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Fatal Spontaneous Tumor Lysis Syndrome: A Rare Presentation in Sarcomatoid Carcinoma

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Abstract

Background: Tumor lysis syndrome (TLS) is a potentially lethal oncological emergency which usually occurs in a hematological malignancy after initiation of chemotherapy. Here we present a rare case of fatal spontaneous TLS with reactive leukocytosis in a patient with localized sarcomatoid carcinoma. *Case Report*: A 73 years old lady a treated case of carcinoma right breast 22 years back presented with a large left infraclavicular mass. Core needle biopsy showed sarcomatoid carcinoma. Laboratory values showed elevated blood glucose, ketones, C reactive protein and procalcitonin, with features of TLS like elevated creatinine, hyperuricemia, hyperphosphatemia. She also had hypercalcemia and leucocytosis. Bone marrow studies revealed hypercellular marrow with myeloid hyperplasia. She was managed for tumor lysis syndrome and probable sepsis. Leukocytosis persisted inspite of hydroxyurea and steroids. Patient had a rapid deterioration and expired inspite of improvement in biochemical parameters. *Conclusion*: We present a rare case of fatal spontaneous tumor lysis syndrome with associated hypercalcemia and reactive hyperleukocytosis in a case of localized sarcomatoid carcinoma. Spontaneous tumor lysis is an extremely rare emergency in a localized solid tumour. A high suspicion in such histology and early recognition may improve patient outcome.

Keywords: Breast Neoplasms, Mastectomy, Biopsy, Hydroxyurea, Steroids, Bone Marrow Aspiration.

Introduction

Tumor lysis syndrome (TLS) is an oncological emergency characterized by metabolic and electrolyte imbalances occurring due to lysis of tumor which is usually seen after cytotoxic treatment. Tumor lysis leads to immediate release of large amounts of various intracellular components. It is associated with hyperuricemia, hyperkalemia, hyperphosphatemia and hypocalcemia. These electrolyte imbalances causes organ dysfunction which are life threatening with acute renal failure and even fatal cardiac arrhythmia and death [1]. Tumor lysis syndrome develops usually after initiation of chemotherapy but rarely may arise spontaneously before antitumor therapy. TLS is common in patients with hematologic malignancies with high tumor burden but rarely seen in patients with solid tumors [2]. Here we present a case of spontaneous tumor lysis syndrome in a localized solid tumor associated with hypercalcemia and hyperleukocytosis.

Case Report

A 73 years old lady with a history of type 2 diabetes mellitus and hypertension was diagnosed with carcinoma right breast and underwent right modified radical mastectomy 22 years back. She presented with history of lump in the left breast and left infraclavicular region for 20 days. Physical examination revealed an ill-defined thickening measuring 2×2 cm in the retro-areolar region of left breast with nipple retraction and another tender, hard mass measuring 10×10 cm in the left infraclavicular region.

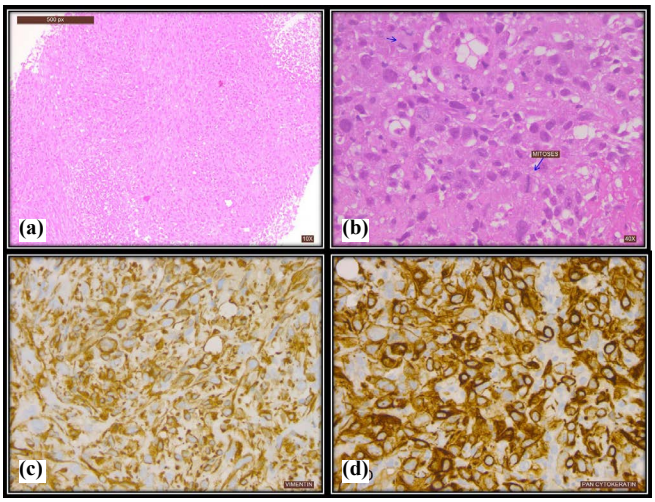

Fig.1: *Hematoxylin and Eosin staining:* **(a):** *10X magnification* **(b):** *40X magnification showing poorly differentiated malignant neoplasm. Immunohistochemistry showing positivity* **(c):** *vimentin* **(d):** *cytokeratin AE1/AE3.*

Mammogram and ultrasound of breasts revealed a 13 mm breast imaging-reporting and data system (BI-RADS) IV lesion in the left breast and a large hypoechoic lesion of size 95×73 mm in the left infraclavicular region and axilla. An ultrasound guided core needle biopsy was performed from the left breast lesion and the infraclavicular swelling. Left breast lesion showed proliferative breast disease with intraductal papillomatosis, infraclavicular mass revealed poorly differentiated malignant neoplasm suggestive of carcinosarcoma. Immunohistochemistry (IHC) showed c-expression of pancytokeratin (CK-AE1/AE3) and vimentin in favour of sarcomatoid carcinoma **[Fig.1]**.

