Case Report

Case Report of a Primary Cutaneous Leiomyosarcoma on the Neck

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Abstract

Primary cutaneous leiomyosarcoma is a rare dermal smooth muscle neoplasm more commonly seen in men in their 50s to 70s. It presents as a painful solitary nodule most commonly on the extremities. Here, we report a case of primary cutaneous leiomyosarcoma on the right neck of a 78 year-old man.

ABBREVIATIONS

SMA: Smooth Muscle Actin; CT: Computer Tomography; IHC: Immunohistochemistry

INTRODUCTION

Primary leiomyosarcoma of the skin is a rare smooth muscle tumor, accounting for about 2-3% of superficial soft tissue sarcomas [1]. These tumors are divided into cutaneous and subcutaneous leiomyosarcomas based on their primary site of origin. Cutaneous leiomyosarcomas are thought to derive from the arrector pili muscle in the dermis and have a relatively benign clinical course while the subcutaneous variant is associated with a more aggressive clinical course and is thought to be derived from the smooth muscle of vessel walls [1,2].

In this article, we discuss the clinical presentation, histological and immunohistochemical features, treatment, and follow up of this rare neoplasm in a 78 year-old man with a primary cutaneous leiomyosarcoma arising on the right neck.

CASE PRESENTATION

A 78 year-old man presented to the Veterans Administration Medical Center with a one-month history of a rapidly growing bleeding, painless mass on the right neck. His past medical history was significant for multiple non-melanoma skin cancers, coronary artery disease, hypertension, and hyperlipidemia.

Clinical exam was remarkable for a 5 cm friable, hemorrhagic nodule on the right neck (Figure 1). There was no palpable cervical or axillary lymphadenopathy. Computer tomography (CT) of the neck taken after a shave biopsy of the entire nodule revealed no soft tissue mass or abnormal lymph nodes. There was a small right lung nodule which was incidentally found that has been stable in a follow-up CT scan. A flexible laryngoscopy did not show any evidence of local invasion or metastasis.

The nodule was shaved flush to the skin with a #10 blade. Gross inspection revealed lobulated tumor mass measuring 4.5 cm x 3.5 cm x 2.5 cm with central ulceration. The cut surface of the mass was tan to white and solid.

Histological evaluation with hematoxylin and eosin staining showed a dermal spindle cell neoplasm composed of cytologically atypical cells with nuclear pleomorphism and mitotic activity (Figure 2). Immunohistochemical (IHC) staining showed strong and diffuse reactivity with smooth muscle actin (SMA) (Figure 3) and no reactivity with S-100 or pankeratin (data not shown). IHC staining of the excision specimen showed residual spindle cell neoplasm staining positive for SMA and desmin. The final

Keywords

• Leiomyosarcoma
• Neck
• CT
• Treatment

Figure 1 Clinical photograph showing a 5 cm irregular, friable, hemorrhagic nodule on the right neck.
The patient was referred to Otolaryngology and a wide local excision with a 1cm margin was performed. The final surgical margins were free of tumor. The patient’s case was presented at a multidisciplinary tumor board and it was felt that the recurrence rate would be low and no additional treatment was recommended. At a four month follow up with dermatology and a five month follow up with otolaryngology, there was no evidence of recurrence of the leiomyosarcoma.

DISCUSSION

Primary leiomyosarcomas of the skin are rare soft tissue tumors, representing about 2-3% of all superficial soft tissue sarcomas [1]. While other types of leiomyosarcomas are more common in women, cutaneous leiomyosarcomas are more common in men. They are usually seen in men in the fifth to seventh decades of life and present as a painful solitary nodule on the extremities. They have occasionally been seen on the head and neck region [1-7]. Our patient was a 78 year-old man who presented with a rapidly enlarging ulcerated nodule on the right neck.

In the past, leiomyosarcomas of the skin were considered a homogenous entity, but in the last couple of decades, they have been divided into two subtypes based on their primary site of origin. Cutaneous leiomyosarcomas are thought to derive from the arrector pili muscle in the dermis. Subcutaneous leiomyosarcomas are thought to arise from the vascular wall and are known to have a higher rate of local recurrence (50-70% of cases) and metastasize to distant sites in up to 30-40% of cases [2,8,9].

Histopathologically, cutaneous leiomyosarcomas are composed of a dermal proliferation of elongated spindle cells that have pleomorphic, cigar-shaped nuclei arranged in interlacing fascicles. They are usually well-to-moderately differentiated tumors which show a high rate of mitotic activity [2,7]. Other spindle cell tumors that are considered in the histopathologic differential diagnosis include a spindle cell melanoma, spindle cell carcinoma, cellular schwannoma, dermatofibrosarcoma protuberans, or an atypical fibroxanthoma. Immunohistochemical studies are required to distinguishing between these different spindle cell tumors. Cutaneous leiomyosarcomas stain positively for SMA, desmin and h-caldesmon. They also stain positive for epithelial membrane antigen (EMA), muscle specific antigen (HHF35), and creatinine kinase (CK) [10-12].

The preferred treatment for cutaneous leiomyosarcoma is wide local excision [2,6,13]. Earlier studies reported a recurrence rate of up to 40% but in some of these case series, a distinction between cutaneous and subcutaneous leiomyosarcomas was not made. [2,6,7] Cutaneous leiomyosarcomas have a lower rate of recurrence and metastases. The local recurrence rate of purely dermal cutaneous leiomyosarcomas in one study was 6.2% [3]. Factors that have predicted recurrence include size of the tumor, high mitotic rate, and extension of the tumor into the subcutaneous tissue. The risk of local recurrence is strongly related to the adequacy of excision with margin status being the strongest predictor of recurrence [1-8]. In a retrospective study performed by Deneve et al primary surgical excision with a median margin of 1cm showed no recurrences in 33 patients at a median follow-up of 15.5 months. Metastasis was not seen in these tumors and they had a favorable clinical course [5]. Factors that are associated with metastasis include depth of tumor on diagnosis and histological grade of the tumor [1-8].

Our patient was treated with wide local excision with a 1cm margin. After presentation at a multidisciplinary tumor board, no further treatment was felt to be necessary and at a 5 month follow-up, there was no evidence of local recurrence or metastasis.

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REFERENCES


