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Primary Myeloid Sarcoma of the Gynecologic Tract: A Report of Two Cases Progressing to Acute Myeloid Leukemia

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Primary gynecologic myeloid sarcomas are rare, and their diagnosis is often difficult. Differential diagnosis includes lymphomas and carcinomas of the gynecologic tract. We report the clinical, morphological, immunohistochemical and cytogenetic features of two cases of chloromas of the female genital tract, which progressed to acute myeloid leukemia in spite of aggressive therapy.

Keywords: Myeloid sarcoma; Extramedullary myeloid tumor; Granulocytic sarcoma; Chloroma; Vagina; Uterus

INTRODUCTION

Extramedullary myeloid cell tumors, also termed chloromas or granulocytic sarcomas, are referred to in the new World Health Organization Classification of Tumors as myeloid sarcomas [1]. They usually occur in patients with hematological malignancies, but also as primary extramedullary tumors. Multiple locations have been described with a predilection for the bone, skin and lymph nodes [2]. Although the involvement of the gynecologic tract is frequent in the autopsy of women dying of leukemia [3], primary cases are rare and only one series including more than ten cases has been reported [4]. Most of the female chloromas involve the ovary and uterine and vaginal locations are less frequent [5]. The management of primary myeloid sarcomas with early antileukemic chemotherapy is associated with a lower rate of progression to acute leukemia and a longer overall survival [6]. We describe one case of primary uterus and ovarian chloroma and another one of vaginal location with their clinicobiological findings and the fatal results of treatment despite surgical resection, antileukemic chemotherapy, radiotherapy and hematopoietic progenitor cell transplantation.

CASES DESCRIPTION

Case 1

A 48-year-old woman presented in 1994 with vaginal bleeding. The histological features of a biopsy from the cervix uteri revealed a diffuse infiltrate of myeloblasts with positivity for CD43 and muramidase, and negativity for CD20, DR and CD3. Pelvic computed tomography (CT) scan showed a homogeneous soft tissue mass involving cervix, left mesosalpinx and ovaries. The CT of the thorax and abdomen showed no other lesions. Bone marrow aspirate and trephine biopsy were normal. After one cycle of induction chemotherapy with daunorubicin, cytarabine and etoposide, hysterectomy and bilateral oophorectomy was performed. A month later, antileukemic consolidation therapy was administered consisting of mitoxantrone and cytarabine and a subsequent cycle of amsacrine and cytarabine. A complete response was achieved. Six months after diagnosis, an autologous bone marrow transplantation was performed, but six weeks later the platelet count dropped, and a bone marrow aspiration showed an infiltration by 34% of myeloblasts. A diagnosis of an acute myeloid leukemia (M2 of the FAB classification)
was made. Palliative treatment was started and the patient died ten months after primary diagnosis.

Case 2
A 48-year-old woman was admitted in December 2000 because of vaginal pain and leukorrhea. Physical examination showed a mass on the right wall of the vagina. Biopsy of the tumor revealed a green macroscopic lesion with ulceration of the mucosa and an infiltrate of medium and large cells of blastic morphology and visible nucleoli (Fig. 1). On imprints, the cells were positive for CD43, naphthol ASD chloroacetate esterase (Leder stain), muramidase and LCA and negative for S100, cytokeratins, CD3, CD20, CD15 and CD34. In the pelvic CT a tumor of 6 cm involving the vagina between the rectum and uterus was observed (Fig. 2). Thorax CT, bone marrow aspiration and bone marrow cytogenetics studies were normal. Induction chemotherapy consisting of idarubicin, cytarabine and etoposide and then, a consolidation cycle of mitoxantrone and cytarabine were administered. After chemotherapy, the uterus and the proximal vagina were removed. No masses were identified and the histologic examination did not show granulocytic sarcoma. An autologous blood cell transplantation was performed. Two months later, two left breast tumors of 2 cm were observed, and a fine needle aspiration confirmed a breast relapse. Bone marrow morphology and cytogenetics were normal again. Salvage therapy with liposomal daunorubicin was started, but two months later, a new tumor on the right thigh was observed with features of granulocytic sarcoma. Bone marrow aspiration showed an infiltrate by monoblasts (Fig. 3), negative for myeloperoxidase and positive for inespecific esterases. Immunophenotypic analysis showed positivity for DR, CD13, CD33 and CD14. An acute myeloid leukemia diagnosis (M5a of the FAB classification) was made. Cytogenetic study revealed a complex cariotype: 47XX, t(1;4)(p34;q31), del(1)(p13), t(2;11)(p23;q13), del(5)(q12;q23), t(6;6)(p25;q15), +8[16]/46,XX [4]. The patient died 10 months after diagnosis.

COMMENT
Gynecologic myeloid sarcomas constitute about 15% of the extramedullary myeloid tumors, and have been described in adults as well as in pediatrics [2,7]. They constitute a rare entity, with about 50 cases reported [2–5,8–11], most of them as anecdotical cases. Clinical presentation may occur de novo or after diagnosis of a myeloproliferative, a myelodysplastic disorder or an acute myeloid leukemia. Post-mortem studies show that the involvement of the gynecologic tract in patients dying of myeloid leukemia is frequent [3]. The typical symptoms of uterus and vaginal granulocytic sarcomas include vaginal mass or bleeding. Morphology may show three types of cells depending on the degree of maturation: blastic, as the two cases reported herein; immature, composed primarily of myeloblasts and promyelocytes, and differentiated, composed of more mature granulopoietic cells [1]. The diagnosis includes immunohistochemical and immunophenotypic analysis, with positivity for myeloperoxidase, naphthol ASD chloroacetate esterase, muramidase and CD43. As in our second case, the monoblastic tumors
may be positive for non-specific esterase and CD14 [1,12]. A few cases of cytogenetic analysis in myeloid sarcomas have been reported, most of them with t(8;21)(q22;q22) abnormality [13]. The second patient described, presented a complex karyotype at the time of bone marrow relapse.

Myeloid sarcomas carry a poor prognosis, regardless of the age of the patient or the previous performance status. It is possible that blastic subtype imposes a worse prognosis, but this hypothesis has still to be confirmed. In a retrospective analysis, Imrie et al. [6] observed that the median survival was 22 months and showed a significantly lower rate of progression to leukemia and a longer overall survival for patients who received antileukemic therapy at the time of diagnosis of granulocytic sarcomas vs. those who did not receive chemotherapy. Moreover, a successful treatment of a case of chloroma of the uterus with surgery, induction therapy for acute myeloid leukemia and autologous bone marrow transplantation has also been reported [14]. Unfortunately, this approach was not effective in our two patients. Further therapy investigations are needed to improve the prognosis of this type of myeloid malignancies [15].

References