



Metastatic Eccrine Carcinoma of The Axilla

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Abstract

Sweat gland carcinoma is very rare with a reported incidence of less than 0.005% of all tumour specimens resected surgically. These are considered as a rare group of tumours with potential for local destruction as well as distant metastasis. Wide surgical excision along with regional lymph node dissection in the presence of clinically positive nodes is the recommended treatment. However, the variability of the histological features even in the same tumour, and its rarity, have contributed to some confusion regarding the classification of sweat gland carcinoma as well as the treatment of specific histological subtypes. Apart from that, documentation is also inadequate regarding management of metastatic cases. Here we present a rare case of metastatic eccrine sweat gland carcinoma in a 50 year old male patient which initially created a diagnostic dilemma and was non responsive to different chemotherapies with gradual disease progression.

Key words: Metastatic eccrine carcinoma, Skin adnexal tumours, Sweat gland carcinoma.

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Introduction

There are two types of sweat glands. Eccrine sweat glands open directly onto the surface of the skin and are widely distributed almost everywhere. Apocrine sweat glands are found in the armpit, areola, perineum, ear and in the eyelid. Rather than opening directly onto the surface of the skin, they secrete sweat into the pilary canal of the hair follicle.¹ Sweat gland carcinomas represent a rare group of tumours with potential for destructive local tissue infiltration and regional as well as distant metastasis. The management of these neoplasms is both complex and cumbersome, mainly due to limited availability of literature. Histological resemblance to the mature gland in biopsy specimen contributes, but diagnosis is primarily based on immunohistochemistry or ultrastructural features. These tumours, therefore, can be considered as clinico-pathological dilemmas with an unpredictable biological behaviour. Rarely diagnosed clinically, they are often encountered as operative and histological surprises.

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The case

Present case was a 50 year old non diabetic, normotensive, smoker patient who initially came in April, 2015 with multiple subcutaneous nodules with ulcerations over his right axilla which he first noticed two years before that. He received homeopathic treatment which resulted in disease progression turning small subcutaneous nodules in multiple discharging chronic non healing ulcers. He gave no previous history of tuberculosis, chronic arsenicosis, psoriasis, chronic skin infection and family history of malignancy. He also did not complain of having hemoptysis, hematemesis, melena, significant weight loss, fever, night sweat, swelling anywhere else in the body, and had normal bowel and bladder habit. On presentation local examination revealed multiple non tender mobile subcutaneous swellings larger one measuring about

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2. Consultant, Department of Oncology, Ahsania Mission Cancer and General Hospital, Uttara, Dhaka, Bangladesh.

3. Associate Professor of Radiology and Imaging, Khwaja Yunus Ali Medical College and Hospital, Enayetpur, Sirajganj, Bangladesh.

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Correspondence: Dr. Deepak Shankar Ray, Senior Consultant, Department of Oncology, Khwaja Yunus Ali Medical College and Hospital, Enayetpur, Sirajganj, Bangladesh. Phone: +8801741982895, Email: deepaksray@hotmail.com 1.5×1.5 cm with erythematous overlying skin, single discharging sinus and unhealthy surrounding tissue. Systemic examination revealed no abnormalities. Routine hematological and biochemical investigations were sent which were normal. But echocardiography revealed aortic sclerosis with trivial aortic regurgitation with normal LVEF. Ultrasonography of whole abdomen revealed gall bladder cholesterolosis, renal cortical cyst and benign prostatic hyperplasia and CXR PA view revealed pneumonitis. Decision was taken for local excision with axillary lymph node biopsy which was done on 29th April, 2015. Histopathology report was consistent with primary adenocarcinoma of adnexal origin or metastatic adenocarcinoma. Report was reviewed and findings were conclusive for sclerosing ductal eccrine carcinoma (microcystic adnexal carcinoma) with involved resection margin (Illustration 1). IHC revealed positive CK-7 and negative TTF 1, CK 20 which was unfavorable for lung and colon primary (Illustration 2). PET/CT scan report showed avid FDG uptake over right axilla, pretracheal, paratracheal, mediastinal, subcarinal and bilateral hilar regions, non FDG avid uptake over sub mental, right parapharyngeal, left axilla, pre aortic, mesenteric, para aortic, ilio-inguinal regions with focal uptake over rectum, ascending colon and D12 vertebra (Illustration 3 and 4). Serum tumour marker revealed raised CA 19-9 (129U/ml) with normal level of Serum LDH, PSA, CEA and CA 15-3. Patient was started on chemotherapy with gemcitabine based regimen (GEMOX - Gemcitabine 1400 mg iv d1 and Oxaliplatin 150 mg iv d2) with significant improvement of subjective response and extensive lymphadenopathy but persistent rise in CA19-9 level. After completion of 12 cycles of chemotherapy, recorded serum CA 19-9 level was 253 U/ml (on 28th Nov, 2015). Patient was kept on regular follow-up and on 1st follow up on 27th Feb, 2016 recorded CA 19-9 was 93.20 U/ml with normal CEA, CA 15-3. Follow up PET/CT scan revealed interval improvement with increased FDG uptake over lower rectum and sacral vertebra. Accordingly colonoscopy and double contrast barium enema was done which was normal. Subsequently he was started on chemotherapy with doxorubicin, cyclophosphamide and capecitabine, however no significant response to therapy was observed.

