

Collagenous Fibroma Mimicking a Retrosternal Goiter: A Case Report

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ABSTRACT

Collagenous fibroma, earlier known as desmoplastic fibroblastoma, is a rare fibrous soft tissue tumor that has been described more often in the extremities, where it presents as a slow-growing and painless mass. We report a case of a 48-year-old gentleman, who presented with an anterior neck swelling with substernal extension, resembling a retrosternal goiter, after clinical examination and radiological and cytological assessments. Intraoperatively, we found it to be a mass abutting but separate from the thyroid, arising from the deep aspect of sternum inaccessible from the neck requiring partial-sternotomy for excision. Histopathological examination revealed a hypocellular tumor made of spindle to stellate cells with surrounding stroma showing collagenization with collagen bundles to myxoid changes. This report documents a rare differential for goiter, only the second case reported in the central neck.

Keywords: Collagenous fibroma, Desmoplastic fibroblastoma, Retrosternal goiter, Soft tissue tumor.

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INTRODUCTION

Desmoplastic fibroblastoma is a soft tissue tumor, first described by Evans in 1995, which was later named collagenous fibroma.¹ It appears clinically similar to a desmoid tumor but is more well-circumscribed, less cellular, less vascularized, and less infiltrative. It commonly occurs in extremities but can present in the anterior aspect of the neck, which may be mistaken for goiter. Here we are describing the case report of a 48-year-old, male, who came with collagenous fibroma in the neck mimicking a retrosternal goiter.

CASE DESCRIPTION

A 48-year-old gentleman presented with a swelling in the neck which he had noticed 6 months ago with a gradual increase in size. He also complained of mild difficulty in breathing and dry cough on lying down. It was not associated with local pain, voice change, or any systemic symptoms like weight loss, breathlessness, or dyspepsia. He got evaluated elsewhere, CT showed bilateral nodules in the thyroid. A large soft tissue mass involving the right lobe of the thyroid extending retrosternally up to the aortic arch displacing the brachiocephalic artery, measuring 5.7 × 7.1 × 9.1 cm in size. Further, USG guided Fine needle aspiration cytology (FNAC) was reportedly suggestive of colloid goiter. As his symptoms worsened over time, an MRI scan for assessment reported an increase in the size of neck swelling to 12 × 9 cm.

He subsequently presented to our hospital for further management where a physical examination revealed a 7 × 8 cm palpable, firm mass with restricted mobility, in the anterior triangle of the neck toward the right. The swelling extended from the mandible superiorly to the clavicle inferiorly. The lower border was not palpable in Rose's position. The trachea was shifted to the left side (Fig. 1). The carotid arteries were not accessible bilaterally, but both superficial temporal arteries were palpable with good volume. Pemberton's sign was negative.

The imaging was reviewed with our radiologist and was suggestive of multinodular goiter with homogenous mass lesion probably from the right lobe (Fig. 2). He underwent surgery, planned for thyroidectomy,

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prepared for sternotomy if needed for the retrosternal component. Intraoperatively, there was a 10 × 15 cm firm fleshy mass in the right paratracheal regions deep to the strap muscle and extending posterior to the sternocleidomastoid muscle with retrosternal extension, this component was firm to hard which appeared fixed posterior to sternum in superior mediastinum unlike a retrosternal extension of benign goiter (Figs 3 and 4). Partial sternotomy was performed, and the tumor was noted to be in a plane close to the bone unlike a retrosternal goiter, needing to be shaved off from the posterior aspect of the sternum. The thyroid gland appeared enlarged with benign hyperplastic change and separate from the retrosternal mass which was closely abutting it but a separating plane was clearly defined. The thyroid gland was left *in situ*. Intraoperative frozen section reported a fibrous tumor, not of thyroid origin. Histopathology revealed a lesion measuring 15 cm, hypocellular, benign spindle cell neoplasm with myxocollagenous changes. Overall features were suggestive of desmoplastic fibroblastoma. It was further discussed in multidisciplinary team meetings; further surgery or adjuvant radiation or chemotherapy was not indicated, and the patient was kept on follow-up.

