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Case Report

A case series of atypical cutaneous malignancies in dermatology out-patient department in a tertiary care centre of eastern India

Prodip Sarkar, Ismt ara Khan, Shiladitya Chowdhury, Sahin Jaman, Dyuti Das, Ramesh Chandra Gharami, Partha Sarathi Nayak

Department of Dermatology, Medical College, Kolkata, West Bengal, India.

Abstract

The skin being a dynamic organ with multiple components, lead to malignancies of an extended spectrum. Keratinocytes, melanocytes and even the endothelial cells may turn malignant. Incidence of skin cancers has been on the rise since the last few decades worldwide. The focus has been mainly on the commonly encountered malignancies like the squamous cell carcinoma (SCC), basal cell carcinoma (BCC) and melanoma. However, certain atypical skin tumors and certain atypical presentation of the common tumors can be quite challenging clinically. Skin tumors may also be a combination of the typical ones. Malignant cutaneous appendageal tumors are quite rare and vascular tumors like hemangioendothelioma are often a forgotten entity. We have encountered four interesting atypical malignancies in our outpatient clinic which include a basosquamous carcinoma of the scalp, porocarcinoma of the hand, epithelioid hemangioendothelioma of the lower limb, and a morpheaform BCC in the neck. Knowledge about the above discussed four cases are required, so that they are not missed.

Key words

Cutaneous malignancies, basosquamous carcinoma, porocarcinoma, epithelioid hemangioendothelioma, morpheaform BCC.

Introduction

The skin is made up of cells of many kind and each of them has a potential to turn malignant. Keratinocytes, melanocytes and even the endothelial cells may start uncontrolled proliferation and lead to cancer formation. Dermatologists are quite familiar with the typical cases of cutaneous cancers like the squamous cell carcinoma (SCC), basal cell carcinoma (BCC), melanoma, to name a few. These diseases when presents in an atypical manner offer challenges in the eyes of a clinician. This case series highlights some of the less commonly encountered cutaneous malignancies.

Address for correspondence
Dr. Partha Sarathi Nayak, Tutor,
Department of Dermatology,
4th Floor MCH Building, Medical College,
Kolkata, 88 College Street, Kolkata-700073,
West Bengal, India.
Email: psnbmoh@gmail.com

Case reports

Case 1 Basosquamous carcinoma of the scalp

A 62-year-old farmer, without any history of addiction and other ailments, presented with an erythematous, patchy hyperpigmented plaque on the right side of the back of the scalp **(Figure 1)**. It had initially started as a papule three years back which was mildly itchy and bled occasionally. It increased in size slowly over the years only to become a localized, circumscribed erythematous plaque of size of 5x6 cm, with few crusted papules located at the center and with areas of atrophy and hemorrhage. It also had yellowish, small scales on most of the region.

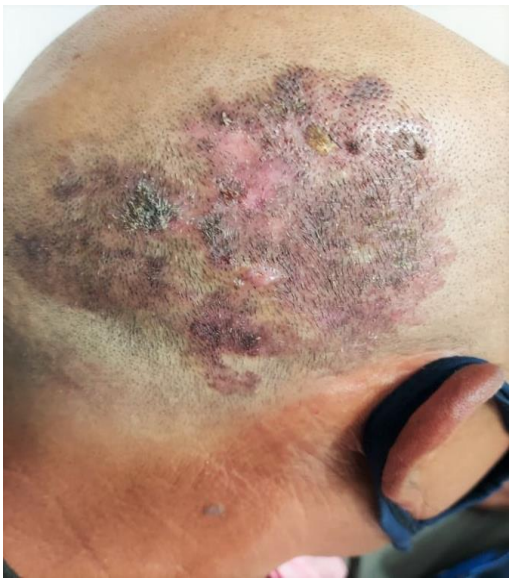


Figure 1 Erythematous, patchy hyperpigmented plaque of basosquamous carcinoma of scalp.

