Case Report / Olgu Sunumu

Malignant eccrine spiradenoma of the external ear

Dış kulağın malign ekrin spiradenoması

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ABSTRACT

Eccrine spiradenoma is a benign tumor of sweat gland origin. Malignant eccrine spiradenoma is a rare neoplasm which almost always arises from a pre-existing spiradenoma lesion. In this article, we present a patient with malignant eccrine spiradenoma of the auricula with a prior diagnosis as benign eccrine spiradenoma. Although malignant transformation is usually associated with aggressive behavior, we did not detect recurrence or metastasis in four years of follow-up after performing wide excision.

Keywords: Auricula; eccrine spiradenoma; malignant.

ÖZ

Ekrin spiradenoma ter bezi kökenli benign bir tümördür. Malign ekrin spiradenoma neredeyse daima daha önceden var olan bir spiradenoma lezyonundan gelişen nadir bir neoplazmdır. Bu yazıda daha önce benign ekrin spiradenoma tanısı konulmuş, dış kulak malign ekrin spiradenoması olan bir hasta sunuldu. Malign dönüşüm genellikle saldırgan davranış ile ilişkilendirilse de geniş eksizyon sonrası dört yıllık takipte nüks ve metastaz tespit edilmedi.

Anahtar Sözcükler: Aurikula; ekrin spiradenoma; malign.

Eccrine spiradenoma is a benign tumor of sweat gland origin that usually consists of a firm, rounded, bluish dermal nodule which is usually seen on the trunk and limbs.[1] It is very rare in the external ear.[2]

Malignant eccrine spiradenoma is a very rare tumor that is considered to originate from a pre-existing benign eccrine spiradenoma, although de novo lesions have also been reported.[3] The incidence is similar in both sexes and the age of patients ranges between 21 to 92 years with the mean age of 55.5 years.[4]

We report a case of malignant eccrine spiradenoma in the external ear, originated from benign eccrine spiradenoma.

CASE REPORT

A 67-year-old male who had previous excision of eccrine spiradenoma from the right cavum conchae was referred to our department. He complained of a crusty persistent, itchy lesion in his right ear. It was occasionally accompanied by a sensation of ear fullness. On physical examination there was a reddish nodular lesion

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in the right auricle, located in the cavum conchae extending to the ear canal (Figure 1). The crus of the helix and entrance of the external ear canal were also involved. The lesion was 1x1.5 cm in diameter, wet-looking, crusty, and slightly protuberant. There was no sign of regional or systemic metastases on examination. The patient had been referred to us two years ago with similar complaints and similar findings. The lesion was excised and pathological examination revealed eccrine spiradenoma (Figure 2). He did not follow-up for two years, although we learned that the lesion recurred approximately six-months after the excision. It was a millimetric rash at the beginning of the recurrence and expanded slowly for two years but the enlargement of the lesion accelerated during the last three months. A high resolution computed tomography of the ear demonstrated pathological contrast enhancement of the lesion described but there was no bone involvement. We performed a wide excision of the lesion. The defect was left to heal by secondary intention. On histopathological examination there was ulceration on one side of the specimen which was covered by stratified squamous epithelium. Tumor infiltration was observed on the ulcerated site, reaching the dermis. The tumor consisted of hyperchromatic nucleated atypical cells with prominent nucleoli and narrowed cytoplasm. There was mild pleomorphism and atypical mitoses (Figure 3a). The tumor predominantly consisted of solid structures. It was positive for epithelial membrane antigen (EMA) (Figure 3b) and S-100 whereas carcinoembryonic antigen (CEA) and P53 were nonreactive. We observed Alcian blue positivity with periodic acid-Schiff-Alcian blue (PAS/AB) in the stroma of the tumor. Considering the previous diagnosis, it was diagnosed as the malignant transformation of pre-existing eccrine spiradenoma. We did not observe any further sign of recurrence over four years follow-up.

