Letters to the editor

The Rosai-Dorfman Syndrome in a 17-year-old woman with transformation into high-grade lymphoma. A rare case presentation

The Rosai-Dorfman Syndrome, better known as sinus histiocytosis with massive lymphadenopathy (SHML), is a rare benign disease of unknown etiology. Persistent painless lymphadenopathy due to expansion of sinuses infiltrated with benign histiocytes and plasma cells is the characteristic feature of SHML [1]. Here, we present a rare case of Rosai-Dorfman Syndrome with transformation into high-grade lymphoma.

Case history

A 17-year-old white woman was admitted to the Cancer Center in Krakow. She had been referred by the local hospital where she was treated with antibiotics due to enlargement of cervical lymph nodes associated with fever. On admission, the patient presented massive cervical, bilateral lymphadenopathy, fever and general malaise. Chest X-ray and abdomen CT scan revealed no pathological changes. Eosinophilia was reported in the bone marrow aspirate. A cervical lymph node was excised and a histopathological diagnosis of Rosai-Dorfman Syndrome was established. An elevated erythrocyte sedimentation rate, leukocytosis, anemia and hypergammaglobulinemia were present. Flow cytometry revealed an immune dysfunction (decreased T-helper lymphocyte subpopulation). The result of an anti-HTV antibody test was negative. Cytogenetic studies were performed on bone marrow cells obtained from a sternal biopsy. All 35 analysed metaphases were normal, with karyotype 46, XX.

No treatment option was chosen. The patient was followed every 3 months and after 5 years of observation a rapid progression of the disease was documented. An excised axillary lymph node revealed a high-grade lymphoma. VACOP-B chemotherapy was administered and resulted in a clinical partial remission. Cervical lymph nodes and pharynx were treated with radiotherapy. A total dose of 5000 cGy in 25 fractions was given. The patient was in remission for 15 months.

The second-line chemotherapy with the ESA regimen was used due to recurrence. The patient has been in remission for 3 months with moderate doses of prednisone (15 mg) as maintenance therapy.

Discussion

The Rosai-Dorfman Syndrome is a very rare condition with a benign course. Spontaneous remissions have been observed, although severe immune dysfunction has been found to be associated with SHML [2]. In a literature review of 462 cases of this disease, the development of lymphomas was observed in 6 patients [3].

According to our observation and information from the literature [4], cases with transformation into non-Hodgkin's high-grade lymphoma require intensive treatment. It is possible that a complete remission cannot be achieved because of heterogeneity of lymph node lesions (the co-existence of lymphoma and SHML cells) and observed remissions are associated with lymphoma component [5].

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References


Adjuvant therapy for breast cancer and psychological distress

Despite continuing improvements in the clinical management of aversive side-effects of chemotherapy treatment for cancer, there remains a widespread perception among health care professionals that patients experience increasing levels of emotional distress across the protracted course of infusions required for therapy [1, 2]. There is little empirical evidence to support this clinical impression, however.

We assessed emotional distress in 33 women receiving a complete course of adjuvant systemic chemotherapy, which consisted of a classic regimen of eight cycles of a standard combination of cytotoxic agents, CMF (cyclophosphamide (600 mg/m²), methotrexate (40 mg/m²), and 5-fluorouracil (600 mg/m²)) i.v. q 21d. Eligibility criteria included: stage I or II breast cancer; post surgery (e.g., mastectomy); 18+ years old; not pregnant; received pretreatment chemotherapy teaching and uniform antiemetic treatments (i.v.), as part of routine clinical care. Few patients (mean = 1.8 patients per infusion) used anxiolytic or antiemetic medications (p.o.) prior to infusions. Emotional distress on the day of each treatment infusion was assessed with a short version of the
Profile of Mood States (POMS), a classic classic mood adjective checklist [3], which patients completed in the waiting room before each infusion. Healthy (self-report), female, hospital employees (n = 31) completed the POMS on a single occasion.

Patients' total distress scores (POMS) were highest prior to the first infusion of chemotherapy and then declined (P < 0.01) to levels comparable to distress scores of hospital employees (Figure 1). Only at Infusion-1 were patient distress levels significantly higher than employee levels (P < 0.05). Patients' distress levels were not predicted by: age, ethnic group, marital status, whether they were scheduled for another treatment modality (e.g., radiation), or by the number of positive nodes (P > 0.05), but were related to tumor size and cancer stage (P < 0.05). None of these factors affected the pattern of reduced distress following Infusion-1.

These results, based on patients receiving CMF, are consistent with our previous studies using single-item measures of distress [4, 5] and provide no support for the widespread view that patients typically develop more distress as they go through repeated cycles of chemotherapy treatment for cancer. To our knowledge all the available data indicate that patients' distress levels are higher prior to treatment than at any other time during chemotherapy. The sources of this pretreatment distress have yet to be determined, but may include negative expectations of side-effects, loss of control, and/or fear related to this novel experience.

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References

Rapid intravenous premedication with dexamethasone prevents hypersensitivity reactions to paclitaxel

Introduction
Paclitaxel is a highly active new drug in the treatment of various types of tumors; however, in early phase I testing [1], the speed of clinical development has been partially hampered by hypersensitivity reactions. In the subsequent clinical trials paclitaxel was administered by continuous infusion over 24 hours and a premedication regimen consisting of oral corticosteroids administered 12 and 6 hours before treatment, orphenadrine and cimetidine, was instituted; this premedication was successful in reducing the incidence of severe hypersensitivity reactions to less than 5% [2].

Moreover, the 3-hour infusion has also proven safe, but the timing of premedication is still rather cumbersome for routine use in the outpatient setting [3]. In the current report we compare the use of a rapid intravenous premedication with dexamethasone with standard prolonged oral premedication in patients treated with paclitaxel given over 3 hours.

Patients and methods

Patients with advanced cancer who had progressed after standard chemotherapy were eligible for paclitaxel.

All patients had histologically confirmed diagnoses of cancer; other eligibility criteria included: age <70 years, an ECOG performance status ≤2, normal bone marrow, liver and renal functions.

Standard premedication consisted of oral prednisone 125 mg 12 and 6 hours prior to paclitaxel infusion.

The intravenous premedication was approved by the local ethical committee and the patients gave their informed consent before treatment. Intravenous premedication consisted of dexamethasone 20 mg administered by intravenous bolus immediately before the start of paclitaxel.

All of the patients were also premedicated with intramuscular orphenadrine 50 mg plus intravenous cimetidine 300 mg one hour before start of treatment The calculated dose of paclitaxel was diluted in 500 ml of saline and administered over 3 hours. Only glass containers and polyethylene-lined tubing were used for drug delivery. In-line filtration of the prepared solution during paclitaxel infusion using cellulose acetate filters of 0.22 µm pore size was performed.

During paclitaxel administration blood pressure, heart rate and respiratory frequency were recorded every 20 minutes.

Severe hypersensitivity reactions were graded, according to WHO [3], as reactions with one or more of the following: angioedema, hypotension (SBP < 80 mmHg), respiratory distress requiring bronchodilators or generalized urticaria.

If any of these symptoms occurred, paclitaxel infusion was stopped and treatment for anaphylaxis with additional corticosteroids, antihistamines and bronchodilators instituted.

If mild or moderate symptoms of hypersensitivity occurred, the infusion was temporarily discontinued and 250 ml of saline were administered before paclitaxel was started again.

Figure 1. Changes in psychological distress scores (mean ± SE) across a regime of outpatient chemotherapy (CMF, i.v.) for breast cancer.

![Graph showing changes in psychological distress scores](image-url)