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Case Report

Squamous cell carcinoma in an atrophic scar of Porphyria Cutanea Tarda

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ABSTRACT

Introduction: Porphyria's are group of inherited metabolic disorders of heme biosynthesis. Porphyria cutanea tarda (PCT) the most frequent type of porphyria worldwide and present with skin symptoms only. PCT develops due to acquired deficiency in fifth enzyme Uroporphyrinogen deaminase (UROD) in hepatocytes. Skin cancers are known to arise from site of trauma or chronic irritation like scars, ulcers, sinuses of diverse aetiology. Squamous cell carcinoma, Basal cell carcinoma, Adenocarcinoma are the common carcinoma identified in these situations. Hereby we present a case squamous cell carcinoma in an atrophic scar of PCT.

Case Report: A 65 years old male, diagnosed case of PCT since 20 years, presented with non-healing ulcer on left cheek since 6 months. It was overlying a preexisting scar of PCT and was progressively increasing in size. Dermatological examination shows single well-defined ulcer on left malar area with rolled out edge and pale granulation tissue on floor. Diagnosis of Squamous cell carcinoma was confirmed on histopathological examination. Patient underwent wide local excision with 1cm margin all around with propeller flap reconstruction which was conducted by plastic surgeons.

Conclusion: Patients of Porphyria Cutanea Tarda are at high risk of facial malignancy and need stringent monitoring for early detection and timely management.

1. Introduction

Porphyrias are metabolic diseases caused by abnormalities of an enzymes in the heme biosynthesis pathway. Porphyria Cutanea Tarda (PCT) develops due to acquired deficiency in fifth enzyme Uroporphyrinogen deaminase (UROD) in hepatocytes. There is inhibition of hepatic uroporphyrinogen decarboxylase (UROD) activity, which leads to accumulation of excess amounts of highly carboxylated porphyrins in liver, plasma, urine and faeces. In immunocompetent individual squamous cell carcinoma (SCC) is the second most common skin cancer after Basal cell carcinoma (BCC), and the most common skin cancer in immunosuppressed organ transplant recipient.[1] Risk factors include ultraviolet (UV)radiation, genetic predisposition, physical and chemical carcinogens, drugs, chronic injury to skin and chronic inflammation, etc.[2] We came across a case of SCC in old atrophic scar in a patient of PCT with photosensitivity. Here we present a case of SCC in an atrophic scar of PCT

2. Case Report

A 65 years old male farmer, presented with non-healing ulcer on left malar area since 6 moths. It started with small erosion which gradually progressed to form a large ulcer of size 5x3x2 cm with rolled out edge and pale granulation tissue on floor. It was associated with pain, without any pus or serous or hemorrhagic discharge. Patient had difficulty in closure of eyelid. Patient took oral and topical antibiotic for the same in initial stages but ulcer showed no response to it. On palpation ulcer was tender with indurated base and had no bleeding on manipulation. No localized lymphadenopathy. He had ectropion over his

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left lower eyelid with conjunctival chemosis. Surrounding area had atrophic scar with hypopigmentation with dry and wrinkled skin over rest of the face. Patient is a diagnosed case of Porphyria Cutanea Tarda since 20 years, Being a patient of PCT, atrophic scars were present all over his face, neck, upper limbs and dorsum of feet. Deformity of distal phalanx of fingers of hands along with onychomadesis and trachyonychia was seen.

Patient's HIV- ELISA and HbsAg were nonreactive. Urine woods lamp examination showed red colored florescence and urine uroporphyrinogen was reported positive. Skin biopsy was done from margin of ulcer. Histopathological examination from ulcer showed hyperplastic, mildly dysplastic, ulcerated stratified squamous epithelium, tumor cell arranged in groups. Individual tumor cells have hyperchromatic pleomorphic vesicular nuclei with prominent nucleoli. Few keratin pears noted. Above findings were diagnostic of well differentiated squamous cell carcinoma.

Patient has undergone wide local excision with 1cm margin all around with propeller flap reconstruction under plastic surgery department. And postoperative biopsy shows well differentiated squamous cell carcinoma with all margin of excision free of tumor.

