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CASE REPORT

Myxofibrosarcoma of the thyroid gland

Maria da Graça Caminha Vidal[1], Onivaldo Cervantes[2], Melissa Falster Daubermann[3], Fernando Cogo Manduca[1], Aloysio Enck Neto[1]*

Abstract

Introduction: Thyroid cancer is the most common endocrine cancer, corresponding to 1% of all malignancies in the 30-74 age group. Cases of primary thyroid sarcomas are rare in the literature. They tend to present an unfavorable clinical course. **Case report:** Female patient, 66 years old, presented a visible neck mass with progressive growth for 2 years ago, reporting dysphagia for solids. Presence of exophytic nodular growth in the left lobe was evidenced through the examinations, and classified as Bethesda I through fine needle aspiration biopsy (FNAP). Total thyroidectomy revealed a low-grade myxofibrosarcoma (MFS). **Discussion:** MFS presents higher incidence in the elderly. Diagnosed by histopathological examination, evaluation through magnetic resonance imaging (MRI) is necessary in order to analyze the surgical resection plans with tumor-free margins. As MFS presents rare occurrence in the thyroid gland and a high local recurrence rate, adjuvant radiotherapy can be considered as a form of recurrence control.

Keywords: thyroid gland; thyroid nodule; thyroid neoplasms.

Introduction

Myxofibrosarcoma (MFS), previously known as a myxoid variant of malignant fibrous histiocytoma (MFH), is a malignant fibroblastic tumor presenting myxoid stroma and cellular pleomorphism[1]. It is one of the most aggressive types of soft tissue neoplasms[2]. The clinical presentation and histological aspects of MFS are highly heterogeneous, frequently delaying or masking the diagnosis[3], which is performed through histopathology. It is typically found in elderly patients, predominantly in men. They are usually located in the upper and lower limbs[2], and are rarely observed in the torso or head and neck regions. To date, cases of MFS have been reported in the larynx, esophagus, sphenoid sinus, pyriform sinus, maxillary sinus, mandible and parotid gland as primary sites, but seldom in the thyroid. These sarcomas must be differentiated from each other, as they are tumors with neural crest cells (spindle cell) that can be mistaken for anaplastic thyroid tumors, spindle cell variants of medullary thyroid carcinoma, melanoma, synovial sarcoma, leiomyosarcoma, fibrosarcoma, among others.

[1]Universidade Federal de Santa Maria (UFSM), Hospital Universitário, Departamento de Cirurgia de Cabeça e Pescoço, Santa Maria, RS, Brasil

[2]Universidade Federal de São Paulo (UNIFESP), Departamento de Cirurgia de Cabeça e Pescoço, São Paulo, SP, Brasil

[3]Universidade Federal de Santa Maria (UFSM), Hospital Universitário, Departamento de Patologia, Santa Maria, RS, Brasil

[4]Universidade do Contestado (UNC), Escola de Medicina, Mafra, SC, Brasil

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Case report

A Caucasian, 66-year-old woman was referred to the Head and Neck Surgery outpatient clinic for thyroid nodule investigation. She reported a noticeable cervical swelling with progressive growth for approximately two years, as well as complaints of throat irritation and dysphagia for solids. She presented a large, left-side mass on the topography of the thyroid gland. She brought an ultrasound (US) scan performed 18 months before the visit showing an exophytic nodule in the left lobe of the thyroid, without vascularization on Doppler, measuring 3.3×4.0×3.2 cm and two previous fine needle aspiration biopsies (FNAB), both Bethesda category I. Results of a new US showed topical thyroid enlarged in the left lobe, presenting parenchyma with heterogeneous echotexture of grossly nodular pattern. The dominant nodule occupied the left lobe almost entirely, presented a hypoechogenic halo, central areas of cystic degeneration, predominantly capsular vascularization on Doppler, and measuring 6.5×5.8×5.0 cm. The patient was submitted to total thyroidectomy. Pathological examination revealed a tumor consistent with low-grade MFS partly occupying the left lobe of the thyroid, with rare mitotic figures and no associated necrosis. A concomitant adenomatous follicular nodule in the right lobe was observed (Figure 1). In view of this result, immunohistochemistry (IHC) (Figure 2) was requested, and the diagnosis of MFS was confirmed. The patient presented no complaints at her return to the outpatient clinic, with mobile and symmetrical vocal folds at nasofibrolaryngoscopy (NFL), and a properly healed surgical wound. She underwent complementary radiotherapy and is undergoing oncological follow-up with no major changes.

