International Journal of Medical Science and Health Research

Vol. 3, No. 06; 2019

ISSN: 2581-3366

Desmoid Tumor After Total Thyroidectomy and Radioiodine Therapy for Papillary Thyroid Carcinoma.

Authors: Antonella Carbone, Domenico Savio Cito, Pasquale Bellitti, Maria Chiara Fabiano and Rocco Bruno

Tinchi Hospital, ASM-Matera

Corresponding author: Bruno R

tel. no. 0835586561

Running title: desmoid tumor

Abstract

Desmoid fibromatosis is an uncommon neoplasm characterized by extensive stromal proliferation of fibroblasts and myofibroblasts that usually develops as a part of an ereditary syndrome or more frequently, in a sporadic form, often after a local trauma (1, 2). We report a case of a locally aggressive desmoid fibromatosis, developed in the thyroid bed after total thyroidectomy and radioiodine therapy (RAIU) for a classic papillary thyroid carcinoma (PTC).

A 42-year- old woman was referred to our endocrine unit because of onset of worsening dysphagia, dyspnea and cough three months after total thyroidectomy and RAIU for a previously diagnosed classic PTC. A palpable firm mass in the neck was detected at clinical examination and ultrasound imaging (US) revealed an hypoechoic inhomogeneous mass in left thyroid bed. Serum ultrasensitive thyroglobulin (Tg) and thyroglobulin autoantibodies (TgAb) were both undetectable. Fine needle aspiration of the mass and Tg dosage on the wash-out liquid of the needle excluded the persistence/recurrence of PTC in thyroid left bed. The evidence of tracheal dislocation and compression at computed tomography (CT) scan indicated surgery. Any way only partial excision of the mass was possible because of tracheal adhesion. Diagnosis of desmoid tumor was made at pathology and patient was referred to the oncologic unit. Chemotherapy (vinorelbine and methotrexate) plus tamoxifen were started. Six months after CT and US revealed a significant reduction of the mass. Actually patient is on levothyroxine treatment (100mcg/die), ultrasensitive Tg is <0.1ng/ml and TgAb is<10mUI/mL. US neck imaging does not reveal any suspect lymph node nor other signs of local recurrence of PTC. In conclusion desmoid fibromatosis should be take into account in the differential diagnosis of any suspect mass in thyroid bed.

Keywords: desmoid tumor, desmoid fibromatosis, PTC

Introduction

Background: Desmoid fibromatosis (DF) is a rare disease, 3% of soft tissues' neoplasms(2). It can interests everybody district, as a part of an ereditary syndrome, as in familial adenomatous poliposis FAP (about 5%-10%), or more frequently, in a sporadic form, usually after a local trauma(3).

The sporadic variant usually affects young adults, particularly female after a prolonged exposition to estrogens (4). Only 10% of these lesions occur in the cervical region and thyroid

Vol. 3, No. 06; 2019

ISSN: 2581-3366

involvement is even less frequent (5). DF does not metastasize, but could be local aggressive due to the adhesion and compression of surrounding organs (6). Pathogenesis of DF is not completely clear. It probably develops from a mesenchymal element. A role of heterozygous somatic activating mutations of CTNNB1 gene which encodes for β-catenin, and the APC/CTNNB1 pathway activation have been reported (7). History of a local trauma and the fibroblast-like cells proliferation with abundant collagen at pathology resemble an uncontrolled wound healing process (1). Here we reported the rare case of a desmoid tumor in the left thyroid bed after total thyroidectomy and RAIU for a classic PTC in the right thyroid lobe.

Case report

A 42yrs old woman was referred to our endocrine unit because of onset of worsening dysphagia and cough three months after total thyroidectomy and RAIU (3700 MBq) for a previously diagnosed PTC of the right lobe.