18-fluoro deoxy glucose positron emission tomography-computed tomography (PET-CT) showed intense uptake (Standardized Uptake Value max-10.2) with central necrosis in left infraclavicular region and axilla of size 113×105 mm. No infiltration into surrounding muscles and elsewhere in the body was noticed **[Fig.2]**. Two weeks from biopsy, she presented to emergency department with history of decreased food intake and altered sensorium. She was drowsy, not obeying commands. There was no history of fever

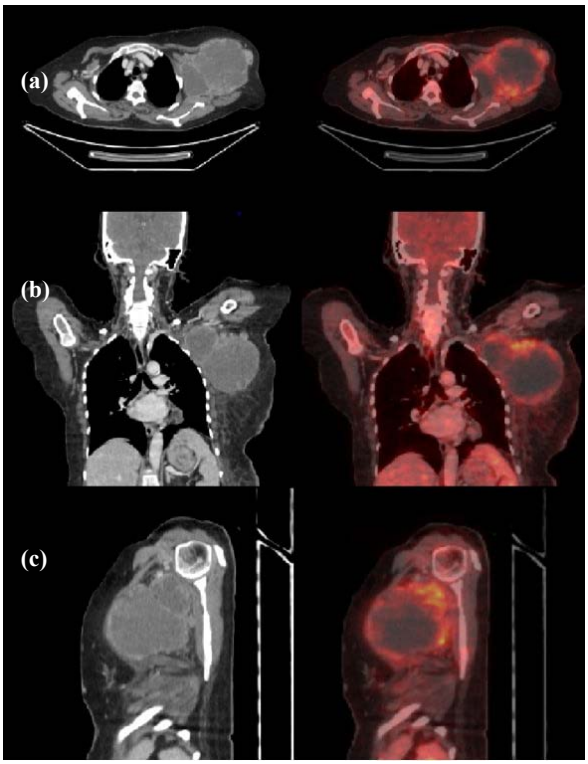

Fig.2: *Computed tomographic (CT) images (left panel) and 18-fluoro deoxy glucose - positron emission tomography [FDG-PET] - CT fused images (right panel) showing intense uptake (SUV max-10.2) with central necrosis in left infraclavicular region and axilla of size 11.3×10.5 cm in axial* **(a):** *coronal* **(b):** *and sagittal* **(c):** *planes.*

or history suggestive of any other focus of infection. There was no history of tumor directed treatment. Her initial investigation revealed elevated blood glucose, creatinine, ketones, uric acid, phosphate, calcium, total leukocyte count, C reactive protein, procalcitonin and lactate dehydrogenase with low bicarbonate. A provisional diagnosis of diabetic ketoacidosis, tumor lysis syndrome and sepsis was made.

She was admitted and managed for tumor lysis syndrome and probable sepsis with intravenous antibiotics cefaperazone and sulbactam, fluids and rasburicase. Her blood and urine culture did not show any growth. She did not have any episodes of fever. Gradually her electrolytes normalised. Inspite of biochemical improvement, she deteriorated clinically. Bone marrow aspiration

and biopsy revealed moderately hypercellular marrow with myeloid hyperplasia and shift to left in maturation **[Fig.3]**. She was started on steroids and hydroxyurea but leukocytosis persisted. Clinically infraclavicular mass became ill defined and softer. Her general condition continued to deteriorate and the patient succumbed 7 days after admission before any tumor directed treatment could be started.

