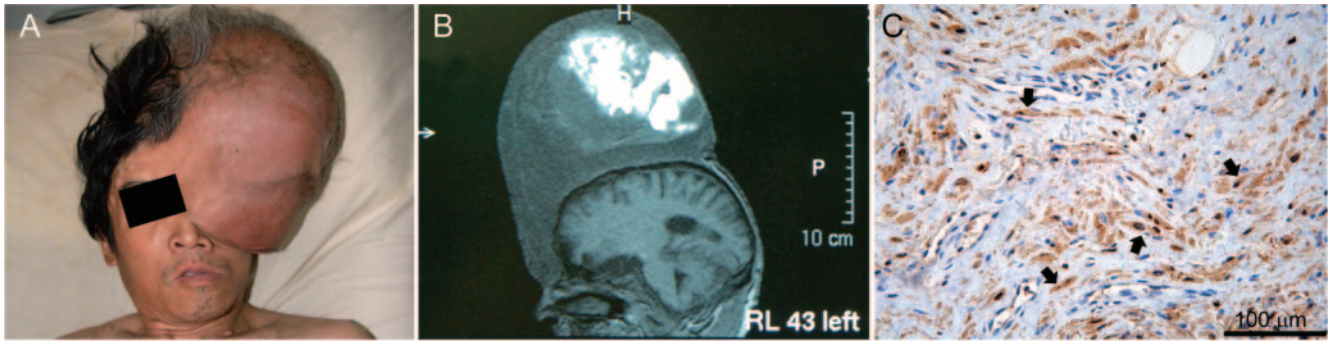
Giant head neurofibroma

Figure Imaging of giant head neurofibroma



(A) Image of the giant head neurofibroma. (B) MRI demonstrated hyperintense signal lesion in the upper part of the mass and mild hypointense signal in the other area on T1-weighted image. (C) Histologic examination revealed a neurofibroma with blood vessel hyperplasia and extensive invasiveness, containing spindle, S100-positive tumor cells (arrows).

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[1]; 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[2] revealed a neurofibroma (figure, C). 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

Author contributions: Xiang Zhang: design of the study, statistical analysis, and writing. Xinggang Mao: analysis of the data. Wei Zhang: drafting the manuscript. Jianning Zhang: revising the manuscript. Weidong Cao: interpretation of the data.

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

1. Messiaen L, Yao S, Brems H, et al. Clinical and mutational spectrum of neurofibromatosis type 1-like syndrome. JAMA 2009;302:2111–2118.
2. Jouhilahti EM, Peltonen S, Heape AM, Peltonen J. The pathoetiology of neurofibromatosis 1. Am J Pathol 2011;178:1932–1939.