Familiarity for breast cancer was present (mother). History revealed prolonged estrogen treatment for visceral endometriosis (about 20 years). A palpable firm mass in the neck was detected at clinical examination. Neck US imaging revealed a hypoechoic inhomogeneous solid mass (27x24x65mm) in left thyroid bed. Tg and TgAb where both undetectable (Tg<0.1ng/ml; TgAb <10UI/ml). Fine needle aspiration biopsy (FNAB) of the lesion showed a small group of epitelial cells, with no atipia, surrounded from chronic inflammatory cells and active fibroblasts. Tg in the wash-out liquid of the needle was <0.1ng/ml. The evidence of tracheal dislocation and compression at computed tomography (CT) scan (see fig 1) indicated surgery. Any way only partial debulking of the mass was made because of tracheal adhesion. Pathology revealed myo fibroblast elements with nuclear atypia; immunostaining was positive for HHF-35, muscle specific actin, β- catenin and desmine. Therefore, diagnosis of desmoid tumor was made and patient was referred to oncologic unit. Estrogen therapy was stopped, and chemotherapy (vinorelbine and methotrexate) plus tamoxifen were started, in consideration of the high rate of local recurrence in case of surgery (1). After six months of therapy, CT (see fig 2) and US showed a significant reduction of the mass (26x21x37mm) with improvement of symptoms. Chemotherapy was than prolonged to 40 administrations. Actually patient is on levothyroxine treatment (100mcg/die), with ultrasensitive Tg <0.1ng/ml and TgAb <10UI/ml. US neck imaging does not reveal any suspect lymph node nor other signs of local recurrence of PTC.

Discussion

Desmoid fibromatosis of the neck is a rare disease. It consists in an uncontrolled monoclonal proliferation of myofibroblasts which usually don't metastasize but can have a local aggressive behavior, because of the compression and the invasion of the surrounding structures. It is not a unique disease but, in reason of its behavior, almost two forms can be distinguish, one with more indolent course and the other characterized by a local aggressiveness. This different disease course is not clearly predictable. Also the presence of free margins after the excision is not necessarily associated to a favourable prognosis(1). Generally the disease develops after a local trauma as the result of excessive reparative process(7). The evidence of a mass in thyroid bed in a patient with a previous diagnosis of classic PTC, although in the other side of thyroid bed,

Vol. 3, No. 06; 2019

ISSN: 2581-3366

forced us to exclude the persistence or the recurrence of PTC. FNAB and the Tg dosage on the wash-out liquid of the needle both excluded this case. Pathology with immunostaining performed after partial debulking of the mass has made it possible the differential diagnosis with other diseases, such as poorly differentiated thyroid carcinoma, anaplastic thyroid carcinoma, thyroid lymphoma and Riedel's thyroiditis, featuring all from the presentation as a solid firm mass in the neck (8). Moreover in our case, PTC with desmoid-type fibromatosis, which is an uncommon variant of PTC, was also rule out. In fact no atypical epithelial cells of PTC were described at pathology, but myo fibroblast elements only (9,11).

The observation of desmoid fibromatosis during or shortly after pregnancy and the reduction or the stabilization of the mass, during therapy with drugs as tamoxifen, supports the hypothesis of a role of estrogens in the pathogenesis of the disease(1,4). In our case history of prolonged estrogen exposition, about 20 years, was present because the woman was on estrogen therapy for endometriosis. For this reason hysterectomy and bilateral ovariectomy have been indicated both to treat endometriosis and to reduce the estrogen stimulated desmoid tumor' growth. Chemotherapy with vinorelbine and methotrexate, combined with anti-hormonal agent as tamoxifen significantly reduced the mass and compressive symptoms as CT scan revealed after six months from the beginning of therapy. Multi disciplinar (oncological and endocrinological) approach was adopted in the follow-up in reason of control both the desmoid tumor progression and PTC course.

Conclusions. Desmoid fibromatosis/tumor is a rare disease which should be taken into account in the differential diagnoses of a neck mass, particularly after surgery. Inpatient underwent surgery for a PTC a clear distinction between desmoid fibromatosis and PTC with desmoid-type fibromatosis is also desirable if both the stromal and epithelial cells are present in the histological specimen. Surgery has a limited role in the management of the disease which prognosis depends to the biological behavior of the tumor, the anatomical site and the response to chemotherapy and to anti-hormonal agents.

The authors don't have conflict of interest.

The patient gave informed consent.

References

Gronchi A., Colombo C, Le Pèchoux C. et al. (2014) Sporadic desmoid-type fibromatosis: a stepwise approach to a non metastasising neoplasm-- a position paper from the Italian and French Sarcoma Group. Annals of Oncology 25:578-583.

