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Multiple Basal Cell Carcinoma of Head and Neck region

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Abstract:
This is a case of non-syndromic, non-familial type of multiple basal cell carcinoma (BCC). A 46-year-old Indian farmer presented with multiple, black-colored papules and plaques of various sizes over pinna, back, limbs, chest and abdomen since 6 years which slowly progressed to form ulcerative lesions since last 1 year. He had no history of irradiation, herb medication, arsenic intake, or exposure to chemical warfare gases. Family history for basal cell carcinoma was negative. In this patient we had ruled out the possibility of Gorlin's syndrome on history, clinical examination, and investigations. Histopathologically, the tumors revealed typical findings of basal cell carcinoma. The case in point is a very rare and unique case of non-syndromic and non-hereditary type of multiple BCC.

Key Word*: Multiple basal cell carcinoma, Gorlin's syndrome, hedgehog pathway, imiquimod*

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I. Introduction

Gorlin syndrome also known as nevoid basal cell carcinoma (nevoid BCC) syndrome or basal cell nevus syndrome, was first reported in 1960 by Goltz and Gorlin. [1]

It occurs in an autosomal dominant pattern. It is a neuro-cutaneous disease characterized by developmental defects which include bifid ribs and palmar plantar pits, and a predisposition to various tumors including Basal Cell Carcinoma, Medulloblastoma, Ovarioma, Cardiac fibroma, and keratocystic odontogenic tumor. [2]

BCC most commonly arises on sun-exposed areas and rarely on the mucous membranes or palms and soles. Basal cell carcinoma is a slow-growing tumor and rarely metastasizes. Usually, clinically, BCC appears as pink-colored, pearly papules with overlying ulceration or vessels with telangiectasia. BCC occurs on the head and neck in the majority of cases but can also involve the trunk and the extremities. [3-4]

II. Case report

A 46-year-old male, farmer by occupation and history of sun exposure, presented with complaints of multiple, black-colored papules and plaques of various sizes which gradually appeared behind left pinna, over the back and dorsum of left hand, face, chest and abdomen since 6 years. This slowly advanced to form ulcerative lesions within a year. On examination, the lesion over pinna was 6 × 2 × 1.3 cm in size with rolled on margins and hemorrhagic crust at the center [Figure 1]. Similar lesion of 1.8 × 1.2 cm size over the back and of size 2.3 × 1.2 cm was present over dorsum of left hand [Figure 2]. Apart from this, the patient also had multiple blackish hyper-pigmented plaques of varying sizes ranging from 0.5 cm × 0.5 cm to 1 × 1 cm over face, chest, abdomen and back [Figure 2]. He also had multiple palmo-plantar pits. [Figures 3] Skeletal examination revealed bony deformities such as short neck. There were no regional lymphadenopathies. There was no history of associated medical or surgical illness. Familial and past histories were non-contributory. CBC, urinalysis, coagulation profile, viral markers, biochemical markers, liver function tests, X-ray chest, electrocardiogram were within normal limits or negative.

Local application. All 3 lesions were completely removed by wide excision and primary closure of skin was done. [Figure 4] The histopathology report of the lesions revealed multifocal basal cell carcinoma with clear margins [Figure 5]. The patient was given 5% imiquimod cream for topical application. No new lesions were noticed after 6 months of healing. Patient is referred for radiation therapy and a definitive plan for pinna reconstruction is thought of after a tumor free interval of 1 year.

III. Figures

Figure 1: Basal cell carcinoma on pinna with rolled on margins and hemorrhagic crust at the center

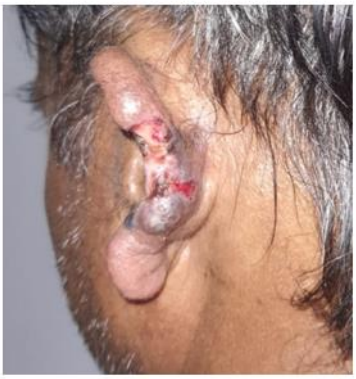


Figure 2: Pigmented basal cell carcinomas in dorsum of hand, face and over the back