DISCUSSION

Eccrine spiradenoma is a benign tumor of sweat gland origin which was first described in 1956 by Kersting and Helwig.[1] It usually occurs on the trunk or extremities, as a hard knot in the skin and is occasionally associated with pain and tenderness.[5] Malignant eccrine spiradenoma is a very rare carcinoma of the skin originating from sweat glands first reported by Dabska in 1972.[6] It usually originates from a pre-existing benign eccrine spiradenoma, as in the present case, although de novo lesions have also been reported.[3]

Malignant eccrine spiradenoma can be seen in various parts of the skin, however, to the best of our knowledge this is the second reported case of malignant eccrine spiradenoma originating from the external ear.[7]

Before malignant transformation of eccrine spiradenoma patients often describe a small lesion that is silent for years that begins to grow

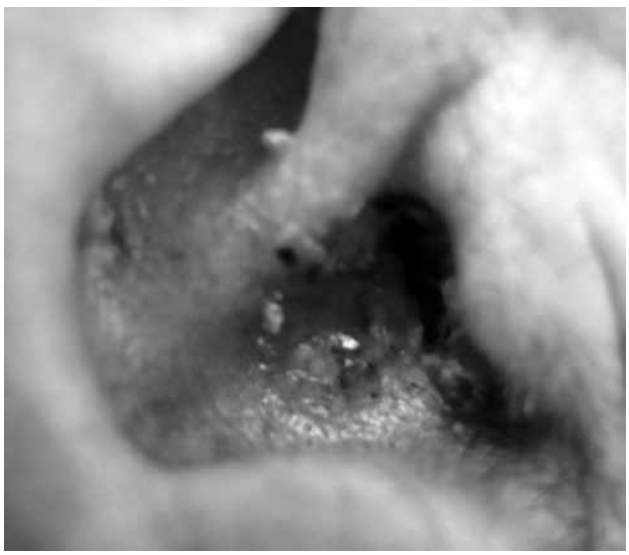

Figure 1. *The lesion located in the right ear. It is crusty wet looking and nodular.*

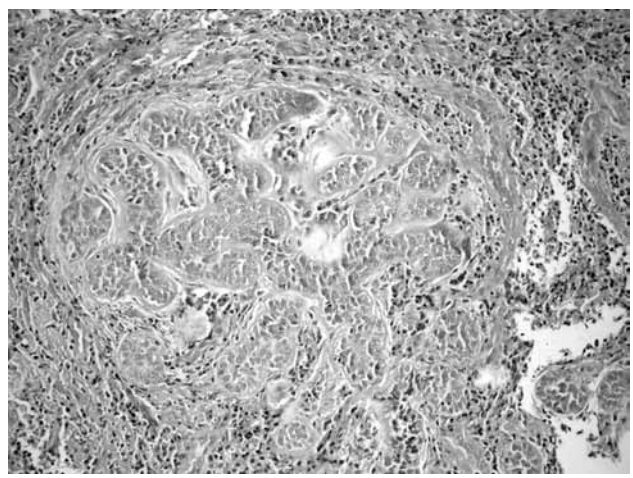

Figure 2. *Benign eccrine spiradenoma. Uniform looking tumor cells located in dermis constitute nodular lesions surrounded by hyaline material (H-E x 100).*

