The Maya Behind Moyamoya—The Two Extremes of the Disease : Authors' Reply

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To the Editor: The main purpose of the article was to highlight “*collaterals*” as the reason behind the varied presentations of moyamoya disease. We did not present these patients to explain their rarity in terms of extreme presentations. We do accept and have also mentioned in the article that moyamoya disease has varied presentations. We just chose these two cases at the opposite ends of the spectrum of manifestations of moyamoya disease as we found an interesting finding of “numerous collaterals *vs.* very minimal collaterals” being the reason behind the varied presentations in these two patients. The first child had episodes of headache occasionally with mostly symptom free intervals and the second child had very rapidly progressive course with profound neurological deficits. They are definitely at the two ends of the spectrum, though we do not argue that they are the most extreme presentations of moyamoya ever reported in literature [1].

Both the patients did have bilateral angiographic findings to qualify a diagnosis of definitive case of moyamoya though we could mention only the significant findings in the brief report. Both the cases had narrowing of the branches of internal carotid artery with collaterals on both sides. The second child did have collaterals though very

minimal, but not absent collaterals. The term used in the article was “almost no/almost nil collaterals” not “absent collaterals” [1].

Bilateral findings on MRI brain with normal caliber of right internal carotid artery could be explained by the narrowing of the branches of internal carotid artery on the right side in the second case.

We quoted the Mugikura et al. study not to explain the severity of presentation of the second case, but to quote the importance of collaterals behind varied presentations of moyamoya [2].

All the secondary causes of moyamoya (arteriosclerosis, autoimmune disease, brain neoplasm, history of cranial radiation, Down syndrome, head trauma, neurofibromatosis, meningitis) were excluded in both cases. Only important, relevant information explaining the main purpose of the article could be discussed in the article because of word limit constraints.

We do accept that “The Maya behind Moyamoya” still remains unclear and more prospective studies are necessary to find the reason behind the varied presentation of the disease.

References

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