Discussion

This case report describes a 73-year-old woman with a history of type 2 diabetes mellitus and hypertension who presented with a lump in her left breast and left infraclavicular region. Physical examination revealed an ill-defined thickening in the retro-areolar region of the left breast with nipple retraction and a tender, hard mass in the left infraclavicular region. She developed spontaneous lysis syndrome with hypercalcemia and hyperleukocytosis. Biopsy revealed proliferative breast disease with intraductal papillomatosis and poorly differentiated malignant neoplasm suggestive of carcinosarcoma, with immunohistochemistry showing co-expression of pan-cytokeratin and vimentin in favour of sarcomatoid carcinoma. PET-CT showed intense uptake with central necrosis in the left infraclavicular region and axilla. To our knowledge there is no report of a TLS due to localised sarcomatoid carcinoma. According to Cairo Bishop definition, our case meets the criteria for laboratory TLS as well as clinical TLS grade III [3]. TLS is usually seen in tumors with a high proliferation rate, high tumor burden and pronounced sensitivity to chemotherapy. It is rare to occur in a solid tumor spontaneously. However, there are few case reports of spontaneous TLS in lung, seminoma, gastrointestinal and uterine cancers with high volume disease [4].

This case highlights the challenges in managing a patient with multiple co-morbidities and advanced-stage cancer. The presence of diabetes mellitus and hypertension can complicate the management of cancer and increase the risk of complications such as diabetic ketoacidosis. Additionally, the development of tumor lysis syndrome, a potentially life-threatening complication of cancer treatment, underscores

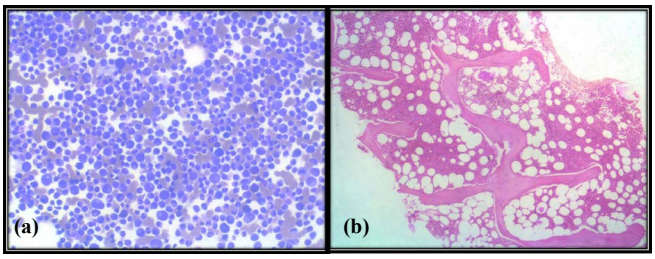

Fig.3: *Bone marrow aspiration cytology* **(a):** *showing solidly cellular marrow with myeloid hyperplasia and bone marrow biopsy* **(b):** *showing moderately hypercelluar marrow with myeloid hyperplasia with no atypical cells.*

Table 1: *Laboratory parameters at baseline and their change during course of treatment.*

Lab parameter	At Baseline (Day 1)	Remarks
Total leukocyte count	123000 cells/mm^3	Persistently high >1 lakh/mm^3 till death
Absolute neutrophil count	86100 cells/mm^3	88330 cells/mm^3 on Day 7
Absolute lymphocyte count	6150 cells/mm^3	7260 cells/mm^3 on Day 7
Absolute monocyte count	1230 cells/mm^3	0 cells/mm^3 on Day 7
Myelocyte count	17220 cells/mm^3	13310 cells/mm^3 on Day 7
Metamyelocyte count	3690 cells/mm^3	3630 cells/mm^3 on Day 7
Band forms	6150 cells/mm^3	7260 cells/mm^3 on Day 7
C reactive protein	222 mg/L	
Procalcitonin	2.31 ng/mL	1.41 ng/mL on day 4
Calcium	12 mg/mL	Normalized on day 3
Ketones	2.7 mmol/L	Normalized on day 2
Phosphorus	6.3 mg/dL	Normalized on day 2
Potassium	4.7 mmol/L	Normal
Bicarbonate	11.8 mEq/L	Normalized on day 6
Urea	195 mg/dL	137 mg/dL on day 7
Creatinine	6.5 mg/dL	1.9 mg/dL on day 7
Lactate dehydrogenase	1189 U/L	
Uric acid	22.3 mg/dL	Normalized on day 3

the need for close monitoring and timely intervention. The diagnosis of carcinosarcoma is also significant as this rare malignancy has a poor prognosis and can be difficult to treat. The co-expression of pancytokeratin and vimentin, as shown by immunohistochemistry, is suggestive of sarcomatoid carcinoma, which is known to be particularly aggressive and resistant to chemotherapy. The patient's rapid clinical deterioration and lack of response to steroid and hydroxyurea therapy highlight the need for more effective treatment options for this rare and aggressive malignancy.

Conclusion

This case report describes a rare occurrence of spontaneous tumor lysis syndrome, accompanied by hypercalcemia and reactive hyperleukocytosis, ultimately resulting in a fatal outcome, in a patient with localized sarcomatoid carcinoma.

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