Farabi Tıp Dergisi 3(3):96-101,2024
Farabi Medical Journal 3(3):96-101,2024
https://doi.org/10.59518/farabimedj.1456392

FARABİ TIP DERGİSİ
FARABI MEDICAL JOURNAL

Olgu Sunumu
Case Report

https://dergipark.org.tr/tr/pub/farabimedj

Skin Spiradenocarcinoma in an Adult Patient: A Case Report

Yetişkin Bir Hastada Cilt Spiradenokarsinomu: Bir Olgu Sunumu

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ABSTRACT

This case report provides important information about spiradenocarcinoma, a rare skin tumor. It underscores the importance of understanding and recognizing this rare malignant tumor, particularly given its potential for aggressive behavior and metastasis. The fact that spiradenocarcinoma often arises from pre-existing benign tumors like spiradenoma highlights the significance of monitoring and evaluating such lesions for any signs of malignant transformation. The case of the 73-year-old male patient with spiradenocarcinoma, especially occurring after the diagnosis of two separate skin cancers, underscores the need for vigilance in patients with a history of skin malignancies. Additionally, the presentation of nodular lesions on the left lower eyelid demonstrates the potential variability in the location of these tumors, which can occur in various sites across the body. The aggressive nature of spiradenocarcinoma, coupled with its high recurrence rate, emphasizes the challenges in managing this condition. While wide local excision is typically recommended as the primary treatment modality, the uncertain effectiveness of adjuvant therapies underscores the need for further research to optimize treatment strategies and improve outcomes for patients with spiradenocarcinoma. Overall, this case report contributes valuable insights into the clinical characteristics and management of spiradenocarcinoma, highlighting the importance of early detection, thorough evaluation, and multidisciplinary management of this rare malignant tumor.

Keywords: Skin cancer, Spiradenocarcinom, Treatment

ÖZET

Bu olgu sunumu nadir görülen bir deri tümörü olan spiradenokarsinom hakkında önemli bilgiler sunmaktadır. Özellikle agresif davranış ve metastaz potansiyeline sahip olan bu nadir malign tümörün anlaşılması ve tanınmasının önemini vurgulamaktadır. Spiradenokarsinomun sıklıkla spiradenoma gibi mevcut iyi huylu tümörlerden kaynaklanması bu lezyonların malign dönüşüm belirtileri açısından izlenmesi ve değerlendirilmesinin önemini ortaya koyar. 73 yaşındaki erkek hastanın spiradenokarsinomuyla ilişkili olarak, özellikle iki ayrı cilt kanseri teşhisi konulduktan sonra ortaya çıkan durum, cilt maligniteleri öyküsü olan hastalarda dikkatli olunması gerektiğini vurgulamaktadır. Ayrıca, sol alt göz kapağında nodüler lezyonların olması, bu tümörlerin vücudun çeşitli bölgelerinde meydana gelebilecekleri potansiyel çeşitliliği göstermektedir. Spiradenokarsinomun agresif doğası, yüksek nüks oranıyla birleştiğinde, bu durumun yönetilmesindeki zorlukları ortaya koymaktadır. Genellikle ana tedavi modalitesi olarak geniş yerel eksizyon önerilse de, adjuvan tedavilerin belirsiz etkinliği, spiradenokarsinomlu hastaların tedavi stratejilerini optimize etmek ve sonuçları iyileştirmek için daha fazla araştırmaya ihtiyaç olduğunu ortaya koyar. Genel olarak bu vaka raporu, spiradenokarsinomun klinik özelliklerine ve tedavisine ilişkin değerli bilgiler sunarak, bu nadir malign tümörün erken teşhisinin, kapsamlı değerlendirmesinin ve multidisipliner yönetiminin önemini vurgulamaktadır.

Anahtar Kelimeler: Cilt kanseri, Spiradenokarsinom, Tedavi

Geliş Tarihi/Received Date: 21.03.2024 | *Kabul Tarihi/Accepted Date: 13.07.2024*

INTRODUCTION

Eccrine glands, also known as cholinergic sweat glands in the skin, play a crucial role in thermoregulation. In 1956, Kersting and Helwig identified a benign tumor of these sweat glands, termed eccrine spiradenoma.[1] Typically presenting as solitary skin tumors, spiradenomas can also manifest as multiple small skin nodules. While these benign lesions commonly emerge in early adulthood, they may display an aggressive clinical course. Although rare, malignant transformation can occur in longstanding, isolated spiradenomas.[2] Terms used to denote this transformation include malignant eccrine spiradenoma (MES), eccrine carcinoma ex-adenoid spiradenoma, and eccrine spiradenocarcinoma (SPC). This exceedingly uncommon malignant tumor, primarily originating from benign spiradenoma, seldom arises de novo from dermal appendages.[3] The English literature has reported 102 cases of MES in all. According to recent accounts, there appears to be little gender predisposition and that the tumor primarily affects older people.[4]

