Table 1. Bone marrow findings during the clinical course.

	1997.2.26	1997.11.7	1998.12.5
Nuclear cell count (μL)	1,500	10,000	12,000
Megakaryocytes (μL)	0	0	0
Blasts (%)	0.0	0.4	0.8
Lymphocytes (%)	78.8	55.2	64.8
Erythroblasts (%)	3.6	7.6	10.4
Monosomy 7#	3.0%	2.0%	60.1%
Karyotypes	46,XX [19 cells]	46,XX [15 cells]	45,XX,-7, t(12,21) (q23; q22) [17 cells]/ 46,XX [3 cells]

Monosomy 7#: detected by FISH analysis using the CEP7 probe.

lative dose and duration of G-CSF therapy may be related to the risk of MDS arising from aplastic anemia.[5] It is interesting to note that 4/7 aplastic anemia patients showed monoclonality of the HUMARA gene, and that 3 of these 4 patients developed MDS, while all patients with polyclonality did not undergo transformation to MDS (*personal communication to Kuriya, S. et al, 1999*). Since our patient showed a monoclonal pattern on the HUMARA gene before the development of MDS and was administered a relatively low total dose of G-CSF, the present case suggests that monoclonality of the HUMARA gene in aplastic anemia is an important risk factor for MDS. Therefore, if patients with aplastic anemia reveal monoclonality of the HUMARA gene, they should be considered to have a borderline disorder between aplastic anemia and MDS.

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Key terms

Aplastic anemia, monosomy 7, G-CSF.

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Primary non-Hodgkin's lymphoma of the vagina

Primary lymphoma of the vagina can be successfully treated by pelvic radiation, but in young woman cytotoxic chemotherapy should be considered to preserve fertility. We report a case of non-Hodgkin's lymphoma of vagina (stage IE) with an optimal response to cytotoxic chemotherapy and disease-free survival for 13 years.

Sir,

Primary lymphoma of the genital tract is rare, accounting for 1% of primary extra-nodal lymphomas and localization in the vagina is ever rares; in fact only twenty-eight cases have been reported in literature,[1-6] although secondary involvement in advanced disease is found about 40% of cases.[7]

We report a case of non-Hodgkin's lymphoma (NHL) of vagina (stage IE) with an optimal response to cytotoxic chemotherapy (MACOP-B) and disease-free survival for 13 years.

A 25-year old female presented a 1-month history of vaginal discharge. Physical examination revealed an extensive tumor in the left wall of the vagina, extending into the pelvic cavity with a mass. The patient did not have fever, weight loss, or night sweats.

CT, US and MRI examinations showed a pelvic mass that measured 9 cm, with spread to the pelvic side wall. Urography showed stenosis of the left ureter. A vaginal biopsy did not show evidence of neoplasm. Because the histologic analysis was not significant despite the imaging signs and ureter is stenosis, the patient was submitted to staging laparotomy with partial exeresis of the pelvic mass and lymph node biopsy. The histologic analysis showed non-Hodgkin's centroblastic-centrocytic type lymphoma according to the Kiel classification. Ann Arbor classification was stage IEA, (Working formulation: F; Rappaport classification: DM). Bone marrow biopsy was negative.

The woman was treated with six courses of chemotherapy using the MACOP-B regimen (MTX, ADM, CTX, VCR, platinum, BLM, and leucovirin).

No hematologic toxicity was observed during chemotherapy, permitting 100% delivery of the planned dose. Grade III hair loss, grade II stomatitis and cystitis occurred.

After six cycles a complete pathologic response was achieved and the patient was disease-free at her latest follow-up in 1999. Four years after the treatment she gave birth to a female-infant.

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The prognosis of NHL of the genital tract seems to be favorable, even when the tumor is extensive.[8] According to one study the 5-year survival is 70%, and the most important predictive factor is the stage according to the Ann Arbor classification.[9]

Diagnosis of genital tract lymphoma is frequently delayed by the lack of specific symptoms, the commonest disturbances being bleeding, discomfort and vaginal discharge. The tumor is usually infiltrative and colposcopy biopsy may give false negative results, as in this case. This rare disease must be kept in mind in the differential diagnosis of the genital tract pathologies, because it can present at any age and may mimic other diseases of the vagina clinically and pathologically.[10] In standard management of lymphoma only stage I disease and, depending on grading and histologic subtype, stage II are treated exclusively with radiotherapy, whereas chemotherapy alone or in combination with radiotherapy is used in all other stages. Primary involvement of the vagina can be successfully treated by pelvic radiation, but cytotoxic chemotherapy should be considered in young woman to preserve fertility.

In conclusion we state that the polychemotherapy is a useful and effective treatment of vaginal NHL which concomitantly protects fertility.

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Key words

Non-hodgkin lymphoma, vagina, chemotherapy, and fertility.

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Homozygous Constant Spring: the first case described in the West

In this work we present the clinical and laboratory data and the molecular identification of homozygosly for Constant Spring in an Argentinian man with parents of Sicilian origin (Palermo). The presence of this in the homozygote form in the West and the difficulty of detecting it in heterozygotes by classical methods suggest that probably more cases eixist.

Sir,

α-thalassemias are a group of diseases caused by reduced synthesis of alpha globin chains. They can be classified into α^+ or α° thalassemia depending on whether the synthesis is totally or only partially absent. Hemoglobin Constant Spring (Hb Cs Sp) is the principal cause of non-deletion α-thalassemia in South-East Asia and Southern China.[1,2] This is caused by a mutation in the terminal codon of the α_2 globin gene (TAA-CAA)[3] which extends the transcription producing an unstable α_2 mRNA that encodes a protein of 172 residues instead of the 141 residues in normal globin. Thalassemic expression and the very low levels of Hb Cs Sp produced are caused by instability of α_2 mRNA and its intracytoplasmatic degradation.[4]

We present the clinical and laboratory data and the molecular identification of an Argentinian man of Sicilian origen.

He had had hemolytic anemia since childhood with splenomegaly and had received blood transfusions on several occasions.

Blood study revealed Hb 9.2 g/dL; RBC 4×10^{12}/L; PCV 32%; MCV 80 fL; MCH 23 pg; reticulocytes 8%. The quantification of Hb Cs Sp+HbA_2 was 8.6% and the HbF of 2.5%. Red blood morphology showed anisocytosis, moderate hypochromia, abundant target cells and basophilic stippling.

The possibility of an α-thalassemic deletion was ruled out by Southern blot and α_2 gene amplification followed by digestion with Hph and Nco I restriction enzymes screen out the existence of a specific mutations corresponding to the non-deletion α-thalassemias α^{Hph} and α^{Nco}. The fragments obtained after digestion with Mse I restriction enzyme correspond to a homozygote for some mutation in the region of the

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