Reviewer's report

Title: Unusual Manifestation of Erdheim-Chester Disease

Version: 1 Date: 21 April 2011

Reviewer: Julien Haroche

Reviewer's report:

Dear Editor,

Thank you very much for the opportunity to review the manuscript entitled “Unusual manifestations of Erdheim-Chester disease”, by Pan et al.

In this manuscript, the authors report the case of a 69-year-old man with abdominal symptoms in whom a laparotomy, along with other typical features, lead to a diagnosis of ECD.

While the diagnosis of biopsy-proven ECD with abdominal involvement is not questionable, as the patient has several features of ECD and a typical histology, I think the discussion and the literature review should be completed, particularly with some of the important data regarding this rare disease published recently.

Minor issues include the following points:

- In the abstract section, please add the PS100 may sometimes be positive (#20%).
- In the case report section please add appropriate reference regarding MRI findings just below figure 6 were reference # 1 is not correct: please refer to Dion et al, Bone involvement in Erdheim-Chester disease: imaging findings including periostitis and partial epiphyseal involvement. Radiology. 2006;238:632-9.
- It would also be interesting to have a dosage of IgG4 in this case. Was it done?
- In the beginning of the discussion section I would specify the number of patients worldwide with less than 400 known ECD cases referring the recent Blood paper of 2011 (see Arnaud et al. CNS involvement and treatment with interferon-α are independent prognostic factors in Erdheim-Chester disease: a multicenter survival analysis of 53 patients. Blood. 2011 Mar 10;117(10):2778-82).
- It would improve the discussion in the first paragraph to mention and refer the gastro-involvement of other histiocytoses, such as Langerhans Cell histiocytosis, which is much well established than in ECD.
- In the fourth’s paragraph I would also refer to more recent data regarding pulmonary involvement as this patient died of respiratory failure with Arnaud et al. Arthritis Rheum. 2010 Nov;62(11):3504-12 as well as Brun AL. Erdheim-Chester disease: CT findings of thoracic involvement. Eur Radiol. 2010 Nov;20(11):2579-87, for the description of the typical pulmonary involvement. It would also be interesting for the iconography of this case to show a picture of the
pulmonary infiltration just after figure 1.

- Again, in the second page of the discussion section, the percentages of lung involvement should also be implemented by more recent data referring for incense to Arnaud et al. Arthritis Rheum. 2010 Nov;62(11):3504-12.

- The last paragraph section of the discussion section should also be enriched and partly re-written with more “up to date” data regarding treatment with interferon alpha with the papers of Braiteh et al. (Successful treatment of Erdheim-Chester disease, a non-Langerhans-cell histiocytosis, with interferon-alpha, Blood. 2005 Nov 1;106(9):2992-4), Haroche et al. (Variability in the efficacy of interferon-alpha in Erdheim-Chester disease by patient and site of involvement: results in eight patients. Arthritis Rheum. 2006;54:3330-6) and also see Arnaud et al. CNS involvement and treatment with interferon-{alpha} are independent prognostic factors in Erdheim-Chester disease: a multicenter survival analysis of 53 patients. Blood. 2011 Mar 10;117(10):2778-82). The last sentence of the discussion should therefore be slightly modified due to recent advances in treating ECD.

Level of interest: An article of importance in its field

Quality of written English: Acceptable

Statistical review: No, the manuscript does not need to be seen by a statistician.

Declaration of competing interests:

I declare that i have no competing interests