MES represents a rare and aggressive tumor emerging from the eccrine sweat glands, although apocrine markers have been detected in some cases.[3,5] Typically affecting regions such as the head and neck, extremities, or trunk, MES tends to manifest more frequently in older individuals, with a mean onset age of approximately 59 years, and it occurs equally in both sexes.[6] One of the notable features of MES is its aggressive nature, with a tendency to metastasize to nearby lymph nodes and distant sites such as bones, lungs, and even the brain.[6,7]

The duration required for malignant transformation of a skin lesion like MES can vary considerably, ranging from as swiftly as 6 months to as protracted as 70 years.[3] Malignant transformation denotes the process whereby a previously stable or benign skin lesion undergoes changes culminating in malignancy, often accompanied by symptoms such as pain and ulceration.[3, 6]

The rarity and diverse presentation of MES render staging and diagnostic processes particularly challenging. While advancements in imaging techniques have facilitated diagnosis and staging, histopathological examination remains pivotal for confirmation. Typically, a biopsy of the lesion is indispensable for obtaining tissue samples for histological analysis, which can unveil the malignant nature of the lesion and furnish critical insights for staging and treatment planning. Histopathological analysis for confirmation can be intricate. High-grade carcinomas may exhibit an abrupt transition between benign spiradenoma and SPC areas, whereas lower-grade carcinomas may necessitate ancillary tests like immunohistochemical staining for markers such as Ki-67, p53, and S100.[6,8,9] MRI and FDG PET-CT are commonly advocated imaging modalities for staging MES, furnishing comprehensive information about tumor extent and potential metastases.[6,10]

As for the prognosis of high-grade SPC, it carries a notable mortality rate. Local tumor recurrence rates post-initial treatment can surpass 20%, with a median recurrence time of approximately 12 months.[6,7,11,12] However, it's crucial to acknowledge that the literature on SPC primarily comprises case reports, short case series, and reviews due to its rarity, precluding the establishment of definitive treatment standards.

While treatment strategies for high-grade SPC remain ill-defined owing to the scarcity of cases, surgical excision is commonly advocated as the primary treatment modality.[6,7,13-15] The role of adjuvant external beam radiotherapy, chemotherapy, and/or regional lymph node dissection remains uncertain and may hinge on individual case characteristics.

Given its rarity and aggressive nature, treatment of MES can pose clinical challenges and often requires a multidisciplinary approach including surgical excision, radiation therapy, and sometimes chemotherapy. Given the risk of recurrence, meticulous monitoring and multidisciplinary management involving oncologists, surgeons, and other specialists are often recommended to tailor treatment plans to each patient's specific needs. Careful monitoring and timely intervention are mandatory for effective management of this condition.

CASE REPORT

The patient, aged 73, with a medical history including bypass surgery in 2012, diabetes mellitus, hyperlipidemia, and arrhythmia, presented with non-healing wound-like lesions on the left eyebrow skin area in 2007 and excisional biopsy was performed. The pathology report indicated SCC with clear surgical margins, and no adjuvant therapy was considered; the patient was put under follow-up. In 2009, an excisional biopsy was performed on a lesion in the lower left eyelid. The pathology revealed carcinoma in situ with a 1 mm surgical margin. In 2023, due to recurrence in the

lower left eyelid, another excisional biopsy was performed, and the pathology report indicated a basaloid malignant epithelial tumor. One year later, on March 22, 2024, the recurrent mass was excised again, and the pathology was reported as spiradenocarcinoma. Since the surgical margin was 1 mm, the patient was referred to the Radiation Oncology clinic, and post-operative radiotherapy was planned.

According to the pathology report, the histopathological characteristics of neoplastic cells were as follows: BerEp4 (+), EMA (+), CK20 (-), INSM-1 (-), CEA-mono (-), CK7 (-), p63 (+), AR (-), Adipophilin (-), p53 (-), S100 (-) (figure 1). In the FDG PET-CT scan performed on the patient, no evidence of distant metastasis was detected. Due to SPC's rarity and aggressiveness, comprehensive treatment planning is necessary. A multidisciplinary approach involving surgical oncologists, radiation oncologists, medical oncologists, and other specialists is often essential. Treatment may require surgical excision, radiation therapy, and possibly chemotherapy, considering disease extent, metastasis, and patient factors. Treatment objectives include local disease control, relapse prevention, and survival improvement, considering the patient's age, medical history, and overall health. The patient underwent 50 Gray radiotherapy and received close monitoring and regular follow-up to assess treatment outcome, detect recurrence or metastasis, and provide supportive care (figure 2).

