Extramedullary haematopoiesis mimicking metastatic lymphadenopathy in a patient with squamous cell carcinoma of the vulva

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Abstract

Introduction

We report a case of extramedullary haematopoiesis manifested as multifocal lymphadenopathy in patient with vulvar cancer. The appearance of haematopoietic elements outside the bone marrow has been associated with solid tumours.

Case report

A 60-year-old woman presented to the Emergency Department at SUNY-Downstate Hospital complaining of generalised abdominal pain. Her medical history was significant for seizure disorder. Her medications were Keppra and Gabapentin. Family history was non-contributory. Social history was non-contributory. Relevant laboratory findings revealed haemoglobin 12 g/dL, haematocrit 40, MCV (mean corpuscular volume) 82 fl, RBC count 5 million/mL, platelets 141,000/mL, WBC count 8490/mL, BUN (Blood Urean Nitrogen) 8 mg/dL, Cr 0.7 mg/dL, albumin 3.4 g/dL, total bilirubin 0.9 mg/dL, AST (aspartate aminotransferase) 29 U/L, ALT (alanine aminotransferase) 16 U/L and INR (international normalized ratio) 1.2.

CT (computed tomography) abdomen/pelvis showed a 4.7 × 4.1 cm portocaval mass abutting the pancreatic head, hepatomegaly, portal, gastrohepatic, aortocaval lymphadenopathy up to 1.8 cm in short axis and 6.1 × 4.4 cm cystic lesion in the left adnexa. Chest CT showed pretracheal and subcarinal lymphadenopathy up to 2 cm in short axis. PET (positron emission tomography) scan revealed hypermetabolic areas in the neoplastic range in the left carotid sheath with SUV (standardized uptake value) of 5.8, paratracheal regions bilaterally (SUV 7.6), gastrohepatic ligament (SUV 6.8) and right vulva (SUV 5.8). The portocaval mass and the adnexal cyst were not metabolically active (Figure 1).

Mediastinoscopy with biopsy of the mediastinal lymph nodes demonstrated reactive lymphadenopathy with extramedullary haematopoiesis (Figure 2). CT-guided biopsy of the portocaval mass showed haemangiomatous pattern. Biopsy of the vulvar lesion showed invasive well-differentiated squamous cell carcinoma of the vulva (Figure 3). Depth of invasion on the biopsy specimen was 1 mm. Given the

Conclusion

Vulvar cancer is a rare malignancy with approximately 4500 new cases and 1000 deaths annually in the United States. Thorough and systematic preoperative workup is essential before treatment planning.

Introduction

Extramedullary haematopoiesis is the appearance of haematopoietic tissue outside of the bone marrow1. Sites most commonly involved are the spleen, liver, lymph nodes and mediastinum1,2. It is usually associated with pathologic entities that are causing ineffective myelopoiesis, leading to peripheral cytopaenias such as bone marrow neoplasms or haemolytic anaemias3,4. Several cases of solid tumours associated with extramedullary haematopoiesis have been reported5. Here we report the first case of extramedullary haematopoiesis occurring in a patient with vulvar cancer.

Figure 1: Whole-body positron emission tomography scan showing hypermetabolic areas in neck, mediastinum, upper abdomen and vulva.

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negative metastatic workup, we proceeded with radical local excision of the vulvar carcinoma and inguinal lymph node dissection. The surgical specimen was negative for residual tumour. All inguinal lymph nodes were negative. Final stage of the vulvar carcinoma was IA.

Discussion

Extramedullary haemopoiesis usually occurs in the reticuloendothelial system with spleen and liver involvement being the most prominent. It has also been described in other sites such as the gastrointestinal tract, lung, pleura, skin, breast, central nervous system, adrenals, kidneys and uterus.\(^1\)\(^-\)\(^5\)

It is considered to be a compensatory phenomenon when associated with anaemia or space-occupying bone marrow pathologic processes.\(^6\)\(^-\)\(^8\) Of note, our patient was not anaemic at presentation. Histologically, cells of myeloid, erythroid and megakaryocytic lineage are identifiable, the latter sometimes being the most obvious.

Extramedullary haemopoiesis may be associated with solid tumours. It has been identified in patients with angiomyolipoma, liposarcoma, spindle cell lipoma, cerebellar haemangioblastoma, lung carcinoma, colon cancer and renal carcinoma.\(^3\)\(^-\)\(^7\) There is evidence to support that the occurrence of extramedullary haemopoiesis in patients with solid tumours without bone marrow invasion is related to growth factors that are being secreted by the tumour and stimulate hyperplasia of circulating haematopoietic precursor cells. For instance, immunohistochemistry of solid tumours has demonstrated positivity for erythropoietin.\(^8\)\(^-\)\(^10\) This pathophysiologic mechanism is not related to the amount of the tumour since small, early-stage solid tumours have been associated with extramedullary haemopoiesis.\(^5\)

Conclusion

Here we present a case of early-stage vulvar carcinoma with concurrent extramedullary haemopoiesis in the lymph nodes. Vulvar cancer is a rare malignancy with approximately 4500 new cases and 1000 deaths annually in the United States. Thorough and systematic preoperative workup is essential before treatment planning. Extramedullary haemopoiesis should be included in the differential diagnosis in a case of vulvar cancer with diffuse lymphadenopathy, and histologic evaluation is required.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

References


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