

**49th ANNUAL DIAGNOSTIC SLIDE SESSION, 2008
DIAGNOSES AND REFERENCES**

MODERATOR: Anthony T. Yachnis, M.D.

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Case 2008-9

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Diagnosis: Cortical fibromatosis in a case of Von Recklinghausen's disease

Comment: An MRI of the lower extremities of this patient revealed greatly enlarged nerves, consistent with a diagnosis of NF1, and the patient had typical cutaneous manifestations also. The resected left leg mass was plexiform, cellular and aggressive in appearance, consistent with a MPNST. The resected brain mass had diffuse cortical thickening, and it was not discrete. The cortex was replaced by fibrous tissue, with islands of GFAP-positive tissue within the fibrous tissue (which stains yellow with the HPS stain). These tissue islands were also positive for vimentin and S-100, and they had CD68 positive cells within them as well. On electron microscopy, the background material was composed entirely collagen. This cortical fibromatosis did not involve the leptomeninges. It also lacked cellularity and mitotic activity. The relationship to NF1 is uncertain.

References:

None. Dr. Ludwin was unable to find any reports of similar cases.