Author's response to reviews

Title: 11 cm Haughton D left cervical aortic arch aneurysm

Authors:

Pankaj Kaul (pankajkaul784@btinternet.com)

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Author's response to reviews: see over
A 56 year old man presented with sudden loss of consciousness while driving and was found to have an 11cm Haughton D type left cervical aortic arch aneurysm with normal brachiocephalic branching and normal descending thoracic laterality but with considerable tortuosity and redundancy of aortic arch. The aneurysm arose between the left common carotid artery and the left subclavian artery. It compressed and stretched the left common carotid artery, compressed the pulmonary trunk and the left pulmonary artery, stretched the vagus, left recurrent laryngeal and left phrenic nerves and caused extreme deviation of trachea, severely compromising the tracheal lumen. Patient underwent successful interposition graft replacement of distal aortic arch under total circulatory arrest and selective unihemispherical cerebral perfusion.

Left cervical aortic arch is a rare developmental anomaly of aorta and only around 70 cases are described in world literature. It is further classified into five distinct types on the basis of brachiocephalic branching, arch and descending aortic laterality and redundancy of the transverse aorta. Left cervical aortic arch, in general, and Haughton D type, in particular, is prone to aneurysm formation due to abnormal flow patterns and tortuosity and redundancy of aorta. The unique features about the surgical anatomy and presentation of this patient include the origin of the aneurysm from the aortic arch in the narrow segment between the left common carotid and the left subclavian arteries with extreme
displacement of both. There was a separation of more than three inches between the origins of left common carotid artery and the left subclavian artery. There was extreme tortuosity and redundancy of the distal aortic arch and disproportionate vertical enlargement of the aneurysm. A number of anatomically diverse structures, including trachea, left common carotid artery, left subclavian artery and main and left pulmonary artery, left vagus and phrenic nerves, had been compressed, displaced and distorted. The aneurysm did not rupture despite reaching a size of eleven cms.

My response to the reviewer’s comments:

**Reviewer:** Chris Klonaris

**Reviewer's report:**

- **Major Compulsory Revisions**

The author needs to rewrite the abstract and discussion sections, so as to justify and explain to the reader why this case is unique. In other words, the author should highlight the points that differentiate this case from all other similar reports already published. Both Abstract and Discussion sections have been rewritten to highlight the differences between this report and the few reports previously published.

- **Minor Essential Revisions**
Some sentences are too large and need to be separated. (e.g. “The unique features about the surgical anatomy and presentation of this patient include the origin of the aneurysm from the aortic arch in the narrow segment between the left common carotid and the left subclavian arteries with extreme displacement of both, a separation of more than three inches between the origins of left common carotid artery and the left subclavian artery, the extreme tortuosity and redundancy of the distal aortic arch, the disproportionate vertical enlargement of the aneurysm with compression of a number of anatomically diverse vital structures, and the fact that the aneurysm did not rupture despite reaching a size of eleven cms”) This sentence has been broken up into smaller sentences to aid comprehension. The manuscript contains also repetitions that should be omitted. All repetitions have been omitted now.
Level of interest: An article whose findings are important to those with closely related research interests

Quality of written English: Needs some language corrections before being published

This has been done as detailed above

Reviewer: Teruhisa Kazui

Reviewer's report:
The author reported a case of left cervical aortic arch complicated with aneurysm formation, rare congenital anomaly which is defined as the apex of the aortic arch is located in the neck, and Haughton type D according to the Haughton Classification (normal brachiocephalic branching, redundant transverse aorta and left-sided descending aorta) which was successfully treated with interposition graft replacement of aortic arch between the arch distal to LCCA and the proximal descending thoracic aorta under total circulatory arrest with hemispherical (left common carotid artery) selective cerebral perfusion.
Although the manuscript is well written, I have the following comments. The figures are redundant and so some of them should be deleted.

1. Figure 2 should be deleted because the MR scan is not clear enough. 
   A new clearer figure has been supplied.

2. The diagram should be added to Figure 3 to delineate the anatomical situation of cervical aortic arch, arch branches, and aneurysm.
   All the anatomical branches have been labelled in the picture now.

3. Figure 4 should be deleted because aortogram is not clear enough. 
   Aortogram has been deleted.

4. Figures 5, 6, 7 should be deleted
   I have labelled all the anatomical structures in the intraoperative picture, called fig 5 in the earlier submission and now fig 4. This gives an unparalleled view of the aneurysm and its extent and the distortion of some of the structures.
   I have deleted fig 6 even though it illustrated the important point that occasionally the distal anastomosis to the descending aorta is facilitated by a small anterolateral extension.
Fig 7 serves the important purpose of showing the distance spanned by the graft after the excision of the aneurysm and hence the sheer size of the aneurysm. Its deletion will detract from that important message and so I feel it is relevant.

All the figures have been renumbered in the text due to 2 deletions.

I thank the reviewers for their kind comments.

All changes in the text have been highlighted in red.