Fig. 1: Single well defined ulcer with rolling margin, pale granulation tissue with crust on left malar area

3. Discussion

Porphyrias are metabolic diseases caused by abnormalities of the eight enzymes in the heme biosynthesis pathway. PCT is the most common porphyria. It develops due to acquired deficiency in fifth enzyme Uroporphyrinogen deaminase (UROD) in hepatocytes

Fig. 2: Post-operative picture with crust on malar area and watery discharge from left eye

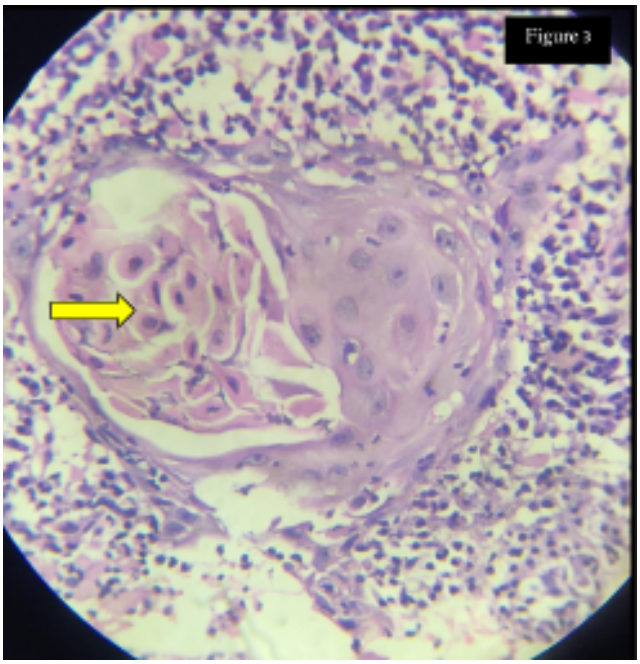

Fig. 3: Histopathological picture showing keratin pearl, hyperchromatic nucleus, pleomorphic cell , apoptotic cells with increased nuclear cytoplasmic ratio

which leads to accumulation of uroporphyrinogen and other highly carboxylated porphyrinogen which are oxidised to corresponding porphyrins leading to tissue damage. It occurs in presence of normal or increased amount of hepatic iron as PCT is an iron related disorder and develops only in the presence of normal or increased amounts of hepatic iron. Multiple susceptibility factors contribute to iron accumulation, oxidative stress, and generation of a UROD inhibitor in hepatocytes, and are important to identify in individual patients. Most common in males usually mid or late life period.[3] Heterogenous UROD mutations are found in around 20% of patients.[4] Presenting features include fluid filled blisters and bulla on sun exposed parts like face and dorsum of hands. Long standing disease increases skin fragility, blisters ruptured leading to erosion with residual scaring associated with hyper or hypopigmentation.[5] PCT is the most readily treated porphyria responding well to phlebotomy or low dose hydroxychloroquine. Increased photosensitivity and atrophic scar increase the risk of skin cancers in PCT patients.

Squamous cell carcinoma (SCC) is the second most common skin cancer after Basal cell carcinoma (BCC). Risk factors include ultraviolet (UV) radiation, genetic predisposition, physical and chemical carcinogens, drugs, chronic injury to skin and chronic inflammation, etc. Incidence is more in men than in women, with age more than 60 years it mostly develops on sun exposed areas like face, head, neck forearm dorsum of hands.[6] Diagnosis of SCC is established histologically. histologic subtypes include spindle cell, acantholytic, verrucous, and desmoplastic SCCs, and keratoacanthoma. The primary mode of therapy for localised SCC is complete surgical excision, preferentially microscopically controlled surgery. Non-surgical interventions include topical therapy such as imiquimod, 5-fluoruracil, cryotherapy[7,8] and for locally advanced unresectable or metastatic SCC, radiation therapy and systemic treatment with chemotherapy or targeted therapy. primary prevention for the development of SCC is based on decreasing UV radiation exposure and concomitant risk factors.

Our case is a classical prototype of Porphyia cutanea tarda with its chronic complications like scarring and skin carcinomas (squamous cell carcinoma). Consistent follow up with cancer surveillance has led to diagnosis of our patient's cancer in stage 1 (TNM) and has helped improve patient care and outcome.

He had undergone wide local excision of his tumour with propeller flap reconstruction by our hospital's plastic surgeons. Post operatively, he had mild ectropion in his left lower eyelid margin which is being followed up in ophthalmology.

Being a patient of PCT, he is advised sun-protection with sunscreens and regular visits in skin OPD to look for any signs of recurrence or any new ulcers.

4. Conclusion

Patients with long standing Porphyria cutanea tarda are at high risk of facial malignancy and need stringent monitoring for early detection and timely management.

4.1. Abbreviation

PCT – porphyria cutanea tarda, SCC - squamous cell carcinoma, BCC- basal cell carcinoma, UV- ultraviolet

4.2. Declaration of Patient Consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understand that his name and initials will not be published and due efforts will be made to conceal the identity, but anonymity cannot be guaranteed.

4.3. Acknowledge

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5. Source of Funding

None.

6. Conflict of Interest

None.

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