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LETTERS TO THE EDITOR

Alveolar rhabdomyosarcoma mimicking acute leukemia at presentation

Dear Editor,

A 26-year-old woman presented with dyspnea on exertion and fatigue of one week's duration. Her past medical history was unremarkable. Overall physical examination revealed neither organomegaly, lymphadenopathy or sternum tenderness. The full blood count demonstrated a white cell count of 15×10^9/l, hemoglobin 9.8 g/dl and platelet count 70×10^9/l. The serum biochemistry profile was unremarkable apart from a mildly elevated lactic dehydrogenase level. Acute leukemia was suspected and the patient was referred for hematologic consultation. Examination of the peripheral blood smear disclosed a "left shift" with 7% myelocytes. Bone marrow aspiration and trephine biopsy were performed. The aspirate smears demonstrated a heavily infiltrated marrow by intermediate to large-sized pleomorphic blasts with primitive nuclei, fine chromatin pattern and vacuolated basophilic cytoplasm without granules. In many of these cells the cytoplasmic vacuoles were coalescent forming clear "lakes" (Figure 1). The malignant cells did not stain with myeloperoxidase or non-specific esterase and only rare cells stained with periodic acid-Schiff. The neoplastic infiltrate was diffusely distributed consisting approximately 80% of the marrow nucleated elements. Malignant cell clustering and rosette-formation were not observed. Based upon the above morphology, a high suspicion of rhabdomyosarcomatic bone marrow infiltration was raised, although rarely met in adults. Pathologic evaluation of the trephine biopsy documented leukemic-type bone marrow infiltration by tumor cells with a high nucleocytoplasmic ratio. The cells were negative for LCA, CD34, CD3, CD20, CD30, myeloperoxidase, Tdt and a variety of cytokeratins, while strongly expressed desmin, S-100, CD56 and smooth muscle actin, findings consistent with metastatic alveolar rhabdomyosarcoma. Whole-body CT scan identified a 2 cm lesion in the right maxillary sinus, indicative of the primary site. No solid organ metastases were found. The patient underwent intensive polychemotherapy with vincristine, actinomycin-D, cyclophosphamide (VAC combination), doxorubicin, etoposide and achieved a rapid remission.

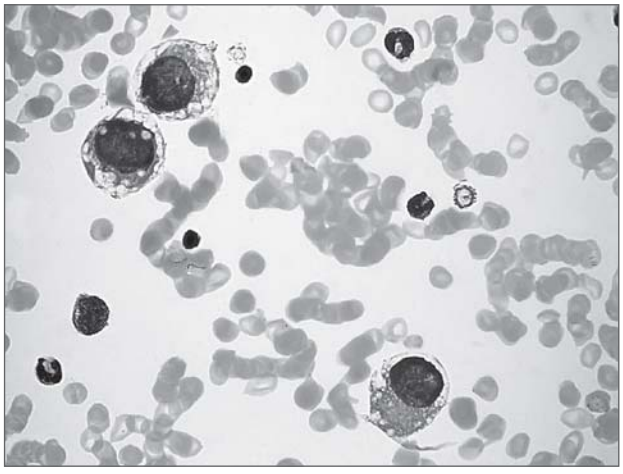

Figure 1. Bone marrow smear revealing rhabdomyoblasts with vacuolated cytoplasm without granules (May-Grunwald-Giemsa, ×1000).

Rhabdomyosarcoma is a highly malignant soft tissue tumor, described initially by Weber in 1845. It originates from primitive mesenchymal cells, the rhabdomyoblasts. It occurs primarily in the pediatric population, being the most common soft tissue sarcoma of childhood and is very rare in adults [1]. This tumor can occur everywhere due to its myogenic nature but the most common primary sites involved are the head and neck region, the genitourinary track and the extremities. There are 4 histological variants, namely embryonal, alveolar, pleomorphic and undifferentiated. Rhabdomyosarcoma is classified as one of the "small, round blue-cell tumors of childhood". This category also includes neuroblastoma, extraosseous Ewing's sarcoma, Wilm's tumor and acute leukemia/malignant lymphoma. Rhabdomyosarcoma may rarely present with extensive bone marrow infiltration and it can be confused histologically and cytologically with acute leukemia or high-grade non-Hodgkin's lymphoma [2,3]. However, the combination of positive immunohistochemical staining

for desmin, myogenin, actin and CD56 is diagnostic of rhabdomyosarcoma [4,5]. Cytogenetics are very helpful since the translocation t(2;13)(q35;q14) is found in 70% of alveolar rhabdomyosarcomas.

Although rare in adults, rhabdomyosarcoma has a particular interest for the clinical hematologist because of its high propensity for bone marrow metastases and its ability to masquerade as an hematological malignancy. Consequently, it should be considered in the differential diagnosis of patients presenting with a leukemic picture and atypical or unusual blasts.