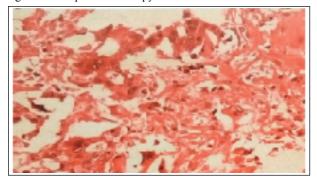


Illustration 1: Photomicrograph showing sclerosing ductal eccrine carcinoma.

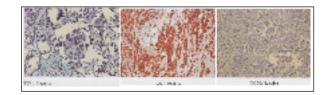


Illustration 2: IHC showing positive CK-7 and negative TTF 1, CK 20.



Illustration 3: MIP FGD PET showing metabolic active areas in right parapharyngeal, pre aortic, mesenteric, para aortic regions with focal uptake over rectum, ascending colon.

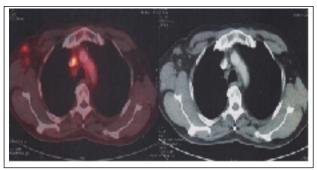


Illustration 4: FDG-PET image showing FDG avid area in right axilla and right paratracheal lymph node.

Discussion

Sweat gland carcinomas occur primarily in adult patients, with a peak incidence in fifth and sixth decades of life.^{3,7,8} Majority occur in the genital skin and perineum (34.5%), followed by trunk (26.4%), head and neck (18.3%) and lower extremities (13.9%).^{7,9} In this case, the patient was 50 years old and his initial presentation was in the right axilla. Apocrine carcinomas manifest as non-tender single or multiple, firm, rubbery or cystic masses with red to purple overlying skin.^{6,9} Tumour cells are PAS (periodic acid-schiff stain) positive due to glycogen granules and diastase resistant.³ Eccrine gland carcinomas possess no distinctive clinical features making diagnosis by gross appearance virtually impossible. They usually manifest as non-tender, subcutaneous nodules,

primarily in elderly individuals. Individual malignant cells are rich in glycogen and stain with PAS and are diastase sensitive with prevalent nuclear changes and propensity for lymphatic invasion.^{8,9} Sites of sweat gland carcinoma metastasis include nodes, lungs, liver and bone. Metastatic deposits from undiagnosed visceral and breast adenocarcinoma are virtually indistinguishable microscopically from sweat gland carcinoma and must be considered before a diagnosis of metastatic sweat gland carcinoma is made.^{3,5,9}

The recommended treatment of all subtypes of sweat gland carcinomas is wide surgical excision along with regional lymph node dissection in the presence of clinically positive nodes. Some authors advocate prophylactic regional lymph node dissection, especially in patients with recurrent lesions after wide excision or with highly undifferentiated tumours. Sweat gland carcinomas are radio resistant, and chemotherapy has been infrequently employed.¹⁰⁻¹⁶ Prognostic factors for sweat gland carcinomas are difficult to identify, again owing to the small number of reported cases. The likely prognostic factors include size, histological type, lymph node involvement and distant metastasis. A 10-year disease free survival rate of 56% in the absence of lymph node metastasis is observed which falls to 9% if nodes are involved.¹⁵⁻¹⁷ To the best knowledge of the authors, this is the first case of ductal eccrine carcinoma of the axilla to be reported from Bangladesh.

Conclusion

Sweat gland carcinomas are a rare and aggressive group of tumours with potential for local destruction as well as distant metastasis. Wide surgical excision along with regional lymph node dissection in the presence of clinically positive nodes is the recommended treatment. Standard treatment guidelines for metastatic eccrine carcinoma have not been established, however the role of conventional chemotherapy is unclear, with these tumours appearing to be chemo resistant in previously reported cases. Longer follow up, clinical trials and reporting of clinical experiences are the way forward in identifying new treatment options.

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