Treatment of primary myelofibrosis

Primer miyelofibroz tedavisi

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To the Editor,

I have read with interest the presentation of three adult patients with primary myelofibrosis by Kar et al. in the recent issue of the journal [1]. The authors, without mentioning dose schedule, stated that parenteral and oral methylprednisolone was used in their patients.

Although our cases of primary myelofibrosis (PM) were not included in Kar et al.'s review, we were the first to successfully treat those patients, beginning in 1980, with megadose methylprednisolone (MDMP) [2-5]. Five children and four adult patients (a 60- year-old woman was not reported) with PM were all treated with MDMP (daily, 30 mg/kg for 3 days, followed by 20 mg/kg for 4 days, and then by 10, 5, 2 mg/kg doses, each dose given 1 week, continued with 1 mg/kg dose until hemoglobin (Hb) level reached 12 g/dl). Each dose was given within 5 to 10 minutes, intravenously earlier and orally recently. Each dose was given before 9 a.m., preferentially between 5 and 6 a.m.

It must be emphasized that MDMP treatment should not be compared with conventional prednisone administration (2 mg/kg, usually divided into 2 or 4 doses). Despite elevation in Hb level in the authors' patients, correction of the myelofibrotic process was not investigated by the authors. In 6 of our 9 patients, normalization of bone marrow was shown by bone marrow biopsy. During the 6-year follow-up period, no recurrences were observed in our patients.

Increased reticulin score of the bone marrow was mentioned by the authors. I would like to call attention to the fact that the presence of increased collagen and osteoblasts in the bone marrow as well as increased number of white cells and megakaryocyte precursors in the peripheral blood were more informative for the diagnosis [6.7].

References

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Author Reply

Dear Editor,

We would thank Prof. Özsoylu for critically evaluating our paper and giving some valuable comments.The review of cases in our paper specifically refers to the distinct entity of primary autoimmune myelofibrosis reported in the literature. In this study we have not included cases of idiopathic myelofibrosis and hence references quoted by Prof. Özsoylu have not been mentioned.

The patients 1 and 2 received iv methyl prednisolone 1g IV OD x 3 days followed by wysolone 50 mg PO. At 5 months and one year follow up, there was improvement in cytopenias and spleen size was regressed. The patient 3 received oral steroids (dose 1mg/kg/day). At one year of follow up the total leucocyte counts and hemoglobin were normal and paravertebral mass had regressed. However, spleen was

palpable 2 cm. below costal margin. Unfortunately, all three patients had a short period of follow up and thus a repeat trephine biopsy could not be performed to assess the marrow fibrosis.

As regards to increased collagen fibrosis and osteosclerosis, that would indicate progression of the same disease process to more advanced stages. None of our cases showed osteosclerosis and there was absence of significant leukoerythroblastosis as opposed to cases idiopathic myelofibrosis.

Sincerely,

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