Reviewer's report

Title: Extrarenal nephroblastomatosis in children: A report of two cases

Version: 2

Date: 15 August 2014

Reviewer: Lydia Pecker

Reviewer's report:

1. Major Compulsory Revisions

* Abstract of case reports should be revised to 1) introduce in the first sentence why two cases of extra renal nephroblastomatosis are of interest and importance to the reader. Next, the two cases should be more briefly summarized. For instance, since each patient underwent surgical resection observation as follow-up, this may be stated in one sentence. Finally, the conclusions of the abstract appear to stray from the information reported in the cases. As your cases are neither extrarenal nephrogenic rests, nor associated with teratoma, I am confused about why these are the ultimate conclusions of your abstract. Conclusions should direct the reader to the instructive nature of the reported cases and may highlight any future work indicated by the unknowns associated with extrarenal nephroblastomatosis.

* Citations in this paper are thoughtful and extensive. However, I am left wondering: Why do two additional cases of extrarenal nephroblastomatosis without unusual clinical sequelae or dramatic presentations warrant publication? The authors are should highlight the educational value, clinical importance or management challenges raised by the cases they identified.

* Given the association of Wilms Tumor with multiple genetic syndromes, the authors should remark on the overall health of the two patients identified. Were these children with cancer predisposition syndromes? Did they have stigmata of Beckwith Weideman, Denys Drash or WAGAR?

* Were pathology specimens tested for genetic testing for WT1 or WT2 mutations? Would this information, if available, have changed management?

* Both patients were managed without adjuvant chemotherapy or radiation. On what basis was this decision made?

* Conclusions must be revised to be drawn directly from the two presented cases.

2. Minor Essential Revisions

* Lines 71-72 seem to conflict with lines 80-81. Was the mass invasive to the testicle or not?

3. Discretionary Revisions
* Management of nephrogenic rests and extrarenal nephroblastomatosis is a clinical challenge because of the concern for malignant transformation. The interesting finding of testicular nephroblastomatosis in an undescended testicle, which also carries a risk for malignant transformation might be explored.

* The authors are encouraged to present the pathology findings once and to tie these findings more closely to clinical concerns and management decisions.

**Level of interest:** An article of limited interest

**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.