Letter to the Editor

Infantile Malignant Osteopetrosis: Delay in Diagnosis Eliminates Chance of Cure

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Megadose Methylprednisolone for Osteopetrosis

To the Editor,

I enjoyed reading the paper of "Infantile malignant osteopetrosis: delay in diagnosis eliminates chance of cure" in the recent issue of the Journal 2002;19(3):411-415.

I believe the authors stressed well the importance of early diagnosis of the disease for cure by bone marrow transplantation.

Although several unsuccessful approaches for the treatment of osteopetrosis were brought to the attention, including very expensive IFNγ, GCSF and, 1α hydroxyvitamin D3, megadose methylprednisolon (MDMP) was not discussed, though included among the references (reference #11) in which given authors were not the real author who was me[1]. If the authors would read my letter carefully which they referred under the other peoples name they would notice that 13 patients with malignant osteopetrosis responded to our megadose methylprednisone (MDMP) administration hematologically and clinically. They may also used this treatment for their patients since it was concluded by me that "our approach would be helpful to those patients in whom bone marrow transplantation can not be carried out" May I remind that oldest patient of mine with osteopetrosis is alive at 21 years of age and teacher in the school of handicapped children though her site is limited and has major deformities.

Hypogammaglobulinemia G was also reported in 9 of our 13 children with malignant osteopetrosis whose immun response did not seem to hampered.

I have noticed marked discrepancy between patients Hb (3.64 g/dL) and RBC (6.55 million/mm^3) in the authors paper. I also could not understand why her hemoglobin electrophoresis, HbA_1, HbA_2, ferritin levels were determined in addition to serum iron and iron binding capacity as of erythrocyte sedimentation rate.

The authors stated that "in our patient blood picture showed leukoerythroblastosis which was suggesting bone marrow infiltration". Did they mean extramedullary hematopoiesis?

I have published some papers on the administration of MDMP for osteopetrosis which had been ignored by Turkish authors[1-4]? Why original papers from this country do not get recognition?

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Professor Şinasi ÖZSOYLU, MD

Department of Pediatrics and Hematology,
Fatih University Medical Faculty
Alparslan Türkeş Cad. No: 57
06510, Emek, ANKARA

Response

Thanks Professor Özsoylu for his interest on our case report published in Turk J Haematol 2002;19(3):411-415 with the following title: "Infantil malignant osteopetrosis: Delay in diagnosis eliminates chance of cure''.

It is nice to hear his experience about good outcomes of patients with malignant osteopetrosis after administration of megadose methylpredinisone. I will be pleased if Professor Özsoylu can submit full texts of his reports on the subject. We may have the chance of administration this therapy to patients

in whom bone marrow transplantation could not be carried out.

I want to correct the mistake about RBC of the patient which is 655.000/mm^3. Hemoglobin electrophoresis, ferritin, serum iron and iron binding capacity and erythrocyte sedimentation rate were determined before the diagnosis of osteopetrosis in an attempt to find the etiology of anemia and extramedullary hematopoesis. Due to rarity of the disease the pediatricians may have some difficulty in diagnosis if personal experience is absent on the subject. But this approach did not delay the diagnosis more than a week in our Center.

By saying "blood picture showed leukoerythroblastosis which was suggesting bone marrow infiltration", we want to mean infiltration of the marrow with sclerotic bone tissue. It is known that leukoerythroblastisis is associated with conditions infiltrating bone marrow (lymphoma, neuroblastoma, histiocytosis) and even with anemias as thalasemia major and other hemolytic anemias.

Professor Özsoylu complains ignorence of his reports by Turkish authors. But as he explained, in medline his name is not published among the authors of Reference 11. In text books also (Nathan and Oski, Hematology of Infancy and Childhood, 4th edition) prednisolon is not presented as a promising therapy for osteopetrosis. The statement is as follows: "Some patients responded to prednisone therapy, with again transient improvement because of decreased hypersplenism and reticuloendothelial suppression" MDMP is not mentioned although this edition was published after his first report on the subject which was in Arch Dis of Child in 1986.

Professor Nazan SARPER, MD

Caferağa Mah. Dr. Şakir Paşa Sok.
No: 7/4 Huzur Apt.
Kadıköy, İstanbul, TURKEY
e-mail: *nazan_sarper@hotmail.com*