ABSTRACT

Objectives: To describe a rare case of eccrine spiradenoma of the upper lip

Methods: Retrospective case review at a university medical center.

Results: A 76-year-old female presented for evaluation of a midline upper lip nodular mass. It was initially appreciated incidentally, but had been present for at least several months in duration. The patient denied any associated pain, drainage, lip trauma, changes in size of the mass or prior history of similar lesions in the head and neck region. Physical examination of the upper lip revealed a midline submucosal, highly mobile, nontender nodule approximately 1 cm in size near the frenulum. An excisional biopsy was performed and revealed a firm, multilobulated and nodular lesion with smooth boundaries. The lesion was completely excised with adequate margins obtained. Surgical pathology was consistent with eccrine spiradenoma with no atypia or malignancy identified. To date, the patient has had no evidence of recurrence.

Conclusions: Eccrine spiradenoma is an uncommon benign adenexal tumor of the eccrine sweat glands. These tumors can present in any part of the body, with approximately one third occurring in the head and neck region. The risk of malignant transformation is a rare phenomenon, but can develop with a long-standing, initially benign solitary lesion. With only one other reported case of eccrine spiradenoma involving the perioral region, we present an interesting case of eccrine spiradenoma of the upper lip with discussion of its clinical presentation, histopathologic characteristics, and management.

INTRODUCTION

Eccrine spiradenoma (ES) represents an uncommon benign adenexal tumor of eccrine sweat glands. Approximately one third of ES occur in the head and neck region; however, only one other case of ES in the perioral region has been reported in the literature. We present an interesting case of ES of the upper lip, focusing upon its clinical presentation, histopathologic characteristics, and management.

CASE REPORT

A 76-year-old female presented for evaluation of a midline upper lip nodular mass. It was initially appreciated incidentally, but had been present for at least several months in duration. The patient denied any associated pain, drainage, lip trauma, changes in size of the mass or prior history of similar lesions in the head and neck region. The patient does have a history of left temple cutaneous squamous cell carcinoma that was treated with Mohs micrographic surgery in 2005 without recurrence. She previously smoked 2-4 cigarettes per day for 20 years, but denied any current use of tobacco.

Physical examination of the upper lip revealed a midline submucosal, highly mobile, nontender nodule approximately 1 cm in size near the frenulum. Both the underlying mucosa and overlying skin were normal. The remainder of the head and neck exam was entirely normal. During the excisional biopsy, the mass was appreciated immediately upon entering the submucosal space as a firm, multilobulated, and nodular lesion with smooth boundaries. The surgical pathology showed eccrine spiradenoma with no atypia or malignancy identified. The lesion was completely excised with adequate margins obtained. To date, there has been no evidence of recurrence.

DISCUSSION

Eccrine sweat glands are cholinergic in nature and are located within the dermis to aid in thermoregulation. Eccrine spiradenoma (ES), as first described by Kersting and Helwig in 1956, is a benign adnexal tumor of these eccrine glands. These tumors can appear in any part of the body, but tend to develop on the upper, dorsal aspect of the body. Approximately one third of ES occur in the head and neck, one third on the trunk, and one fifth on the extremities. Only one other case of perioral ES has been reported in the literature. Although ES can appear at any age, the highest incidence occurs in young adulthood, especially during the second to fourth decades of life. No gender predominance has been identified. Eccrine spiradenoma can also occur in the setting of Brooke-Spiegler syndrome, an autosomal dominant disease consisting of multiple spiradenomas and a predilection towards developing cutaneous adnexal neoplasms, such as trichoepitheliomas and dermal cysts.

 Clinically, patients often report spontaneous pain or tenderness on palpation. These symptoms have been theorized to stem from a large plexus of nerves surrounding the connective tissue capsule. Eccrine spiradenomas are typically solid skin tumors, but may also appear in multiple, giant, linear, blashkoid, or grouped patterns. The lesions often present as bluish-red dermal or subcutaneous nodules that can range in size from 0.5 cm to 3 cm in diameter. These tumors are arranged in sheets, cords or islands. Alternatively, a trabecular arrangement may be seen. The cells are periodic acid Schiff (PAS) negative. A few duct-like structures, squamous eddies, small cysts, and lymphocytes can infiltrate the tumor nests. Fine needle aspiration, ES can appear similar to adenoid cystic carcinoma as both contain hyaline globules. This can be of particular concern when tumors occur along the normal distribution of salivary glands. The treatment of choice of eccrine spiradenoma is surgical excision with clear margins, given its potential for malignant transformation.

 Recurrence has been documented in the literature. Kersting and Helwig reported 11 of 98 patients experienced recurrence of the tumor 2 weeks to 8 years after the initial excision, but felt the regrowth may have been secondary to incomplete excision.

 Malignant transformation of eccrine spiradenoma is a rare phenomenon, but can occur within a long-standing initially benign solitary lesion. Enlarging, increasingly painful, and/or ulcerative lesions may signify malignant transformation. Malignant ES is quite aggressive, with high recurrence rate and subsequent metastasis that can prove fatal. Historical nomenclature of malignant ES has included malignant eccrine spiradenoma, sweat gland carcinoma, eccrine spiradenocarcinoma, and ex-ecrine spiradenoma. Histologically, markedly atypical cells with frequent mitotic changes can be seen. Malignant ES metastasizes via lymphatic and hematogenous routes. The primary treatment of malignant ES consists of wide local excision with intraoperative frozen sections, given the tumor’s potential for infiltration into normal tissues. If tumor metastases are suspected, regional lymphadenectomy should be performed. The roles of adjuvant radiation therapy, and chemotherapy have yet to be established. The average survival of malignant ES is approximately 36 months after diagnosis.