

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

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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

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