and the recommended treatment is surgical resection.

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