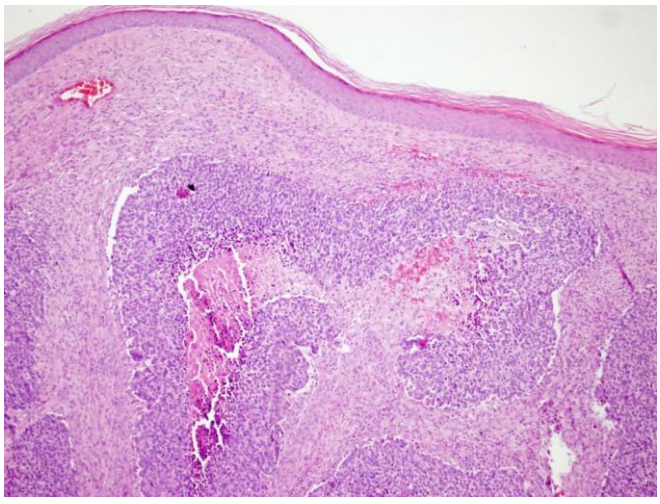

Figure 1. Regions of spiradenoma free of atypia and displaying its characteristic nodular appearance

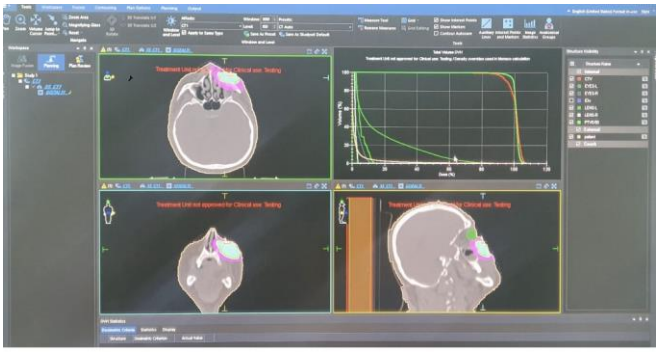

Figure 2. Treatment planning

DISCUSSION

SPC is indeed an exceptionally rare malignant tumor originating from the eccrine sweat glands. It was first described by Dabska et al. in 1971.[2] Typically, SPC arises from a benign precursor lesion known as spiradenoma, although there have been sporadic reports of de novo development without a preceding benign lesion. Histologically, SPC is characterized by the malignant transformation of eccrine cells.[3] However, it's noteworthy that in some cases, apocrine markers have also been identified, indicating a possible differentiation towards apocrine sweat glands. Due to the rarity of SPC and its varied histological features, accurate diagnosis often requires a combination of clinical, histopathological, and sometimes immunohistochemical examinations. Additionally, given the aggressive nature of SPC, prompt diagnosis and appropriate management are crucial for optimizing patient outcomes. Treatment typically involves a multidisciplinary approach, including surgical excision, radiation therapy, and possibly chemotherapy, tailored to the individual patient's needs and disease characteristics.

SPC typically manifests as nodular lesions on the trunk, extremities, or head and neck region.[3] While the incidence of this malignancy is comparable between genders, it tends to occur more frequently in older individuals, with a typical age of onset around 59 years.[16] The malignant transformation of spiradenoma into SPC is associated with aggressive behavior. It often exhibits a high recurrence rate and has the potential to develop fatal metastases.[17] Due to its aggressive nature, SPC requires prompt diagnosis and appropriate management to optimize patient outcomes.

Treatment usually involves a multidisciplinary approach, including surgical excision, radiation therapy,

and possibly chemotherapy, tailored to the individual patient's needs and disease characteristics. Close monitoring and regular follow-up are essential to assess treatment response, detect any recurrence or metastases early, and provide timely intervention as needed.

Metastases in SPC typically involve sites such as the bone, lung, liver, and brain, but lymphatic infiltration can significantly worsen the prognosis.[7,11] While metastases may not be initially detected, the likelihood of their occurrence increases if sarcomatous changes are found on histopathological analysis. Sarcomatous changes have been reported in 21–57% of SPC cases and are associated with a higher risk of metastasis and poorer prognosis.[18,19] In the case you mentioned, the absence of sarcomatous foci or syringoma on histopathological analysis is a positive finding, as it suggests a lower likelihood of metastasis. However, it's essential to remain vigilant for any signs of recurrence or metastasis during follow-up evaluations. Regular monitoring, including clinical examinations and possibly imaging studies, will be crucial to detect any potential disease progression early and initiate appropriate treatment interventions if necessary. Additionally, a multidisciplinary approach involving oncologists, radiologists, and other specialists may be necessary to ensure comprehensive management and optimize patient outcomes.