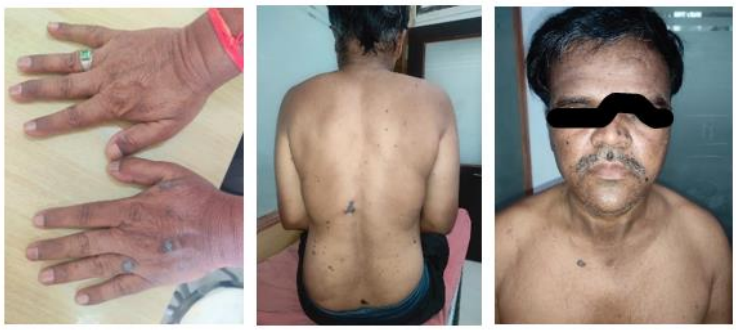


Figure 3: Palmoplantar pits

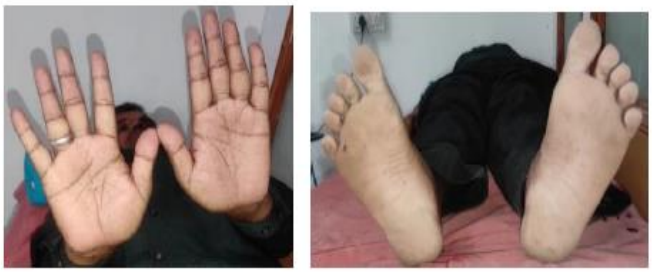


Figure 4 : Wide excision of BCC and primary closure

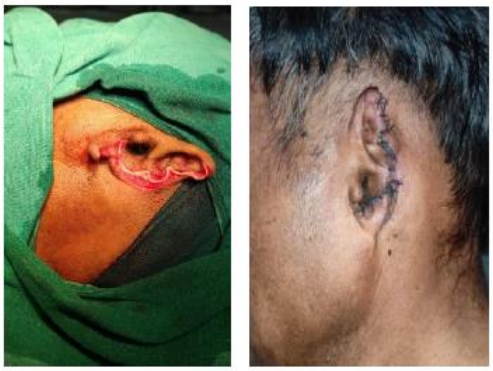
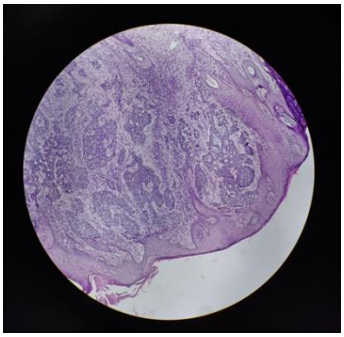


Figure 5 : Histopathological findings suggestive of Basal Cell Carcinoma



III. Discussion

Basal cell carcinoma (BCC) of the skin constitutes approximately 75% of all malignant skin tumors. [5] Presentation of multiple BCC's is also not uncommon as there is a 36%–50% increased risk of development of additional BCC within 5 years of the first lesion.[6]

The disease can be classified into non-syndromic and syndromic forms. In the first group, which comprises majority of the cases, the affected individuals did not have any geno-dermatosis susceptible to developing cutaneous malignancies. BCC lesions occur more commonly in middle-aged and old-adults. In the second group, the patients suffered from a genetic skin disorder (e.g., Gorlin-Goltz syndrome, Bazex syndrome, xeroderma pigmentosum), which predisposes them to BCC development at an early age. although most individuals demonstrate only a single lesion at each presentation, others may suffer many tumor clusters at various locations. [7-9]

Basal cell carcinoma lesions occur more commonly at sites frequently exposed to sunlight. BCC lesions arise most often in head and neck, and amount to more than 80% of cases.[10] However, they may occur anywhere, even in areas not exposed to direct sunlight.

The most commonly involved pathogenesis of basal cell carcinoma is exposure to ultraviolet rays, especially in the UV-B spectrum (290 to 320 nm), which activates mutations in tumour suppressor genes. Other factors involved in the pathogenesis include mutations in regulatory genes; radiation exposure , arsenicals, polyaromatic hydrocarbons, ultraviolet therapy and alterations in immune surveillance.

Basal cell carcinoma is associated with extremely low metastatic potential, but it does invade locally. It tends to grow along the path of least resistance. Metastases have been reported in the lungs, lymph nodes, skin, esophagus and oral cavity.

The treatment of basal cell carcinoma may be surgical or nonsurgical. The initial goal in the treatment of primary lesions is complete tumor removal, either by conventional surgical excision, cryosurgery, Moh's micrographic surgery, electro-desiccation and curettage, or topical methods.[10] Mohs' micrographic surgery is a special technique that offers high cure rates for basal cell carcinoma at high-risk sites (central face), and recurrent tumors, with maximum preservation of normal tissues which comprises of taking serial sections which are examined histologically until clear margins are obtained . It is the treatment of choice for all recurrent and infiltrative basal cell carcinomas.[11]

Nonsurgical techniques include, photodynamic therapy, topical fluorouracil, radiotherapy and topical imiquimod. Sonic hedgehog antagonist in the form of cream (cyclopamine) together with oral medication vismodegib (GDC-0449) is a recently tested modality of treatment. It has been approved by the FDA for use in adults with advanced and inoperable BCCs.[12] There is not enough evidence to make recommendations on topical solasodine glycol-alkaloids, topical ingenol mebutate, and intralesional 5-FU and IFN-α.

Vismodegib is a first-in-class hedgehog pathway inhibitor and recent addition to the armamentarium for the treatment of advanced basal cell carcinoma. [13-14]

The ideal treatment modality for older patients is Radiation therapy, particularly those with extensive lesions over lower limbs, eyes, or eyelids. Radiation therapy is not indicated for recurrent lesions and for young patients.

For superficial lesions topical photodynamic treatment has been found to be effective.

The case in discussion is different from those reported in literature. We had ruled out Gorlin's syndrome, based on history, clinical examination, and other investigations. Our case had no positive family history either. Exposure to irradiation, arsenicals, and sulphur mustard gas was also ruled out. There was no evidence of xeroderma pigmentosum or keratoacanthoma. Glutathione S transferase (GSTM1 and GSTT1) and cytochrome P450 (CYP2D6) genotypes have been found to be associated with multiple presentation phenotype of basal cell carcinoma. Hence, this patient was listed as a rare case of non-syndromic and non-hereditary type of multiple basal cell carcinoma.

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