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Diagnosis: Moyamoya disease

Comment: A gross photograph of the cerebral arteries, which had been dissected from the specimen, revealed thread-like main channels, with extensive proliferation of small collateral vessels. The middle cerebral arteries were thrombosed, and there was also recanalization of vessels. This patient was not of Japanese ancestry. Moyamoya (Japanese for “something hazy,” the appearance of the cerebral circulation on angiography, in which the small collateral vessels look like a puff of smoke) disease is common in Japan, with two peaks in incidence, one in the first decade of life and the other in the fourth decade. Various predisposing causes have been proposed for it, including growth factors, prostaglandins and EB virus, but the etiology is unknown. Moyamoya syndrome is more common, and it is seen in association with a variety of conditions, including in sickle cell anemia, after cranial irradiation, and in Down syndrome.

References:

