Reviewer’s report

Title: Pheochromocytoma as a rare cause of hypertension in a 46 X, i(X)(q10) Turner syndrome: a case report and literature review

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Reviewer: Troy Puar

Reviewer’s report:

This is a unique case of Pheochromocytoma occurring in a patient with Turner's syndrome, which is worthwhile to report.

The novelty of this case, is that in patients with Turner's syndrome developing hypertension, we classically are taught to consider differentials like cardiovascular abnormalities (aortic stenosis / coarctation), however, physicians may not consider other important secondary causes like pheochromocytoma. This point should be emphasized, and appropriately expressed, in the abstract and discussion. Failure to consider can lead to mortality (as in the other reported case) Pheochromocytomas is now known to be hereditary in 30-40% of cases, and it is even more likely in young patients. Hence, it will strengthen the case report if appropriate genetic screening could be done for this index case before submission. Are patients with chromosomal abnormalities (Turners) at even greater risk? This will be something important to add to the current knowledge.

My other minor comments will be:

1. the presentation of the case report.

   - In the initial presentation, authors may want to mention absence of radio-femoral delay (impt feature of coarctation of the aorta)

   - should ideally mention the laboratory workup of pheochromocytoma, Before the abdominal CT results, as abdominal CT should not be used as a first line investigation to workup 'sudden onset hypertension' (this was mentioned in the abstract, but should be mentioned first in the case presentation, before the CT)

   - discuss about the high washout of the pheo, which is often quoted as a marker of benign adenomas, but is not 100% when used to rule out a pheo (pheos can mimic many lesions). more importantly, the lesion on CT appears very heterogenous and vascular, which should be used to determine the type of lesion more than the washout.

   - MIBG - reason for doing this? Not routinely recommended in the latest endocrine society guidelines, but in view of her young age, and possibly other paragangliomas this would be warranted.
- line 51 you may want to clarify "no recurrence developed at 24 months of follow-up" via biochemical tests or CT imaging?

- biochemical results. This pheo was predominantly secreting noradrenaline / normetanephrine, and you may want to comment on that (and also compare with the other cases in literature)

- Was there a reason the urinary normetanephrine was normal? This is unusual as normetanephrine is more sensitive to norepinephrine. What was the urine volume and renal function?

- AS this is a literature review, there should be greater discussion about the other cases in the literature and how they compare to the current index case, and if the authors feel that Turner's may predispose to Pheo, or is it just two unusual diseases occurring in the same patient by chance. Again the genetic tests will be very relevant.

- Figure 3 is not adequately labelled (A-B-C-D)

Are the methods appropriate and well described?
If not, please specify what is required in your comments to the authors.

No

Does the work include the necessary controls?
If not, please specify which controls are required in your comments to the authors.

Unable to assess

Are the conclusions drawn adequately supported by the data shown?
If not, please explain in your comments to the authors.

No

Are you able to assess any statistics in the manuscript or would you recommend an additional statistical review?
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Not relevant to this manuscript

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Please indicate the quality of language in the manuscript:

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