Surgical excision with wide margins is indeed considered the mainstay of treatment for SPC. This approach aims to achieve complete removal of the tumor and surrounding tissue to minimize the risk of local recurrence. Several studies, including those by Andreoli et al. have reported a 100% survival rate with wide surgical excision in cases where there is no evidence of metastasis.[7] The role of adjuvant treatments such as external beam radiotherapy and chemotherapy in the management of SPC remains unclear and controversial. Some studies suggest that chemotherapy is typically reserved for cases where the cancer has spread to other organs such as the brain.[7,11] However, the decision to use adjuvant therapies should be made on a case-by-case basis, taking into consideration factors such as the extent of the disease, tumor characteristics, and the patient's overall health status. Further research and clinical trials are needed to better understand the role of adjuvanttreatments in improving outcomes for patients with SPC.

The diagnosis of SPC typically requires histopathological analysis, as it can be challenging to distinguish between benign and malignant lesions based solely on clinical presentation. Histological examination allows for the evaluation of cellular characteristics and tissue architecture, which are crucial for accurate diagnosis. Immunohistochemical staining for specific markers can also be helpful in distinguishing SPC from benign lesions such as spiradenoma. Markers commonly used in this context include Ki-67, p53, and S100. Ki-67 is a marker of cellular proliferation and can help assess the growth fraction of the tumor.[3] Elevated Ki-67 expression may indicate increased cellular proliferationand thus suggest malignancy. p53 is a tumor suppressor gene that plays a role in regulating cell cycle progression and apoptosis. Abnormal expression or mutation of p53 protein is associated with many malignancies, including SPC. S100 is a marker commonly expressed in cells derived from the neural crest, including sweat gland cells. While S100 staining alone may not differentiate between benign and malignant lesions, its expression pattern in conjunction with other markers can provide additional diagnostic information. The combination of histopathological analysis and immunohistochemical staining helps pathologists accurately diagnose SPC and differentiate it from benign lesions. This information is crucial for guiding treatment decisions and predicting prognosis in patients with suspected SPC.

Indeed, imaging methods such as MRI (Magnetic Resonance Imaging) and FDG PET-CT (Positron Emission Tomography with Fluorodeoxyglucose) are commonly recommended for tumor staging in SPC.[7,16] These modalities provide valuable information about the extent of the tumor and the presence of any metastases. High-grade SPC is characterized by a high rate of local recurrence and metastasis. Due to the rarity of SPC, treatment guidelines are currently lacking. Management strategies are generally based on case reports, small case series, and reviews, as mentioned by Andreoli and Gupta et al. (2000).[7,17] Given the limited evidence and variability in clinical presentations, more research is needed to establish standardized treatment protocols and improve outcomes for patients with SPC. Multidisciplinary collaboration among oncologists, surgeons, radiologists, and other specialists is essential to tailor treatment plans to individual patients and optimize their care. Clinical trials and prospective studies may help to further elucidate the optimal management strategies for SPC. This case report highlights the presentation, diagnosis, and management

of SPC, a rare skin tumor. The presence of nodular lesions on the left lower eyelid in the patient's history is significant, as it raises suspicion for malignancy. The evolution of the tumor and treatment options described in the literature support the clinical findings observed in this case. Wide local excision is highlighted as a primary treatment option, but it's noted that treatment standards for SPC have not been firmly established due to its rarity. This underscores the importance of individualized treatment plans based on the patient's specific circumstances and tumor characteristics.

The case report also suggests that SPC should be considered in the differential diagnosis of eyelid tumors. Additionally, it acknowledges that in certain cases where there is a low risk of metastasis or a short life expectancy, observation rather than aggressive surgical intervention may be a viable option. Furthermore, the challenges in managing SPC are highlighted, emphasizing the need for further research to better understand the optimal diagnostic and treatment strategies for this rare malignancy. This case serves as a reminder of the complexity involved in managing rare skin tumors like SPC and the importance of ongoing research efforts to improve patient outcomes.

Authorship contribution statement

Consept and design: MK, FRY.

Acquisition of data: MK, FRY.

Analysis and interpretation of data: MK, FRY, ŞE, RBG.

Drafting of the manuscript: MK, FRY, ŞE, RBG.

Critical revision of the manuscript for important intellectual content: MK, FRY.

Ethical approval/Informed Consent

Written informed consent was obtained from the patient who participated in this case.

Declaration of competing interest

None of the authors have potential conflicts of interest to be disclosed.

Availability of data and materials

All data generated or analyzed during this study are included in this published article.

Funding

The authors declared that this study has received no financial support.

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To Cite: Yesilyurt FR, Gulcebi RB, Ersoz S, Kandaz M. Skin Spiradenocarcinoma in an Adult Patient: A Case Report. Farabi Med J. 2024;3(3):96-101. doi:10.59518/farabimedj.1456392