Yonsei Medical Journal
Vol. 45, No. 5, pp. 941 - 943, 2004

Collagenous Fibroma (Desmoplastic Fibroblastoma)

Muharrem Dagli[1], Adil Eryilmaz[1], Aydin Acar[1], Sezer Kulacoglu[2], and Halit Akmansu[1]

[1]Department of Otolaryngology-Head and Neck Surgery and [2]Department of Pathology, Ankara Numune Hospital, Ankara, Turkey.

A Collagenous Fibroma (Desmoplastic Fibroblastoma) is a rare, benign, slowly growing, fibroblastic, soft tissue lesion. Here, the case of a 28-year-old woman, who presented with a 1-year history of a slowly growing painless mass in the right anterior aspect of her neck, is described. This type of tumor was first described by Evans in 1995, and named as a Desmoplastic fibroblastoma but was renamed, by Nielsen in 1996 as a Collagenous Fibroma. This type of tumor is frequently reported in men with a mean age at occurance of 50 years. Clinically, a Collagenous fibroma presents as a firm, well-circumscribed subcutaneous, or intramuscular, painless mass of long duration. They are mostly located in the neck and extremities. The tumors range in size from 1 to 20 cm and predominantly occurs within the subcutaneous tissue, but fascial and skeletal muscle involvement is common. The treatment of a Collagenous Fibroma is a total surgical excision. No tumor recurrence has been reported the literature during the follow-up period and no tumor recurrence was observed in our case at the 1-year follow-up.

Key Words: Collagenous fibroma, desmoplastic fibroblastoma, fibroblastic, soft tissue lesion, neck mass

INTRODUCTION

A Collagenous Fibroma (Desmoplastic Fibroblastoma) is a rare, benign, slowly growing, fibroblastic soft tissue lesion. This type of tumor was first described by Evans, in 1995, and given the name a Desmoplastic fibroblastoma.[1] This tumor was subsequently renamed, by Nielsen, as a Collagenous Fibroma one year later.[2] Since then, the largest series of collagenous fibroma, with 63 cases, was published by Miettien and Flesch, and their clinicopathological characteristics better defined.[3]

To date, approximately 85 cases have been reported in the English literature. These tumors have a male predominance, and occur between the ages of 16 and 83 years.[3,4] The tumors predominantly occur within the subcutaneous tissue, but fascial and skeletal muscle involvements are common. They can occur at various anatomic location, including the shoulders, posterior neck, feet, legs, hands, abdominal wall, hips, parotid gland and palate.[3,5,6]

On gross examination the tumors range from 1 to 20 cm in maximal diameter, and appear oval, disc-shaped, or fusiform, with a firm, homogeneous pearl-gray color consistency.

Recurrences of the this type of tumor have not been reported. The treatment is a total surgical excision.

CASE REPORT

A 28-year-old woman presented with a 1-year history of a slowly growing painless mass in the right anterior aspect of her neck. She had no history of trauma, infection or weight loss.

A physical examination revealed a 5×4 cm firm mass located in her right lower neck. There was no pathological change to the overlying skin. She had no palpable lymphadenopathy in her neck. The results of her blood chemistry, urinalysis, thyroid function tests and chest x-ray were normal.

An ultrasound examination showed a $4 \times 2.6 \times 3.5$ cm smooth, heterogenous hypoechoeic mass, surrounded by fat tissue, in the lower part of the neck. The thyroid gland was normal. A computed

Received April 26, 2003
Accepted July 21, 2003

Reprint address: requests to Dr. Muharrem Dagli, Cemal Gursel Cad. No: 48/1 Cebeci 06600 Ankara Turkey. Tel: 90 312 310 3030-2208, Fax: 90 312 4358832, E-mail: mdagli@hotmail.com

tomography scan showed a 4 × 3.5 cm soft tissue dense mass, with no invasion to the adjacent tissue, located posterior to the sternocleidomastoid muscle (Fig. 1). The mass density was measured as approximately equal to that of fat tissue.