DISCUSSION

Evan was the first to describe “desmoplastic fibroblastoma” in a case series of seven patient in 1995.¹ It was renamed by Nielsen

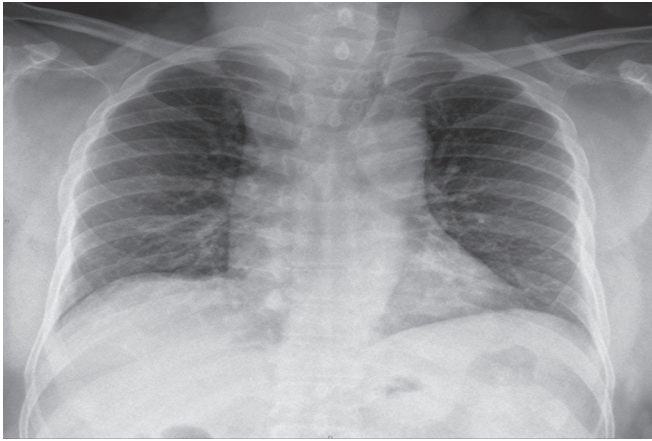


Fig. 1: Chest X-ray shows a gross deviation of trachea to left



Fig. 4: Gross specimen

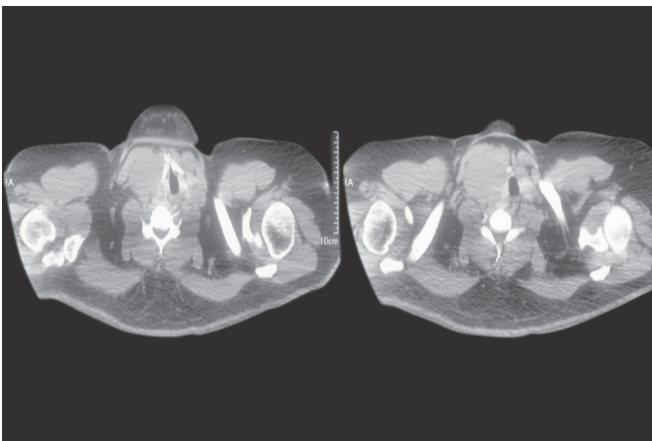


Fig. 2: CT of neck and thorax shows a large mass with heterogeneous density Right thyroid nodule

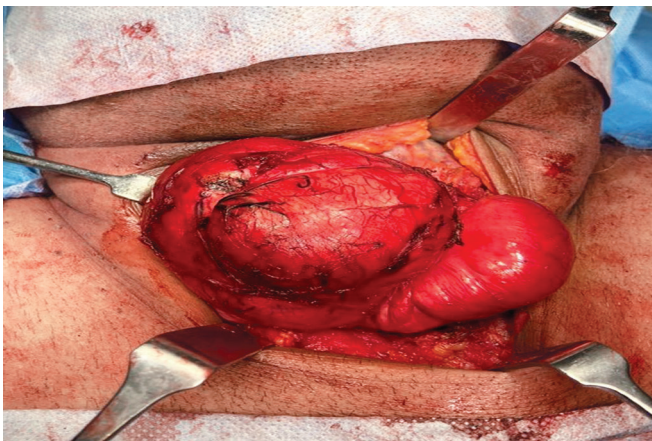


Fig. 3: Intraoperative image showing large neck mass extending to the anterior mediastinum

et al. as “collagenous fibroma,” as it may be perceived wrongly as a neoplasm composed of immature cells that induce scirrhous reaction; however, these tumor cells are mature cells and the collagenous stroma is not because of a reaction to them.¹⁻³ It mainly affects males in their fifth to seventh decades of life.⁴ It usually presents as a painless, slow-growing, large tumor (1–20 cm).⁵⁻⁷ Most common

sites of presentation are arm, shoulder, posterior neck, upper back, foot, ankle, leg, and hand.⁸ Fascial and muscle involvements are common.⁵ MRI remains the preferred imaging modality. Its low signal intensity at T2 weighted images is ascribed to the low cellularity of the lesion.⁹⁻¹² Other differentials to be considered are desmoid tumor, nodular fasciitis, fibroma of the tendon sheath, nuchal fibroma, calcifying fibrous pseudotumor, neurofibroma, and myxoma.^{5,10} This tumor is described pathologically as well-circumscribed tumor, composed of widely separated stellate or spindle-shaped cells embedded in a hypovascular fibrous or fibromyxoid stroma with less mitotic activity, calcification, or necrosis. Immunohistochemically, it is positive for vimentin and negative for desmin, CD 34, S100, cytokeratin.^{1,5,13} Surgical excision is the treatment of choice.⁵ No recurrence and metastasis have been reported. No role for External beam radiotherapy (EBRT) has been described for this. As described in a previous study, the origin of this tumor would be the fascia surrounding the thyroid gland or carotid sheath.⁸

In 2000, Christopher Wilson et al. reported a unique presentation of collagenous fibroma as goiter in an 88-year-old male, the only one in the literature before this report. Radiological evaluation plays an important role in ruling out thyroid pathology in such cases.

CONCLUSION

Collagenous fibroma is an extremely rare differential diagnosis of anterior neck swelling. It can resemble goiter with or without retrosternal extensions and present as an intraoperative surprise to the surgeon. As metastasis and recurrence are very unlikely hence the patient can be kept on follow-up postoperatively.

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