Reviewer's report

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

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Reviewer: Gordan M Vujanic

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This is a nicely written paper presenting two cases of a relatively uncommon lesion - extrarenal nephroblastomatosis. The authors say that they have identified two cases in their 10-year material - it would be interesting to know how many cases of Wilms tumour they have had in the same period.

Major points:

line 142: "the distinguishing characteristics of nephroblastoma are frank atypia, atypical mitoses and marked pleomorphism" - it is not quite clear what is meant by this, are the authors listing histological features of anaplasia to which they refer in the following sentence? If so, than the diagnostic features of anaplasia are not precisely listed as they include atypical mitoses, marked nuclear enlargement and hyperchromasia. Anyway, the features they have listed as "characteristics of nephroblastoma" are not quite right, or the wording of this sentence is not good, so should be rephrased.

line 146: "... and moderate pleomorphism, but no atypia." - the rests do not show any pleomorphism and if it is present, than the diagnosis of Wilms tumour is more likely.

line 175: "In the case of renal nephrogenic rests, however, approximately 40% are associated with Wilms tumor." - it is actually the other way around, ~40% of unilateral Wilms' tumours are associated with nephrogenic rests. The incidence of nephrogenic rests not associated with Wilms tumour is not known.

line 179: "some nephrogenic rests are associated with teratoma and therefore, have a poor prognosis [27]; these lesions require surgical treatment with additional chemotherapy." - this is new to me, presumably, the authors want to say that some extra-renal nephrogenic rests may be associated with teratoma? If so, it is still inaccurate to say that such cases require additional chemotherapy - teratoma in children, even the immature ones, are treated with surgery only, and I don't think that there are any accepted protocol which advise chemotherapy for teratomas associated with nephrogenic rests. In the quoted reference (27), the authors only comment that in some cases chemotherapy but do not recommend it routinely.

Minor points

line 134: "Differential diagnosis between benign extrarenal nephroblastomatosis
and malignant nephroblastoma is mandatory..." - there is no benign nephroblastoma, so the word malignant should/could be replaced with "extra-renal"

line 140: "on cytological examination..." presumably, the authors wanted to say "on histological examination..."?

line 265-274: Figures legends are not what they should be, describing what is presented in the figures, we do not need to know which staining was used (that can go in the brackets, with the magnification) but what we are supposed to see in them. I am not entirely convinced that Fig 2C shows a blastemal component.

line 155: "In the second patient, the ectopic nephrogenic rests were thought to have originated from mesonephric or metanephric tissue." - what about the first case, is it not the same possibility?

**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:**

no competing interests