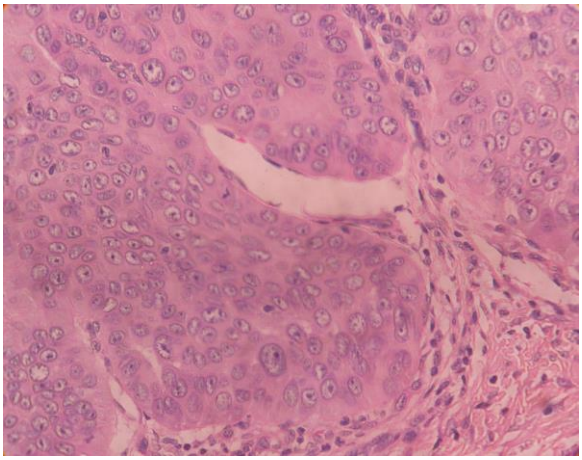


Figure 2 HPE of basosquamous carcinoma showing atypical squamous cells.

Numerous islands of atypical cells suggestive of malignancy is seen in the entire dermis showing basaloid as well as squamous differentiation in histopathology **(Figure 2)**. SCC dominant areas had multiple eosinophilic polygonal squamous cells with nuclei being round, large, polymorphic and hyperchromatic, with numerous mitotic figures. The area which exhibited features of BCC showed large and rounded basaloid cells. A transition zone between the two regions of SCC and BCC showed intermediate cells and appeared abruptly. As the size of the lesion was quite significant, the patient was referred to a plastic surgeon for complete excision of lesion along with skin grafting.

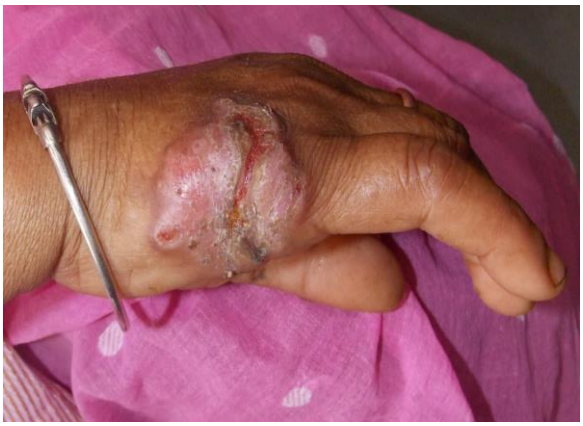


Figure 3 Enlarged tumor of porocarcinoma.

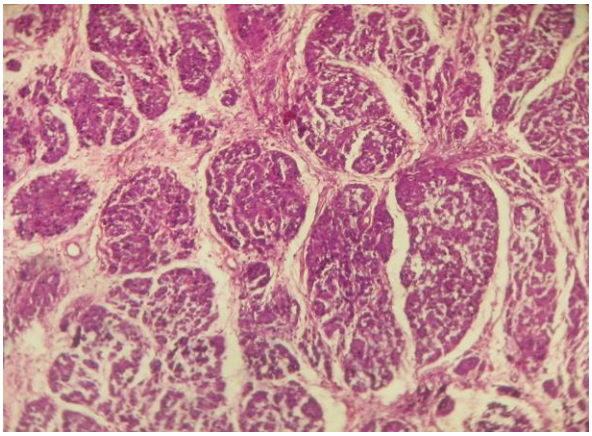


Figure 4 HPE of porocarcinoma showing broad anastomosing epithelial trabeculae with ductal differentiation and intracytoplasmic lumen formation, pleomorphism and mitotic activity.

Case 2 Porocarcinoma of the hand A 38-year-old housewife had come to our outpatient department with complaint of a tumor on the hand for 6 months. The mass had rapidly grown and was painful. On examination, a red, well defined tumor 3x2 cm in size with a central fissuring was observed on the radial aspect of her right hand **(Figure 3)**. She had no lymhadenopathy and other systemic examination was normal. Punch biopsy followed by histopathological examination showed nests of epithelial cells with intracytoplasmic lumen formation and multiple mitotic figures **(Figure 4)**. Immunohistochemistry was not performed.

