THE SINGULAR PRESENTATION OF CUTANEOUS SPINDLE CELL SQUAMOUS CELL CARCINOMA OF THE INGUINAL REGION

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PRESENTATION OF CASE

A 33-year young male presented with chief complaints of an ulcerated lesion in right inguinal region for 4-months. Patient was apparently asymptomatic 4-months back when he had swelling the size of a lemon in the right inguinal region, associated with continuous dull aching pain, non-radiating and not associated with bowel or bladder complaints. The swelling gradually progressed in size and subsequently developed an ulcerated irregular lesion involving inner aspect of the entire thigh with necrotic slough and pus discharge. There was no history of weight loss, loss of appetite or breathlessness. There were no other significant systemic illnesses. Patient was a chronic smoker for 15-years, non-alcoholic and directly exposed to sun for prolonged periods due to occupational reasons. Systemic examination was normal. All haematological and biochemical parameters were within normal limits. On examination, there was a confluent asymmetric ulceroproliferative lesion of size 20×15 cm, involving inner aspect of right thigh and groin with serosanguinous discharge. The neoplastic cells illustrated wide spread positivity for cytokeratin (CK), epithelial membrane antigen (EMA) and vimentin immunocytochemical stain, while stain for leucocyte common antigen (LCA) was negative. Magnetic resonance imaging revealed a lesion in right inguinofemoral region of 14×3 cm in transverse and antero-posterior diameters. The lesion is seen in cutis and infiltrates the subcutaneous tissue. Multiple prominent and mildly enlarged bilateral inguinal nodes are seen with lesion seen in right half of sacrum. The histopathological appearance, IHC profile of the biopsy tissue and MRI scan confirmed the diagnosis as spindle cell squamous cell carcinoma, stage IV. Subsequently patient received palliative radiotherapy of 800 cGy single session to the involved site with adequate margins by Co-60 external beam radiotherapy and received pain palliation to some extent. However, given the patient's poor functional status and very rapid progression of the disease, the patient elected to get discharged and was lost to follow-up.

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CLINICAL DIAGNOSIS

Aggressive cutaneous malignancy

DIFFERENTIAL DIAGNOSIS

The clinical presentation of spindle cell squamous cell carcinoma (SCSCC) is misleading as it presents as a rapidly growing tumor. The diagnosis of this rare entity mainly depends on the pathological and immunohistochemical (IHC) profile with the latter playing major role in excluding various differentials. The pathological and clinical differentials for SCSCC are Squamous Cell Carcinoma (SCC), sclerosing basal cell carcinomas, microcystic adnexal carcinomas, desmoplastic trichoepithelioma, basosquamous carcinoma, atypical fibroxanthoma, spindle cell malignant melanoma, cutaneous T cell lymphoma and leiomyosarcoma.

PATHOLOGICAL DISCUSSION

Spindle cell squamous cell carcinoma (SCSCC) is a rare variant of squamous cell skin cancer and was first reported in the era of King George V i.e. in year 1935.[1] Clinically, it overlap with conventional SCC in many aspects such as site and pathogenesis but these lesions are pathologically distinct as these lesions comprise spindle cells that infiltrate the dermis as single cells or cohesive nests; that lack features of keratinization.[2] It mimics sarcoma microscopically and has high frequency of aggressive clinical course, extensive local disease, recurrence and metastasis.
It is a biphasic tumor with epithelioid and spindle shaped neoplastic cells. Due to biphasic morphology**,** it is also known as carcinosarcoma. It commonly occurs in elderly population with very high male preponderance i.e. male to female ratio of 11**:**1.[3] Contrary to this our case is a young immunocompetent male. Spindle cell carcinoma accounts for nearly 7.4% of squamous cell carcinoma.[4] As with other skin

malignancies tumor thickness of 0.5 mm or infiltration beyond the dermis are indicators of poor prognosis.[5] Overall rates of metastasis are 2-5%, with about 85% of metastases occurring in regional lymph nodes and the remainder occurring at distant sites. As observed in most of the reported cases**,** if regional lymph nodes are involved then 10-year survival rates are less than 20% and less than 10% for patients with distant metastases.[6,7]

Previous skin injury, including trauma and prior radiotherapy are postulated to be causative for spindle cell SCC. Like other cutaneous malignancies they tend to occur on sun damaged skin with the sites of predilection being head and neck region,[8] especially ears but in our case the site was very unusual being inguinal region.

The spindle cell squamous cell carcinoma generally presents as an exophytic or ulcerated polypoidal mass which is firm and non-tender without other specific clinical features.[9] The duration of symptoms is short due to rapid progression of this malignancy which is generally observed to be less than 1-year in approximately 95% of patients. Histologically, dermis infiltration is in the form of single atypical spindle cells with elongated nuclei often arranged in short fascicles but with no signs of the keratinization that are associated with conventional SCC therefore routinely employed cytokeratin stains such as AE1/AE3 are often negative.[10] Hence, in order to establish the diagnosis, a large panel of immunohistochemical (IHC) markers for epithelial, mesenchymal and neurotropic origin differentiation are employed to assign a tumor as spindle cell squamous cell carcinoma. The commonly used IHC markers are cytokeratin (CK), epithelial membrane antigen (EMA), high molecular weight CK (HMWCK), vimentin, calponin, P63, CD31, CD34, S-100, leucocyte common antigen (LCA) and human melanoma black (HMB)-45.[11] There is strong expression of P63 found in most cases.[12] Among these, the markers which are used to rule out other differentials are CD31 & CD34 for angiosarcoma, calponin for leiomyosarcoma, LCA for lymphomas, HMB45 for malignant melanomas as well as for malignant fibrous histiocytomas.

