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Sebaceous carcinoma of scalp with parietal bone destruction: A rare case presentation

Parietal kemik yıkımlı saçlı derinin sebasöz karsinomu: Nadir bir olgunun sunumu

Kafil Akhtar [1], Noora Saeed [1], Sumbul Warsi [1], Shafaque Zabin [1]

[1] Department of Pathology, Jawaharlal Nehru Medical College, Aligarh Muslim University, Aligarh (U.P), India

ORCID ID of the author(s)

KA: 0000-0002-3482-1195
NS: 0000-0002-3245-1234
SW: 0000-0002-1234-2134
SZ: 0000-0002-4321-3214

Abstract

Sebaceous carcinoma is a rare malignant tumor of the skin appendages, which occurs in the elderly. The most commonly involved site is the orbital region and it is rarely seen in the extra-orbital sites. We report a rare case of extra-orbital sebaceous carcinoma of the scalp with parietal bone destruction in a 45-year-old male. He was treated with wide local excision of the tumor with no regional or distant metastases. The patient received adjuvant chemo-radiation therapy, with no evidence of any local recurrence after one year of follow-up period. It is imperative to diagnose sebaceous carcinoma at an early stage to execute the primary treatment i.e., wide local excision and prevent recurrence of the tumor.

Keywords: Scalp, Sebaceous carcinoma, Bone destruction, Histopathology

Öz

Sebasöz karsinom, yaşlılarda ortaya çıkan, deri eklerinin nadir görülen kötü huylu tümörüdür. En sık tutulan bölge orbita bölgesidir ve orbita dışı bölgelerde nadiren görülür. 45 yaşında bir erkekte pariyetal kemik yıkımı ile ilerleyen, kafa derisinde orbita dışı sebasöz karsinomlu nadir bir olguyu sunuyoruz. Bölgesel veya uzak metastaz olmaksızın, tümörün geniş lokal eksizyonu ile tedavi edildi. Hasta, bir yıllık takip süresinden sonra herhangi bir lokal nüks olmaksızın adjuvan kemo-radyasyon tedavisi aldı. Birincil tedaviyi, yani geniş lokal eksizyonu gerçekleştirmek ve tümörün nüksetmesini önlemek için sebasöz karsinomu erken aşamada teşhis etmek zorunludur.

Anahtar kelimeler: Saçlı deri, Sebasöz karsinom, Kemik yıkımı, Histopatoloji

Introduction

Sebaceous carcinoma is a rare aggressive malignant tumor derived from the adnexal epithelium of sebaceous glands, seen frequently in the Asian population [1,2]. It commonly occurs in the peri-ocular region. Extra orbital sebaceous cell carcinoma is extremely rare, occurs on head and scalp due to the presence of abundant sebaceous glands [3]. Most sebaceous carcinomas have no obvious etiology, but few are associated with Muir-Torre syndrome and thought to arise from sebaceous glands in the skin [4].

Sebaceous carcinoma is classified into 2 groups, those arising from the ocular adnexa, particularly the Meibomian glands and glands of Zeiss, and those arising in extra ocular sites [4]. Ocular sebaceous carcinomas comprise 1% to 5.5% of all eyelid malignancies [5]. These tumors have a high incidence of local recurrence and regional metastasis. Early diagnosis is crucial in reducing the morbidity and mortality associated with the tumor [6]. We report a case of sebaceous carcinoma of the scalp with parietal bone destruction in a 45-year-old male, who presented with a painful gradually increasing scalp swelling over the parietal bone for the last 4 months.

Corresponding author / Sorumlu yazar:
Kafil Akhtar
Address / Adres: Department of Pathology, Jawaharlal Nehru Medical College, Aligarh Muslim University, Aligarh (U.P), India
E-mail: drkafilakhtar@gmail.com

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Case presentation

A 45-year-old male presented to the surgical outpatient clinic with a painful, gradually increasing scalp swelling over the parietal bone for the last 4 months. There was no discharge, fever, or any family history of malignancy. On examination, a solitary ill-defined tender erythematous mass measuring 4x2 cm was observed over the left parietal bone. The mass was mobile and firm to cystic in consistency. No regional lymph nodes were palpable and systemic examination was unremarkable. His routine blood investigations were within normal limits. Skull X-ray showed lysis of the parietal bone. CT scan of the head, neck, thorax, and abdomen was performed, which showed no evidence of distant metastasis, but destruction of the parietal bone. Magnetic resonance imaging of the brain revealed a small focal area of altered signal within the scalp in left posterior parietal region with cortical destruction of the underlying bone.