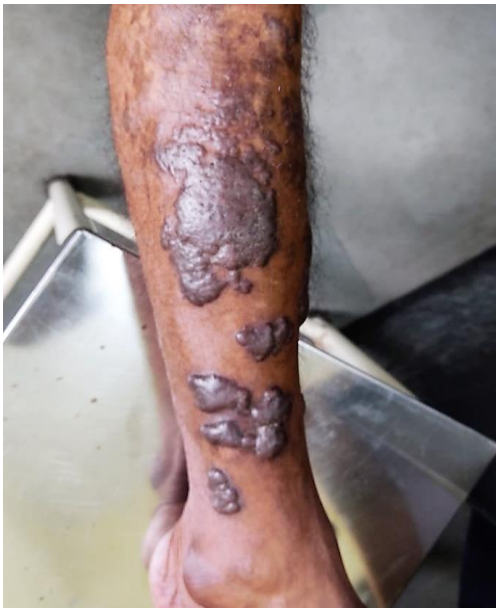


Figure 5 Dark red plaques in the leg in a patient with epithelioid hemangioendothelioma.

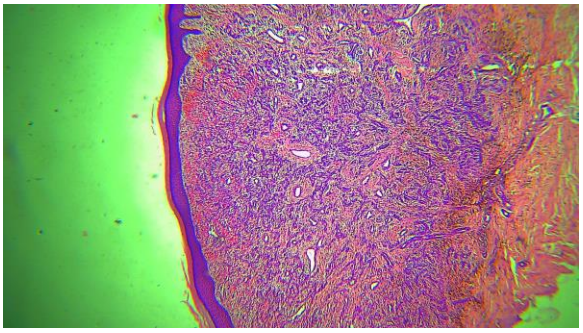


Figure 6 HPE of epithelioid hemangioendothelioma showing Eosinophilic spindle and ovoid-shaped tumor cells are found arranged in nests lying over hyalinized or mucoid stroma. Vascular channels are usually absent, intracytoplasmic vacuoles are seen with occasional erythrocytes. However, nuclear pleomorphism and mitotic figures are less or absent.

We referred the patient to the department of surgery for further management.

Case 3 Epithelioid hemangioendothelioma of the lower limb A 42-year-old man presented with a year long history of multiple growths unilaterally present over the right lateral aspect of the leg. It initially started as a flat lesion near the lower part of the leg and it gradually increased in size to form darkish red plaques which bled occasionally **(Figure 5)**. The lesions also increased in number and involved the upper part of the leg also. Routine examinations like complete blood count, liver and kidney function tests and urine examination were done which were within normal limits. There was no connection with the underlying bones in the computed tomography. Imaging studies of the thorax and abdomen did not reveal any feature suggestive of distant metastases. Histological examination with H&E stain showed infiltration of the tumor cells in a diffuse manner till lower dermis. Sinusoids were lined by large epithelioid tumor cells along with few mitotic figures **(Figure 6)**. However, well-formed vascular channels were not seen. Immunohistochemistry was not done. Eventually, the patient was referred to the general surgery department for further treatment and follow-up.

Case 4 Morpheaform BCC A 56 years, Female, normotensive, non-diabetic housewife, attended our OPD with history of multiple slightly itchy scaly lesions, some of which were ulcerated since last 7 years **(Figure 7)**. The lesions were present in chest, forehead, face, scalp and back. The lesions were hyperkeratotic, hyperpigmented, well demarcated ranging from size 0.5x 0.5 cm to 2.0x1.5 cm. The border of the lesion was round and hyperpigmented and

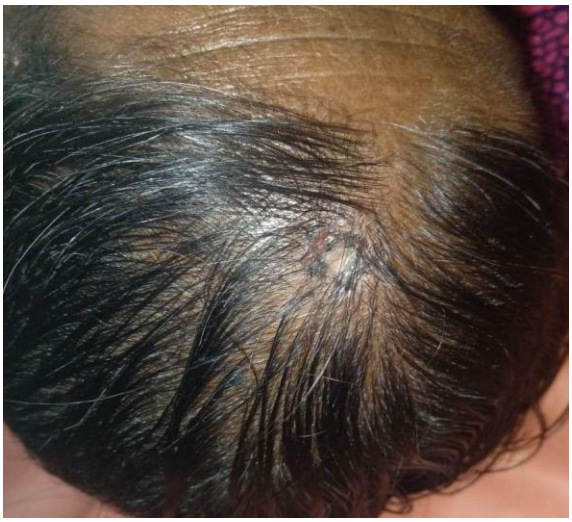
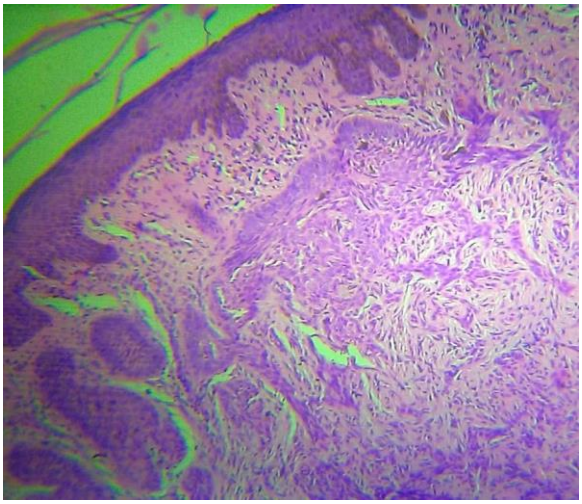