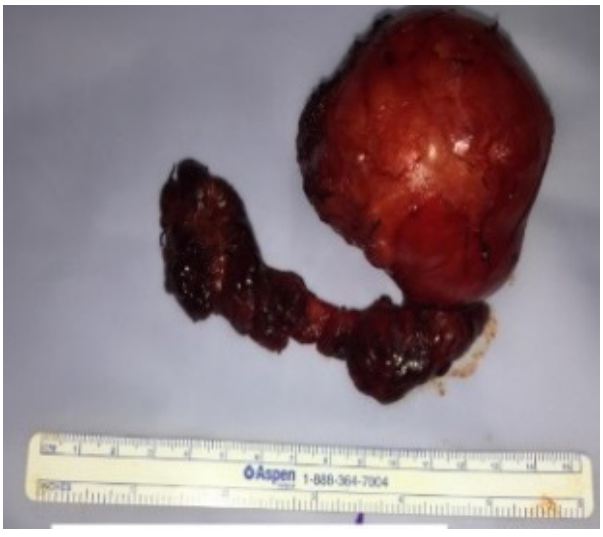

Figure 1. Macroscopic image of the surgical specimen.

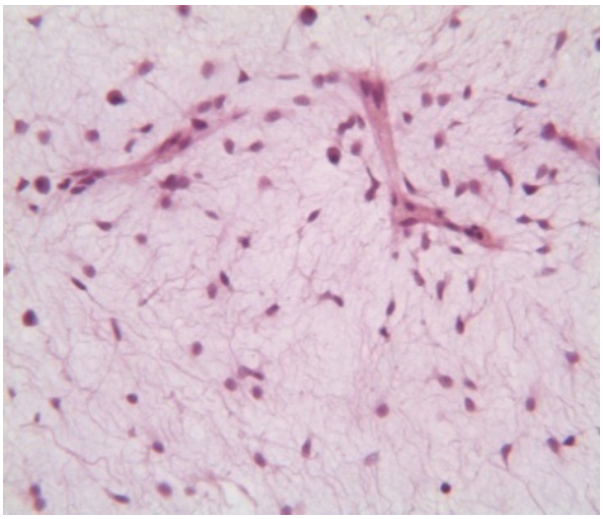

Figure 2. Myxofibrosarcoma - H&E staining.

Discussion

The MFS is one of the tumors of mesenchymal origin most frequently found in the elderly, with slight predominance in men[4]. Mean age of occurrence is 66 years, with a sex ratio of 1 (man) to 1.4 (woman). It usually presents slow and painless growth[4]. The most common locations are the upper and lower limbs, followed by the trunk, pelvis, and the head and neck and genital regions[4]. The proportion of myxoid areas defines the lesion grade, being more prominent in low-grade neoplasms[3]. Histopathology is the "gold standard" for establishing the definitive diagnosis[2].

According to previous studies, grade II and III tumors are truly malignant, as they present metastatic potential, contrasting with grade I lesions, which are only locally aggressive. To the best of our knowledge, there are no reports of grade I MFS with distant metastasis[2]. The likelihood of recurrence seems to be independent of the depth and histological grade of the tumor, and it can occur in up to 50-60% of cases.

Risk of metastasis is greater in deep and high-grade lesions, with the lungs and bones as the most common locations. The 5-year survival rate is 60-70%[2].

MRI is essential, and should precede treatment whenever is possible. In addition to the histological diagnosis, immunohistochemistry (IHC) should be performed to differentiate between them, as well as to distinguish from the spindle cell variant of medullary carcinoma or anaplastic thyroid carcinoma (ATC)[5].

The treatment for sarcomas of the head and neck regions is surgical, and should include extensive excision to obtain tumor-free margins. Radiotherapy is usually indicated for recurrent, non-resectable lesions, or for neoplasms with positive or close excision margins, to suppress local recurrence and the risk of histological progression, especially in low-grade MFS. Low-grade MFS seldom causes distant metastases, and thus presents a good short-term prognosis. However, the local recurrence rate of low-grade lesions is as high

***Correspondence**
Aloysio Enck Neto
Universidade do Contestado,
Faculdade de Medicina
Av. Presidente Nereu Ramos, 1071,
Jardim Moinho
CEP 89306-076, Mafra (SC), Brasil
Tel.: +55 (47) 99191-1706
E-mail: consultorioaloysioenck@gmail.com

Authors information
MGCV - Master and Senior Physician at the Head and Neck Surgery Service-HUSM (University Hospital of Santa Maria), Santa Maria/RS. Brazil
OC - Head and Neck Surgeon. Professor Doctor of UNIFESP (Federal University of São Paulo), São Paulo/SP. Brazil
MFD - Pathologist at Pathology Service of HUSM Santa Maria/RS Brazil
FCM -Head and neck surgeon
AEN - Head and neck surgeon, Professor of UNC (University of Contestado). Mafra / SC. Brazil

(50-60% in 5 years) as that of high-grade lesions. Moreover, MFS typically behaves more aggressively after each local recurrence. Therefore, MFS patients should be kept under long-term follow-up with regular MRI assessments[3,4].

The MFS is a subtype of soft tissue sarcoma of rare occurrence in the cervical region, mainly in the thyroid gland. When diagnosed, this tumor requires regular follow-up using highly sensitive image techniques (MRI) owing to the high local recurrence rate and the prospect of histological progression. Due to the difficulty in excising it, with wide surgical margins in the region, adjuvant radiotherapy can be an important alternative in local recurrence control.

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