

Reply to Commentary on “A Case of Erdheim-Chester Disease with Asymptomatic Renal Involvement”

Hyun Jung Lee, MD, Tae Min Kim, MD, PhD

Department of Internal Medicine, Seoul National University Hospital, Seoul National University College of Medicine, Seoul, Korea

We express our thanks to Dr. Cavoli for interest in our case [1] and shared experience of your case. A 60-year-old female with Erdheim-Chester disease (ECD) suffered from azotemia with bilateral hydronephrosis within 2 years of initial diagnosis. Author regarded a metformin-associated lactic acidosis combined with azotemia as the cause of her presentations. Although nearly 10% of cases with metformin-associated lactic acidosis were met for all three criteria (i.e., arterial pH < 7.35, blood lactate > 5 mmol/L, and detectable plasma metformin concentration) [2], plasma metformin concentration was not given in this patient. In addition, fever, abdominal pain, and leukocytosis raised a possibility of urinary tract infection accompanied by azotemia.

Regardless of causality, a wait-and-see policy was possible after the correction of azotemia in this patient. Similarly, our patient did not any specific treatment for ECD until August 2012 (more than 4 years from initial detection of left renal mass) and did not any genito-urinary symptoms [3]. Therefore, a wait-and-see policy might be acceptable for asymptomatic ECD patients. Considering that interferon- α appeared to be effective against bilateral hydronephrosis [4], its response might be important to design a treatment strategy in this patient. Recently, 13 (54%) of 24 ECD patients harbored *BRAF* V600E mutation [5] and the identification of *BRAF* V600E mutation would provide a theoretical rationale for the use of *BRAF* inhibitors in patients with *BRAF* V600E-positive ECD with any symptoms or disease progression.

References

1. Li Cavoli G. Commentary on “A case of Erdheim-Chester disease with asymptomatic renal involvement”. *Cancer Res Treat.* 2012;44:279.
2. Kajbaf F, Lalau JD. The criteria for metformin-associated lactic acidosis: the quality of reporting in a large pharmacovigilance database. *Diabet Med.* 2012 Sep 21 [Epub]. <http://dx.doi.org/10.1111/dme.12017>.
3. Lee HJ, Lee KY, Shin DY, Lee YG, Choi SY, Moon KC, et al. A case of Erdheim-Chester disease with asymptomatic renal involvement. *Cancer Res Treat.* 2012;44:146-50.
4. Haroche J, Amoura Z, Trad SG, Wechsler B, Cluzel P, Grenier PA, 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.
5. Haroche J, Charlotte F, Arnaud L, von Deimling A, Helias-Rodzewicz Z, Hervier B, et al. High prevalence of BRAF V600E mutations in Erdheim-Chester disease but not in other non-Langerhans cell histiocytoses. *Blood.* 2012;120:2700-3.

Correspondence: Tae Min Kim, MD

Department of Internal Medicine, Seoul National University Hospital, Seoul National University College of Medicine, 101 Daehak-ro, Jongno-gu, Seoul 110-744, Korea
Tel: +82-2-2072-3559, Fax: +82-2-764-2199, E-mail: gabriel9@snu.ac.kr

Received December 4, 2012 **Accepted** December 10, 2012