A 42-year-old man presented with a tense, tender 24-cm cranial lump in the left face and frontal-temporal-parietal region (figure, A and B). Slow growth and limited local medical resources delayed presentation for care. Firm subcutaneous nodules were observed in other locations. A diagnosis of neurofibromatosis type I-like syndrome was made; confirmatory genetic testing was not performed. The 5-kg tumor was completely excised surgically, including an intracranial portion that grew through an orbital-frontal cranial defect. Histologic examination revealed a neurofibroma with blood vessel hyperplasia and extensive invasiveness, containing spindle, S100-positive tumor cells (arrows). After 4 years of follow-up, besides the removed left eye damaged by the tumor, he had a satisfactory recovery.

X. Zhang, MD, PhD, X. Mao, MD, PhD, W. Zhang, MD, PhD, J. Zhang, MD, PhD, W. Cao, MD, PhD, Shaanxi, PR China

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Correspondence & reprint requests to Dr. Xiang Zhang: xzhang@fmmu.edu.cn