In the operative procedure, the mass was found in the subcutaneous layer, extending to the visceral layers of the supraclavicular region, posterolateral to the sternocleidomastoid muscle. The fibrous, smooth, firm mass was surrounded by fat tissue, with no invasion, or fixation, to the adjacent tissues. The tumor was totally removed by surgical excision. The resected mass was well-circumscribed and disc-shaped. On cross-section, a pearl gray color and homogeneous smooth surface, surrounded by fat tissue, was observed.

Microscopically, the tumor was composed of spindle shaped cells embedded in a collagenous stroma with moderate vascularity (Fig. 2 and 3). Niether mitotic activity nor necrosis were observed. The tumor infiltrated the subcutanous fat at the periphery, despite being well circumsribed on gross examination. Vimentin (Vq, NeoMarkers), smooth muscle actin (1A4, NeoMarkers), desmin (D_{33}, NeoMarkers), S-100 protein (4C4-9, Neo Markers) CD_{34} (QBEnd/10, NeoMarkers), cytokeratin (PAN-CK Cocktail, NeoMarkers) and epithelial membrane antigen (EMA, E29, NeoMarkers) Immunohistochemical stainings were performed.

The tumor cells were positive for vimentin, but there was no immunoreactivity for actin, desmin, S-100, EMA, cytokeratin and CD_{34}.

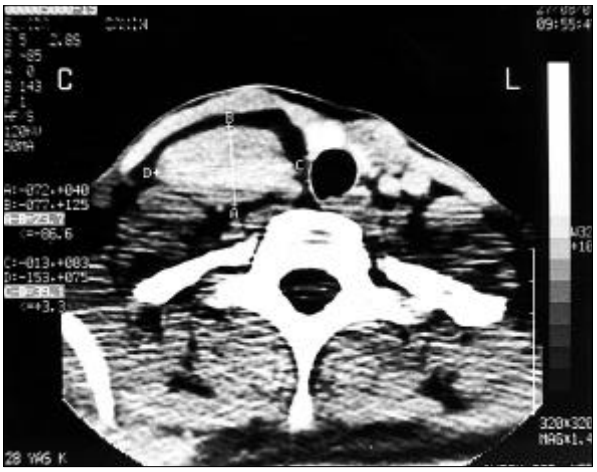

Fig. 1. The CT scan showed a 4 × 3.5 cm soft tissue mass, with a measured density approximately equal to that of fat tissue, in the right lower neck.

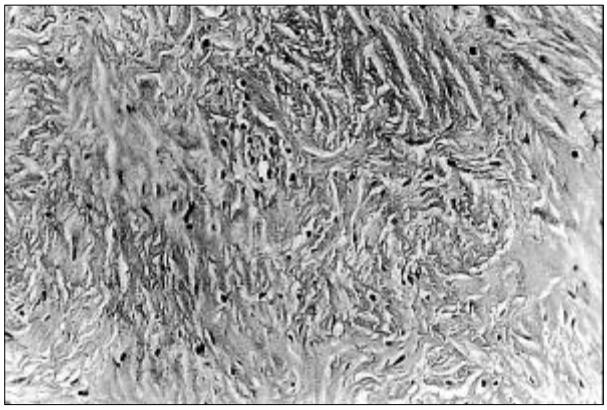

Fig. 2. The lesion is hypocellular, and composed of spindle-shaped fibroblasts embedded in a collagenous stroma. H-E × 100.