A wide local excision of the scalp swelling with local template flap was performed. Macroscopically, a polypoidal skin covered mass measuring 4x3 cm was seen. The cut section showed lobulated solid to cystic grey-white mass containing gelatinous material. Microscopically, the tumor comprised large round to polygonal cells arranged in trabeculae, nests, and cords in a hyalinized stroma, with abundant clear cytoplasm and oval hyperchromatic nuclei with distinct nucleoli (Figure 1 and 2) PAS stain showed strong positivity (Figure 3). On immunohistochemistry, the tumor cells showed cytoplasmic positivity for cytokeratin (Figure 4) and focal positivity for epithelial membrane antigen (EMA). The microscopic features demonstrated a malignant skin appendageal tumor compatible with sebaceous carcinoma. With the intent of decreasing the chances of local recurrence, adjuvant radiotherapy in the dose of 50 Gy in 20 fractions was delivered over 4 weeks using electron beam therapy. Currently, he has completed a follow-up period of one year after therapy, with no evidence of disease.

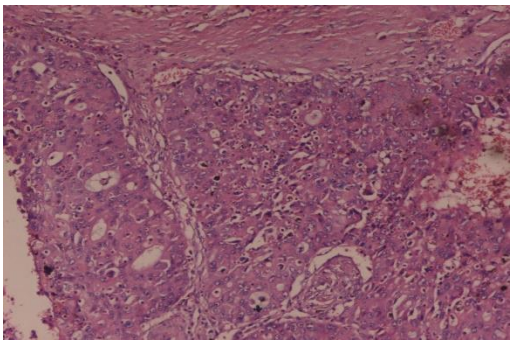

Figure 1: Microscopically, the tumor showed large round to polygonal cells arranged in trabeculae, nests, and cords in a hyalinized stroma, with abundant clear cytoplasm and oval hyperchromatic nuclei with distinct nucleoli. Hematoxylin and Eosin x 10X.

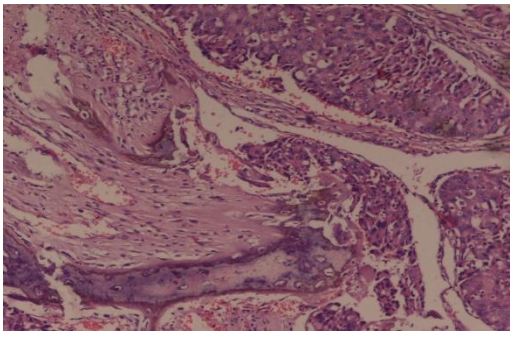

Figure 2: Section shows malignant cells in distinct lobules with strip of lysed bone. Hematoxylin and Eosin x 40X.

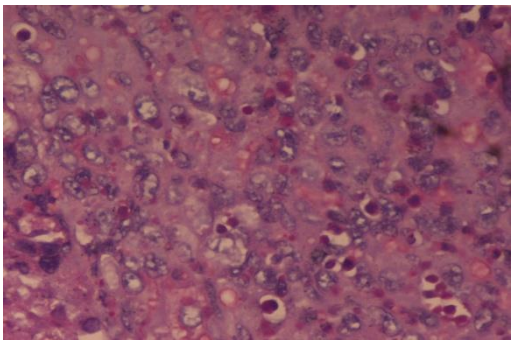

Figure 3: Section showed strong cytoplasmic PAS positivity. PAS stain x 40X.