Figure 7 Morpheaform BCC of the scalp.



Figur 8 HPE showing thin strands of basaloid cells in a dense fibrous stoma suggestive of morpheaform BCC.

center erythematous, scaly, atrophic and some ulcerated. Systemic examination and routine blood examinations are within normal limit.

Histopathology from the margin of the lesion showed nest of basaloid cells arranged in a palisading manner **(Figure 8)**. The tumor cells are large, having oval nucleus and little cytoplasm. Narrow strands of basaloid cells in dense sclerotic stroma are also present. Hence the diagnosis of morpheaform BCC was made. The patient treated with imiquimod and the lesions improved a lot.

Discussion

Dermatologists mainly confront melanoma and nonmelanoma skin cancers but there is an array of other cutaneous malignancies that needs attention. Malignancies of the skin has mainly been divided into those that differentiate from the epidermis, dermis, adnexal structures of the skin and also those that arise systematically and undergo cutaneous metastases. Therefore, this article sheds light on the lesser discussed cutaneous malignancies.

Basosquamous Carcinoma The non-melanoma skin cancers (NMSC) are usually classified as BCC or SCC supported by histopathology. When the combined features of both SCC and BCC are exhibited by a single tumor, it is known as basosquamous carcinoma (BSC).[1,2] The main site of origin of the BSC is the head and neck region, especially on the sun exposed areas like the nose and in the auricular and periocular areas and it occurs mostly in Caucasian men.[3] The patient in our case is a man and having the lesion in the occipital region. He is a farmer by occupation and had a history of prolonged sunlight exposure. Though it resembles BCC, but unlike BCC, BSC is locally invasive, aggressive and has an increased chance to metastasize, these features in turn resembles SCC.[4,5] This is the reason why BSC cases needs long-term follow-up so that the local recurrences and distant metastasis can be detected earlier.

BSC is mainly a histopathological diagnosis.[6] Combined features of BCC and SCC are present along with a transition zone. Retraction artefacts as seen in BCC are absent, infact, some basaloid cells in BSC have eosinophilic cytoplasm. Peripheral palisades have uniform, small, hyperchromatic cells along with mitotic figures, surrounded by stroma with collagen deposition and fibroblasts which proliferates.[6] The area having features of SCC have large, eosinophilic, polygonal cells marking keratinization along with multiple dyskeratotic cells and mitotic figures. The nuclei are large with visible nucleoli and intercellular bridges.[6]

Treatment of choice for BSC is surgical excision with a wide margin. BSC frequently involves the lymph nodes and the chances of metastasis increases with perineural invasion.[7]

Epithelioid hemangioendothelioma In 1975, Dail and Liebow described an aggressive, rare, vascular tumor with malignant potential known

as Epithelioid hemangioendothelioma (EH).[8] Though it was described in lungs as bronchoalveolar carcinoma.[9] It metastasizes to bones and liver through lymphatics and blood.[10] It occurs equally in both sexes. Genetics and bartonella infection has been considered to play a role in the pathogenesis of EH. Reciprocal translocation t(1;3)(p36.3;q25) leads to fusion gene formation known as WWTR1-CAMTA1 which in turn may trigger the pathogenesis.[10] Cutaneous EH may present as a single or multiple dome-shaped erythematous nodules or as a tumor. It occurs mostly on the extremities. It may also present as nonhealing ulcer or scar.[11-13] In this case, the patient presented with multiple tumor masses in the leg.