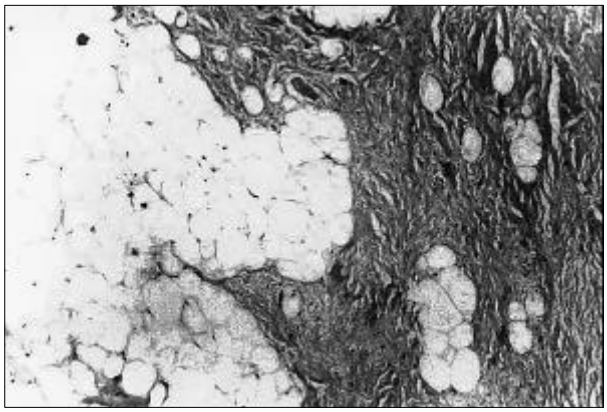

Fig. 3. Despite being well-circumscribed on macroscopic examination, microscopically, infiltration of the surrounding adipose tissue at the periphery is observed. H-E × 40.

DISCUSSION

Clinically, a Collagenous fibroma presents as a firm, well-circumscribed subcutaneous, or intramuscular, painless mass of long duration. They are mostly located in the neck and extremities. According to the literature, they frequently occur in men with a mean age at occurance of 50 years. The tumors range in size from 1 to 20 cm. Grossly, the tumors appear oval, disc-shaped, or fusiform, and have a firm, homogeneous pearl-gray colored consistency on cut section.[3]

The differential diagnosis of a Collagenous Fibroma includes a variety of soft tissue lesions: Neurofibroma, fibromatosis, nodular fasciitis, fibroma of the tendon sheath, solitary fibrous tumor, perineuroma, nuchal fibroma, sclerotic fibroma of the skin, calcifying fibrous pseudotumor, low grade fibromyxoid stroma and myxoma. However the recognition of a Collagenous Fibroma is not difficult if the clinical findings (age of patient, the location and the depth of the lesion) and the pathological features (the low cellularity and relatively monotonous appearance of the tumor, with no mitotic activity, necrosis and infiltrative margins of most of these tumors microscopically) are evaluated together.

Also, immunohistochemistry aids the correct diagnosis when doubt exists. Especially, S-100 protein, CD34 and EMA have proven to be useful markers for excluding neurofibroma, solitary fibrous tumor and perineuroma, respectively.[3]

Adjacent skeletal muscle and fat tissue entrapment are a common histopathological findings in Collagenous Fibromas. Mietteinen and Fetsch observed entrapment of fat in 51% of their 63 cases.[3]

In our case, both the CT scan and ultrasonography showed the mass, surrounded by fat tissue, and intraoperatively, the mass was actually observed to be surrounded by fat tissue. The entrapment of fat tissue has also been reported by Mietteinen and Fetsch. A hypothesis relating the preoperative disclosure of this fat tissue entrapment on CT scan and US can be proposed. Therefore this finding can help in the preoperative diagnosis of the disease.

The treatment of a Collagenous Fibroma is a total surgical excision. No tumor recurrence of this type of tumor has been reported in the literature pertaining to the follow-up period. No tumor recurrence was observed in our case at the 1-year follow-up.

REFERENCES

1. Evans HL. Desmoplastic fibroma: A report of seven cases. Am J Surg Pathol 1995;19:1077-81.
2. Nielsen GP, O'Connell JX, Dickersin GR, Rosenberg AE. Collagenous fibroma (desmoplastic fibroblastoma): A report of seven cases. Mod Pathol 1996;9:781-5.
3. Miettinen M, Fetsch JF. Collagenous fibroma (desmoplastic fibroblastoma): A clinicopathologic analysis of 63 cases of a distinctive soft tissue lesion with stellate-shaped fibroblast. Hum Pathol 1998;29:676-82.
4. Jang JG, Jung HH, Suh KS, Kim ST. Desmoplastic fibroblastoma (Collagenous fibroma). Am J Dermatopathol 1999;21:256-8.
5. Ricardo AM, Edson O, Waldir AJ, Vera C de A. Collagenous fibroma (desmoplastic fibroblastoma): A case of report. Oral Surg Oral Med Oral Pathol 2001;91:80-4.
6. Fumio I, Tetsuo S, Norio H, Hiroshi T. Collagenous fibroma (desmoplastic fibroblastoma) presenting as a parotid mass. J Oral Pathol Med 1999;28:465-8.