Clinics in Surgery

Case Report
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Eccrine Porocarcinoma of Scalp: an Unusual Malignancy of Sweat Glands

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Abstract

Eccrine Porocarcinoma (EPC) is a very rare malignant sweat gland tumor, first described by Pinkus and Mehregan in 1963 as Epidermotropic eccrine carcinoma. The term "eccrine porocarcinoma" was later coined by Mishisma and Morikoin 1969. It is considered as the malignant counterpart of eccrine poroma, a benign tumor of the intraderamal sweat gland. Porocarcinoma is commonly found in elderly, usually in the lower extremities. The scalp is a rare site of affliction. The tumor may present as nodular, infiltrative, ulcerated or polypoid growth. Surgical resection is the treatment of choice. We report a case of EPC of scalp arising from a pigmented eccrine poroma in a 66-year-old male treated with surgery.

Case Presentation

A 66-year-old gentleman presented to surgical oncology outpatient department with complaints of non-healing ulcer over the scalp of 1 year duration (Figure 1), which according to him was growing rapidly from the past 2 months. On examination, there was an ulcer of size 10 x 8 cms in the left frontal region of the scalp, edges were everted, and was bleeding on touch. Medially it is 1 cm to right of midline, inferiorly 2 cm above the eyebrow, superiorly 12 cm from the eyebrow with no associated regional lymphadenopathy. An incisional edge biopsy was done which was consistent with infiltrating carcinoma with possibilities of either squamous cell carcinoma or skin adnexal carcinoma with squamous differentiation. The Computed tomography scan of the skull revealed irregular ill-defined soft tissue density lesion measuring 6.2x7x1 cm in the left frontal region of scalp with no bone erosion or regional adenopathy (Figure 2). A wide local excision of the tumour with 1 cm of gross margins all around was done. The defect was reconstructed with a rotation flap, the donor area being covered with split thickness skin graft (Figure 3). The final histopathology report was consistent with eccrine porocarcinoma of the scalp (Figure 4). Microscopy showed structure of skin with foci of ulceration, multiple foci of infiltrating cellular lesion arising from the basal layer of the skin, the cells are polygonal with granular formations, central keratinisation and foci showing intercellular bridges. Cell clusters showed peripheral palisading, vesicular nucleoli with an average mitotic figure of 12 per high power field (HPF). All the resected margins were tumour free. On Immunohistochemical staining the tumor cells showed CEA and CK 6 positivity (Figure 5 and 6). Postoperatively the wound healed well and patient was treated with adjuvant radiation. At the time of last follow up he was doing well without any evidence of recurrence.

Discussion

Eccrine porocarcinoma is a rare malignant adnexal tumor arising from the intra dermal part of the sweat gland accounting for 0.005% of all epithelial cutaneous tumors. It is the malignant counterpart of common benign adnexal tumor (eccrine poroma) and is also termed as malignant hidroacanthoma simplex, eccrine poroepithelioma, malignant syringoacanthoma, dysplastic poroma. They occurring equally in both the sexes usually in older individuals [1], more than half of them are occurring in lower extremities. On rare occasion, it may involve head and neck region, upper limbs, trunk and abdomen [2,3]. Eccrine porocarcinoma may arise as de-novo or secondary to any pre-existing lesions like eccrine poroma, nevus sebaceous, chronic lymphocytic leukemia and actinic keratoses [3-6]. The most common presentation is reddish nodular cauliflower like growth or infiltrative verrucous plaque with superficial ulceration and bleeding due to trivial trauma [5]. The differential diagnosis when on extremities, should include more common entities like seborrheic

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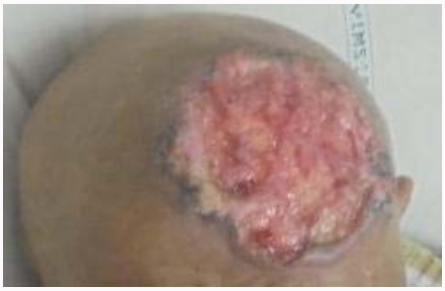

Figure 1: Clinical photograph.

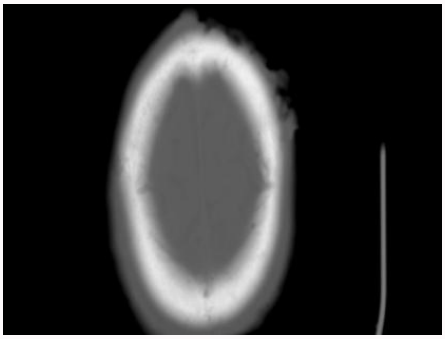

Figure 2: CT Showing soft tissu density lesion with normal bone.