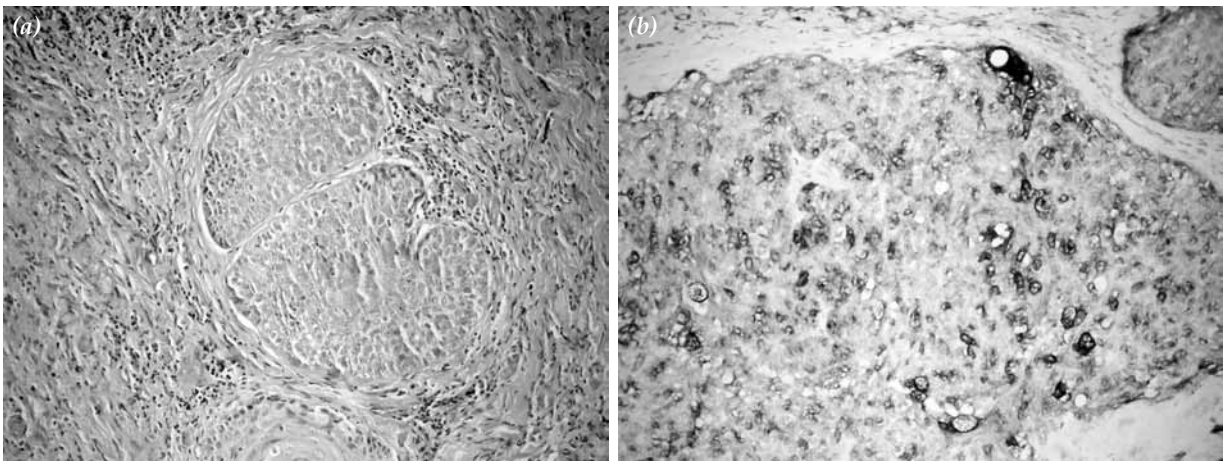

Figure 3. *Malignant eccrine spiradenoma* ***(a)*** *Atypical tumor cells constituting solid masses which infiltrate dermis (H-E x 100).* ***(b)*** *Epithelial membrane antigen positivity in tumoral cells (EMA x 200).*

in size or becomes painful.[7] Rapid enlargement, pain, and change in color often prompt the patient to seek medical attention.[8] This case had also experienced similar symptoms except for pain over two years after the recurrence and there was acceleration in the growth of the lesion for the last few months. The latency period was approximately two years in our case. There are reports where the latency period before transformation into malignant disease ranges from six months to 70 years.[9]

In lesions of malignant eccrine spiradenoma, the benign and carcinomatous regions associated with areas of transition are typically seen.[8,10] Lesions consisting of well-demarcated zones of spiradenoma and spiradenocarcinoma have also been described.[8] We must keep in mind that the areas of malignant transformation can be very small and can remain undetected if the lesion is not completely excised.[4] Because the malignant changes may be focal in the lesion inadequate excision may leave residual malignant tumor.[11]

Malignant transformation of eccrine spiradenoma is associated with aggressive behavior, high recurrence rate, and subsequent development of fatal metastases.[7] Metastases usually affect the lymph nodes, bones, and lungs.[4] We did not observe any sign or symptom of recurrence or metastasis in four years of follow-up.

There are around 100 reported cases of malignant eccrine spiradenoma in the literature. In a meta-analysis conducted by Andreoli et al.[12] in 2011, 102 patients reported in the English literature as malignant eccrine spiradenoma were surveyed and 72 that had sufficient data were analyzed. Among these patients 36 were nonmetastatic, 12 had lymph node metastasis without distant metastasis, and 24 had distant metastasis. Thirty-six patients who had no metastasis were treated with local resection of the lesion and had a disease free survival rate of 100% with a mean follow-up of 33 months. Patients with distant metastasis had a median survival of 16 months. They found that in patients with distant metastasis median survival time was longer when there was adjuvant therapy to the surgery.[12] However it is stated that neither radiotherapy nor chemotherapy which are given adjuvant to surgery has proven to be effective in controlling the disease progression.[4]

S100 protein, which stains for myoepithelium of the sweat gland or eccrine sweat gland, CEA, which is a tumor marker of the sweat organ, epithelial membrane antigen, or p53 protein may provide some assistance in diagnosis. However, these patterns of expression vary case-by-case.[5]

Conclusion

Malignant eccrine spiradenoma is a rare skin appendage tumor and the ear is an unusual location. Wide surgical resection with is essential for management. Adjuvant radiotherapy or chemotherapy may be of choice for recurrent or metastatic tumors. Although eccrine spiradenoma rarely transform into malignant

form, total resection of the lesion is mandatory, in order not to leave malignant foci in a benign looking lesion.

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