Histopathology plays a cardinal role in diagnosis. Tumor cells are arranged in nests, some cells are ovoid in shape and some are spindle, with eosiniphilic cytoplasm. Stroma is mucoid or hyalinized. No organized vascular channels are seen; however, few erythrocytes are seen within intracytoplasmic vacuoles. Ironically, mitotic activities and nuclear pleomorphism are minimal or absent.[9]

EH tumors are aggressive with high infiltrative growth rates. Prognosis is uncertain. 10-15% of the tumors recur. 20-30% metastasize.[11]

The main mode of treatment is surgical excision. Radiation has minor role.[10]

Porocarcinoma Another atypical skin malignancy is the eccrine gland tumor known as porocarcinoma. Its benign counterparts, hidroacanthoma simplex and poroma are better known to a dermatologist. In 1963, it was first described by Mehregan and Pinkus.[14] In 1969, Moriko and Mishisma had coined the term 'eccrine porocarcinoma'.[14] It is a very rare tumor and only a handful of cases have been reported so far.[15] It may arise either from a pre-existing eccrine poroma or de novo.[14]

Porocarcinoma arises from the intraepithelial ductal portion of the eccrine glands.[16] It mostly occurs on the lower extremities in middle aged individuals with an equal sex predilection and in all races. It is usually less than 2cm in size and appears as an asymptomatic, erythematous, firm nodules.[17] It may also be dome-shaped, erosive or infiltrative plaque or an ulcerated polygonal growth. The tumor may have an accelerated growth phase but usually grows slowly.[18] Our patient had developed the lesion in the upper extremity, in the radial aspect of the right hand. The tumor was painful and it had a short course of only 6 months only to exceed more than 2cm in size. Porocarcinoma may be associated with immunocompromised states mainly like human immunodeficiency virus (HIV) infection and also other conditions like sarcoidosis, diabetes, organ transplantation.[18] However, there was no such association in our patient.

Histopathology establishes the diagnosis. The entire dermis is covered by multiple epithelial aggregates of poroid cells with atypias and ductal differentiation. The cells are stained with immunoperoxidase, pancytokeratins and Ki-67. Ductal structures stain with CEA (Carcinoembryonic antigen)[19] in immunohisto-chemistry.

20% of porocarcinoma recurs, and another 20% metastasize through lymphatics, this necessitates an early diagnosis. Treatment of porocarcinoma remains challenging. Most patients undergo wide local excision.[20]

Morpheaform BCC is the most common cutaneous malignancy. There are many histological variants, some histologic variants of BCC like the infiltrative, morpheaform, basosquamous and the micronodular ones have

clinically aggressive behavior and show higher rates of recurrence. These subtypes also metastasize frequently and have significant depth of invasion.[21,22]

Morpheaform BCC is a rare subtype of BCC with an incidence rate of 5 to 10 %.[23] As it resembles indurated-ivory coloured plaque of morphea, hence its name. Lesions appear either as shiny, ivory-white, indurated plaques or as ill-defined depressions, located mostly in the head-neck region. Frequently, scarring or atrophy may be seen. Small crusts, erosions or telengiectasias may also develop on the lesions. In our case, however, the clinical features strikingly resembled classical BCC with well-defined hyperpigmented borders, central atrophy and ulceration. However, the number of lesions were multiple.

Histopathology is again essential for establishing the diagnosis which shows strands of basaloid cells. It varies in thickness from one cell layered to five cell layered, dispersed in between dense collagen bundles. The tumor is poorly defined and shows diffuse infiltration in the reticular dermis and may even reach the subcutaneous fat. There is no retraction artifact and peripheral palisading of basal cells.

Morpheaform BCC is also known as infiltrating BCC and is more aggressive than nodular and superficial BCC as it tends to exhibit subclinical spread with the potential for extensive local destruction.

As the tumor extends microscopically far more than what is visible clinically, the treatment of morpheaform BCC is challenging. This causes high recurrances as compared to other BCC variants.[24]

Conclusion

Hereby we saw that cutaneous malignancies in clinical practice itself covers a vast range of cancers. The lesser discussed topics are important to revisit.

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