Fletcher JA, Bridge JA, Hogendoorn PCW et al. Desmoid-type fibromatoses. In WHO Classification of Tumors of Soft Tissue and Bone. Lyon: IARC Press 2013; 72–73.

Clark SK, Phillips RKS. Desmoids in familial adenomatous polyposis. Brit J Surg 1996; 83: 1494–1504.

Deyrup AT, Tretiakova M, Montag AG. Estrogen receptor-β expression in extra-abdominal bromatoses—an analysis of 40 cases. Cancer 2006; 106: 208–213.

Seper L, Bürger H, Vormoor J, Joos U & Kleinheinz J 2005 Aggressive fibromatosis involving the mandible – case report and review of the literature. Oral Surgery, Oral Medicine, Oral Pathology, Oral Radiology Erratum in: Oral Surgery, Oral Medicine, Oral Pathology, Oral Radiology 99 254.

Abdelkader M, Riad M & Williams A 2001 Aggressive fibromatosis of the head and neck (desmoid tumours). Journal of Laryngology and Otology 115 772–776. (doi:10.1258/0022215011909116

Cheon SS, Cheah AYL, Turley S et al. β-Catenin stabilization dysregulates mesenchymal cell proliferation, motility, and invasiveness and causes aggressive fibromatosis and hyperplastic cutaneous wounds. Proc Natl Acad Sci USA 2002; 10: 6973–6978.

Joana Simões-Pereira, Rafael Adame Cabrera and Valeriano Leit. A case of thyroid fibromatosis, a rare lesion of this gland.Endocrinology, Diabetes and metabolism 2016 Oct; 1-4.

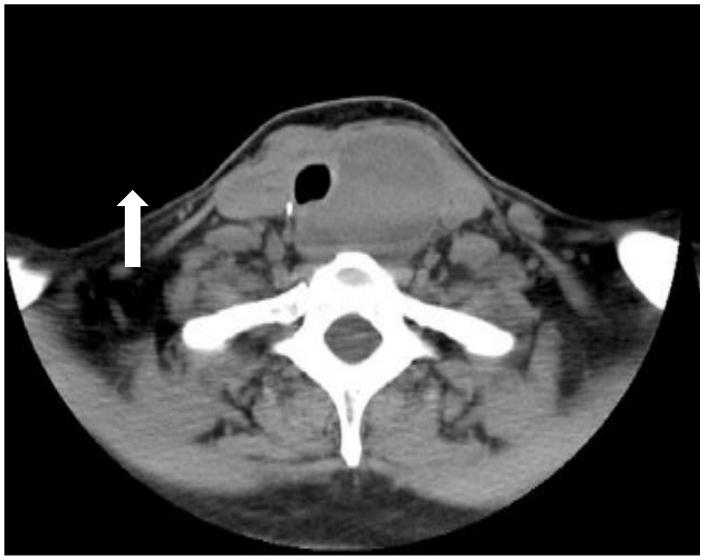
Chan JK, Carcangiu ML & Rosai J 1991 Papillary carcinoma of thyroid with exuberant nodular fasciitis-like stroma. Report of three cases. American Journal of Clinical Pathology 95 309–314. (doi:10.1093/ ajcp/95.3.309)

Wu Z, Chu X, Fan S, Meng X & Xu C 2013 Papillary thyroid carcinoma with fibromatosis-like stroma: a case report and review of the literature. Oncology Letters 5 215–217. (doi:10.3892/ ol.2012.993)

Inaba M, Umemura S, Satoh H, Ichikawa Y, Abe Y, Kirokawa K, Sakai H & Osamura RY 2002 Papillary thyroid carcinoma with fibromatosis-like stroma: a report of two cases. Endocrine Pathology 13 219–225. (doi:10.1385/EP:13:3:219

Eve M.Roth, Courtney E. Barrows, Michiya Nishino, BarrySacks, Per-Olof hasselgreen, Benjamin C. James. Papillary thyroid cancer with extrathyroidal extention desmoid - type fibromatosis. A case report of an aggressive presentation of an uncommon pathological entity. International Journal of Surgery Case Reports(2019) 63: 5-9.

Legend Fig.1: CT scan at the first diagnosis and at the start terapy.



Vol. 3, No. 06; 2019

ISSN: 2581-3366

Legend nFig. 2 CT scan performed six months after therapy which show a significant reduction of the mass.