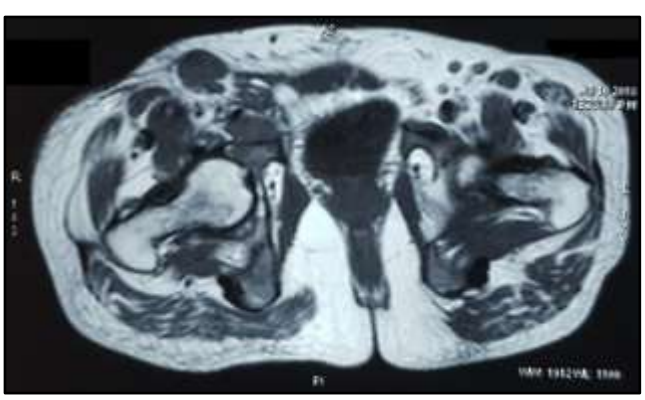

Figure A

Figure A: Sagittal T1-weighted MR image of pelvis showing a lesion in right inguino femoral region cutis infiltrating the subcutaneous tissue.

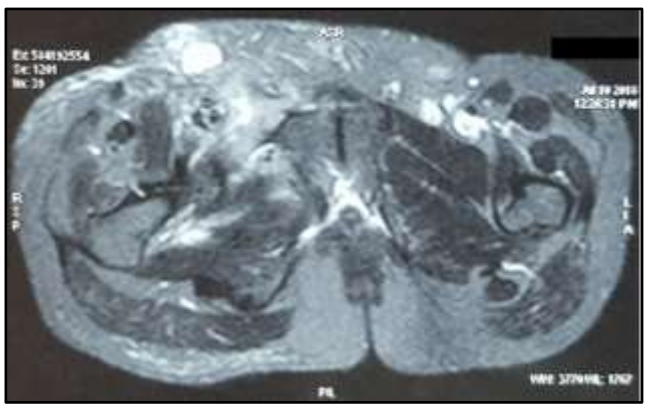

Figure B

Figure B: Sagittal T2-weighted MR image of pelvis showing hyper intense lesion in right half of sacrum along with lesion in inguinofemoral cutis infiltrating the subcutaneous tissue.

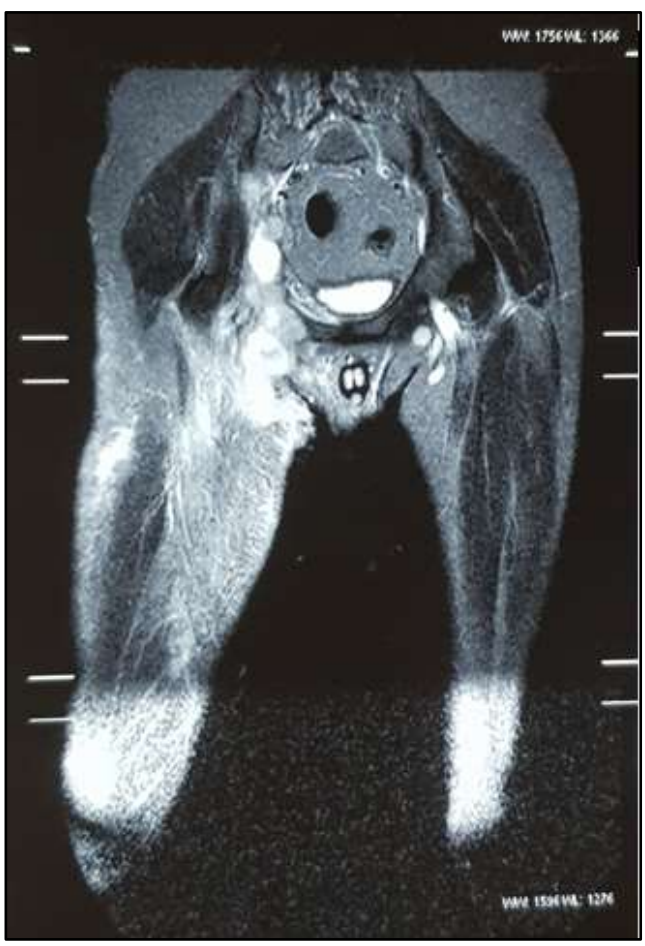

Figure C

Figure C: Coronal T2-weighted MR image showing hyperintense lesion in right half of sacrum along with lesion in right inguinofemoral region extending from inguinal region to lower part of right thigh.

DISCUSSION OF MANAGEMENT

Due to such rarity of this subtype of SCC, there is no standard of care regarding the paramount treatment approach for advanced stage spindle cell SCC. In general, it

is treated as other SCC types. Treatment of nodal disease involves local external beam radiation therapy (mostly done for palliation) or less commonly lymph node dissection with curative intent if feasible, or a combination of both the modalities if residual disease is detected after surgery. Systemic chemotherapy, palliative radiotherapy or biologic response modifiers are various approaches for metastatic disease. On account of the minimal number of cases reported so far, the efficacy of any of these methods has not been established.[13]

FINAL DIAGNOSIS

Spindle cell squamous cell carcinoma, stage IV.

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