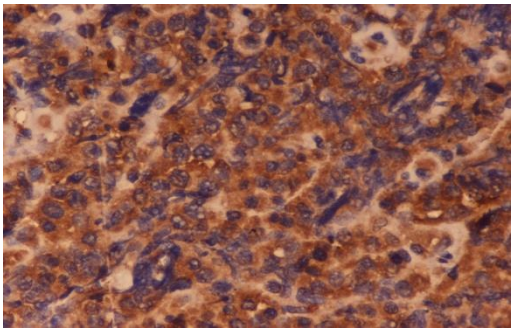

Figure 4: Immunohistochemistry showed strong cytoplasmic positivity for cytokeratin. IHC Cytokeratin x 40X.

Discussion

Sebaceous gland carcinoma is a rare aggressive cutaneous tumor, which occurs in the elderly [1]. The most frequent extra-orbital site for this tumor is skin in the head and neck region wherein sebaceous glands are most abundant [1]. Peri-orbital sebaceous carcinoma is three-times more common than the extra-orbital carcinoma. Extra-orbital tumors are known to show rapid growth with distant metastases [5].

Sebaceous carcinoma may occur in pre-existing dermatoses like naevus sebaceous and actinic keratosis or post radiation therapy for some cancerous diseases [6]. It has been associated with Muir-Torre syndrome which is an autosomal dominant dermatosis consisting of sebaceous neoplasms like sebaceous adenoma and sebaceous carcinoma, with associated visceral malignancy in the same individual, without any precipitating cause such as radiotherapy or AIDS [7]. Our patient was screened for Muir Torre syndrome with negative colonoscopy, ultrasonography of abdomen and chest and routine and microscopic urine examination findings.

Sebaceous carcinoma presents as a solitary, erythematous or pale yellow-colored, firm to hard, slowly growing nodule, with ulceration. It is seen with an increased frequency in the Asian population [2]. The gender predisposition of extraocular sebaceous carcinoma is equal in males and females, with mean occurrence age of 63 years [3]. This malignancy occurred in our patient at an early age of 45 years.

The four common histological patterns reported are lobular, comedo-carcinoma, papillary and mixed. The main histological picture is the lobular architecture, with cells showing marked nuclear pleomorphism and foamy vacuolated cytoplasm [4]. Some mature cells may show multiple cytoplasmic vacuoles with scalloped nuclei. Fat stains on frozen sections demonstrate fine lipid globules. Necrosis may be also present in the center of the tumor [8]. Siddhi et al reported local recurrence rates of 29.0%, regional nodal metastasis in 15%, and a disease-related

mortality of 20.0% in 91 cases with extra-ocular sebaceous carcinoma [4]. Adverse prognostic factors include poor differentiation, infiltrative growth pattern, multicentric origin of the tumor and size of the tumor more than 10 mm. Samarasinghe et al. have reported a case of sebaceous carcinoma of scalp with nodal metastasis [2].

The tumors of sebaceous glands are separated into three major categories: Sebaceous adenoma, basal cell carcinoma with sebaceous differentiation and sebaceous carcinoma [1]. Sebaceous carcinoma cells are large and may show squamous changes, and it should be differentiated from squamous cell carcinoma by hydropic changes in the cell cytoplasm. Tumor cells may show basaloid differentiation with presence of inconspicuous lipid vacuoles, and hence the tumor must be distinguished from basal cell carcinoma with sebaceous differentiation [9]. Immunohistochemical staining for EMA can differentiate sebaceous carcinoma from basal cell carcinoma and squamous cell carcinoma [10].

Complete surgical excision is the treatment of choice [11]. Sebaceous carcinomas tend to show 9 to 36% local recurrence within 5 years [12]. Metastasis to liver, lungs, bones, and brain have been reported in 14-25% of the patients [13]. Radiation therapy and chemotherapy have been used for regional and metastatic disease, with varying degrees of response. Only a few studies have shown satisfactory results of adjuvant chemotherapy in the treatment of sebaceous carcinoma with metastasis. There is one such report of complete response to systemic chemotherapy in metastatic sweat gland carcinoma to pleura, pericardium, and chest wall with 5‑fluorouracil after 3 months of follow up period [3].

Conclusions

Sebaceous carcinoma needs to be considered as a differential diagnosis for the cutaneous malignancies in all age groups. The timely recognition enables execution of the primary treatment i.e., wide local excision. Adjuvant radiotherapy may be considered to improve the clinical outcome of recurrent tumors.

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