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K. Liapis[1], N. Alevizopoulos[2], S. Delimpasi[1], J. Apostolidis[1], N. Harhalakis[1], D. Rontogianni[3], E. Nikiforakis[1]

[1]Department of Hematology and Lymphoma, [2]Department of Oncology, [3]Department of Histopathology, Evangelismos Hospital, Athens, Greece

Correspondence to: Konstantinos Liapis, MD. E-mail: kosliapis@hotmail.com

Glomus cells tumors in the male genital area

Dear Editor,

The penis glomus cells' tumor is a rare benign dermatic tumor deriving from special cells of the vessels called glomus cells. The tumor consists of broad vascular spaces with local clusters of glomus cells. The presence of glomus tumors in the male genital area is extremely rare and is manifested at an early age.

A 17-year-old male with a free personal and family medical history presented with a varicose lesion in the glans penis. The lesion pre-existed since his childhood and suddenly started to increase. The clinical examination revealed two varicose cyanotic lesions of 1 cm diameter each. The lesions were asymptomatic.

After local anaesthesia, we excised both tumors and applied photopexy with ND-Yag Laser (CW-10W) in the bottom of the lesions in order to achieve a better haemostatic and cosmetic result. The healing of the surgical wound and the cosmetic result were absolutely satisfactory one month after the operation. Histology revealed a typical glomus cells tumor.

Grauer and Burt first described such a tumor in the genital area in 1939 and since then only 9 cases have been published in the medical literature [1-5].

Glomus cells participate in the rapid thermoregulatory mechanism of the body through arteriovenous anastomoses during childhood and then tend to atrophy [1-3].

Local excision of the lesion is sufficient enough to deal with the problem, without the risk of recurrence.

In our case, we used for the first time a ND-Yag Laser photopexy, in addition to the surgical excision. Through this intervention we succeeded in having two advantages:

1) We achieved a better haemostatic result, since a postoperative bleeding could cause problems in the wound area.

2) We achieved a better cosmetic result, as just the

surgical excision could create deformed wound healing in this sensitive area of the body.

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N. Dimasis, K. Grekos, K. Efstathiou, S. Peftoulidis

Urology Department, Theagenion Cancer Hospital, Thessaloniki, Greece

Correspondence to: Nikolaos Dimasis, MD. E-mail: makedoni@otenet.gr

Multiple second primary tumors in a woman following breast cancer

Dear Editor,

The occurrence of a second malignancy in a patient with a known malignant tumor is not uncommon. It has been observed that a person with a malignant tumor may be more prone to develop another malignancy than would be expected by mere chance alone.

Metachronous primary malignancies are becoming increasingly frequent because of increase in the number of elderly patients and improvements in diagnostic techniques.

A 48-year-old female had undergone a lumpectomy 11 years ago for breast cancer. Histopathology had revealed an infiltrating ductal carcinoma of high and at certain sites moderate differentiation, T1N0M0, c-erbB2-positive, p53-negative, PCNA-positive in about 20% of the nuclei. Postoperative adjuvant radiation therapy to the whole breast and tamoxifen 20 mg/d for 5 years had been administered.

Ten years ago the patient had undergone a Whipple operation due to an extensive pancreatic adenocarcinoma stage IV (Yamaguchi classification).

Two years ago she underwent right colectomy for colon adenocarcinoma. Biopsy revealed a highly differentiated villous adenocarcinoma. Pericolic and mesenteric lymph nodes and surgical margins were negative.

In January this year the patient presented with vaginal bleeding for one week. Diagnostic work-up including abdominal and thoracic CT scans showed no evidence of disease. Mammography was negative. Because of the patient's medical history of pancreatic, colon and breast carcinomas we performed colonoscopy (negative) and gastroscopy which revealed a nodal formation on the mucosa in the anastomotic region (Whipple operation). Biopsies obtained from that region were negative.

Following diagnostic work-up the patient underwent a total abdominal hysterectomy with bilateral salpingo-oophorectomy and histopathology revealed a grade II endometrial adenocarcinoma, which invaded the whole myometrium up to 1 mm from the serosa. Radiation therapy to the pelvis was delivered postoperatively.

Multiple primary malignant neoplasms in a single patient have been well documented in the literature. The lesions can be limited to a single organ or may involve multiple organ systems. These lesions generally fall into 2 categories: 1) synchronous – in which the cancers occur at the same time or within 2 months apart; and 2) metachronous – in which the cancers present more than 2 months apart. Although a gynecological cancer may occur as a second primary cancer in patients with previous breast cancer as well as colorec-

tal cancer, the occurrence of 3 new second primary non-breast malignancies in a woman after breast cancer treatment is extremely rare.

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I. Kantzou[1], G. Sarris[1], N. Kalinoglou[2], V. Karagiannis[1], K. Hrisanthou[1]

[1]Department of Radiation Oncology-A and [2]Department of Gynecologic Oncology, Metaxa Cancer Hospital, Piraeus, Greece

Correspondence to: Ioanna Kantzou, MD. E-mail: ioannakantzou@mail.gr