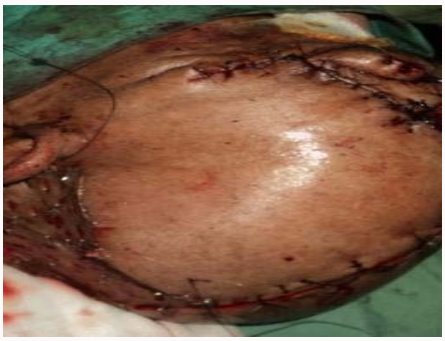

Figure 3: Scalp rotation flap covering the surgical defect.

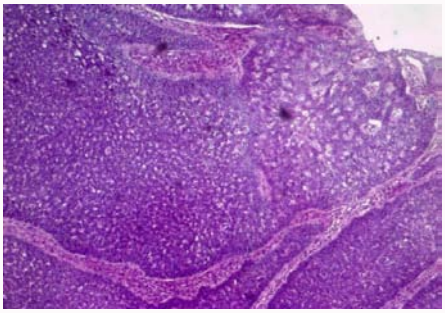

Figure 4: Microphotograph of the lesion (40X).

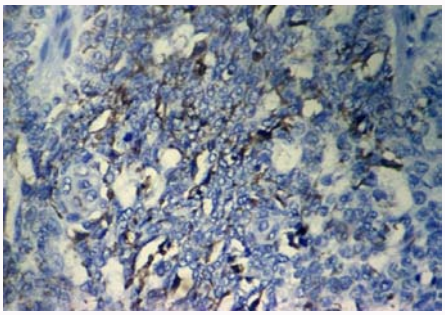

Figure 5: IHC staining.

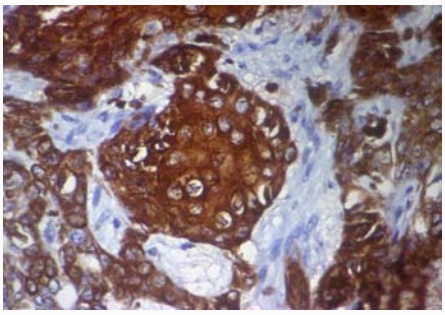

Figure 6: CK6 positivity.

keratoses, pyogenic granuloma, amelanotic melanoma, squamous cell and basal cell carcinoma and verruca vulgaris [5,7]. When the lesion is on scalp, the usual differentials include Cylindroma, eccrine poroma, sebaceous adenoma, sebaceous carcinoma, pilar tumor and metastatic carcinoma [3]. In their large series, Robson et al. [7] found that the specific clinical diagnosis was never correct on histopathological evaluation [7]. In our case it was clinically diagnosed as squamous cell carcinoma of the scalp. Microscopic appearance usually shows atypical tumor cells arranged in cords and lobules which may involve both dermis and epidermis. Tumor cell shows nuclear atypia with frequent mitosis and necrosis. Robson et al. reported few histopathological factors that are predictive for poorer clinical outcome and death [7], such as (i) presence of more than 14 mitosis per high power field (ii) lymph vascular invasion (iii) tumor depth >7 mm and (iv) infiltrating margins. Immunohistochemical features like positivity for CEA, EMA, CK-7 and negative staining for S-100, CK-20 help in the diagnosis of difficult cases where histopathological findings are not conclusive [3,7,8]. The standard treatment consists of wide surgical excision or Mohs micrographic surgery. The Surgery alone is curative in 70% of cases. The regional lymph nodes should be assessed as porocarcinoma has shown propensity to invade dermal lymphatics which cause lymph nodal disease in about 20% of the cases [3,6-9]. In case of regional lymph node involvement, lymph node dissection should be done. Adjuvant radiotherapy is necessary in margin positive cases; chemotherapy is suitable and effective for management of metastatic lesions. The reported rates of local recurrence is 17-25% and distant metastasis of 11% [3,8].

Conclusion

Eccrine porocarcinoma of scalp is a rare malignant adnexal neoplasm and those affecting scalp are still rare with less than 20 cases being reported in the literature. Local excision with negative margin is the cornerstone of management. Our case is a classical example of this rare malignancy and the diagnostic confusion and it emphasizes the inclusion of the lesion in the list of differential diagnosis during evaluation of any ulcerative lesions of the scalp. It needs a high index of suspicion and has to be managed aggressively.

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