Formation of bilateral arteriovenous malformations

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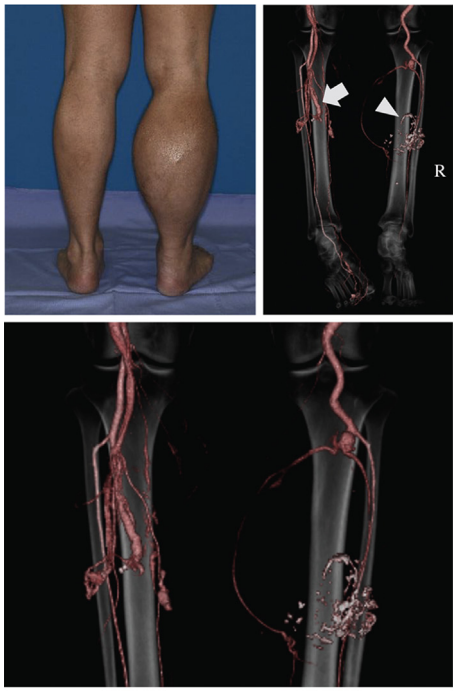
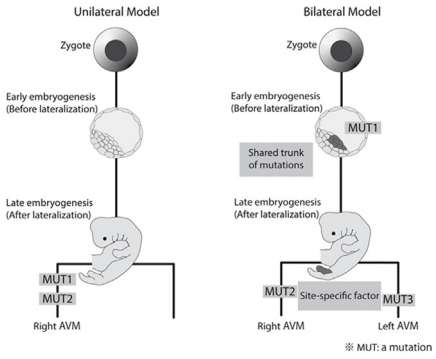
A 58-year-old Japanese man, with no family history of vascular anomalies, had presented with swelling of the right lower extremity (*A*). He did not have a thrill, edema, or any symptoms specific to well-described syndromes. Computed tomography angiography revealed an arteriovenous (AV) shunt and a mosaic pattern of various signal intensities, indicating a chronic expanding hematoma in the right lower extremity (*A* [*arrowhead*]; and *B*/Cover). It also described an AV shunt in the left lower extremity (*A* [*arrow*] and *B*/Cover). These findings suggested the presence of bilateral AV malformations (AVMs). He underwent partial resection of the AVM lesion on the right side because of continuous bleeding (*S1* [*arrow*]; and *S2-S4*). He had had anemia before surgery; however, he had recovered. No other abnormal changes were observed in the laboratory tests. Genetic examinations were not performed because they are not routine for the diagnosis of AVMs. The lesions were well-controlled even 1 year after the surgery (*S5*). The patient provided written informed consent for the report of his case details and relevant imaging studies.



DISCUSSION

An extracranial AVM is a type of vascular anomaly characterized by AV shunts.[1] Similar to venous malformations, sporadic AVMs are thought to arise from somatic mosaicism caused by somatic mutations at embryologic stages.[2,3] Because the number of reported bilateral extracranial AVMs has been far less than that of unilateral AVMs,[4] one working hypothesis is that a unilateral AVM can arise from mutations that have occurred in the leg after lateralization (*C*, unilateral model). For bilateral sporadic lesions, another working hypothesis is that the first hit mutations occur early in embryogenesis before



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lateralization.[5] In the present case, we believe that site-specific factors might have played a significant role in the manifestation of the lesions because of the high similarity in the location of the bilateral lesions (*C*; bilateral model). Because an asymptomatic AVM was coincidentally found at an identical location on the contralateral side of the symptomatic AVM lesion, it is possible that the number of multiple or bilateral AVM cases could be greater than previously reported. We believe the details of the present patient will help in understanding the etiology and pathogenesis of "sporadic" AVMs in the future.

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