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Neurofibromatosis**

Tony Kim, Robert J. Foust and Saeid Mojtahedi

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MR Imaging of Asymptomatic Brainstem and Spinal Cord Lesions in Sisters with Neurofibromatosis

The increased incidence and multiplicity of intracranial and intraspinal neoplasms associated with neurofibromatosis are well-known phenomena. We report here two sisters who have neurofibromatosis and multiple CNS neoplasms and in whom asymptomatic brainstem and spinal cord lesions were discovered and evaluated by MR imaging of the head and spine.

Case Reports

Two sisters, ages 37 and 26 years, developed bilateral acoustic neuromas and multiple intracranial meningiomas. Their mother and two other siblings also had bilateral acoustic neuromas.

During a routine MR study of the brain of the older sister, an abnormality (possibly a mass) was found at the cervical medullary junction. This patient had no symptoms referable to this area, and postoperative changes were extensive. Therefore, we decided to follow the patient clinically and to obtain another MR study later. She eventually developed symptoms in her right leg, and the MR study was repeated (Fig. 1). A cystic brainstem glioma was discovered and drained during surgery.

The younger sister underwent a routine MR study of the brain and spine. A large expansile lesion was discovered arising from the medulla and extending inferiorly to the level of the C2 vertebral body (Fig. 2). In addition, three cystic intramedullary lesions, which later proved to represent syringohydromyelia, were found in the thoracic and lower cervical spinal cord. The patient had no symptoms referable to her lesions and was followed clinically.

The younger sister developed ataxia 1 year later, but a repeat MR study showed no detectable changes in the previously seen lesions.

The older sister remains asymptomatic at 1-year follow-up, although reoccurrence of the cystic brainstem lesion was shown by follow-up MR.

Discussion

Neurofibromatosis is a hereditary disease that is transmitted as an autosomal dominant trait with an almost 100% penetrance rate, but a highly variable degree of expressivity. Neurofibromatosis arises in the general population in 1 of 3000 people, half from spontaneous

mutation [1]. Neurofibromatosis consists of two distinct forms [2]. Neurofibromatosis 1, described by von Recklinghausen, is characterized by café au lait spots, intertriginous freckling, Lisch nodules, and skin neurofibromas. Neurofibromatosis 2, which afflicts the family of these two sisters, is characterized by bilateral acoustic neuromas, which are rarely found in neurofibromatosis 1.

The incidence of CNS tumors in patients with neurofibromatosis 1 may be up to six times that of the general population with a frequency of 5–10% [1, 3]. Intracranial tumors typically include optic and trigeminal nerve gliomas, acoustic neuromas, astrocytomas, meningiomas, hamartomas, and glioblastomas [4]. Intraspinal tumors include intradural extramedullary neurofibromas or meningiomas and intramedullary ependymomas, astrocytomas, or syringohydromyelia [4, 5].

The incidence of CNS tumors in patients with neurofibromatosis 2 is even higher. In a review of 49 patients with neurofibromatosis 2, Rodriguez and Berthrong [5] found CNS gliomas in 45% and syringomyelia in 20%. The distribution of the CNS gliomas in their series was 5.9% cerebral, 16.4% cerebellar and brainstem, 22.4% cervical, 23.9% thoracic, 17.9% lumbar, and 13.4% sacral and below. Syringomyelia occurred in 28.6% of the cases involving the cervical, thoracic, and lumbar spinal regions, and in 14.2% of cases involving the sacral region.

The value of CT as an intracranial screening examination in neurofibromatosis has been described [7]. The sensitivity of MR in the detection of these CNS tumors is equal to and in most cases superior to CT, particularly in the posterior fossa and spine, where the majority of these tumors occur. MR has additional diagnostic advantages: lack of ionizing radiation, freedom from intrathecal iodine contrast, and the abilities to show spinal cord tumors in their complete cranio-caudal extension and to differentiate between cystic and solid tumor areas [7].

The asymptomatic lesions described in these sisters illustrate the need for thorough and periodic examinations of the brainstem and spinal cord in addition to the brain, especially if the patient suffers from neurofibromatosis 2. Early detection of treatable lesions will permit earlier and more effective intervention. MR imaging, with its great sensitivity and high resolution of the posterior fossa and spinal cord in addition to the other advantages already described, may be

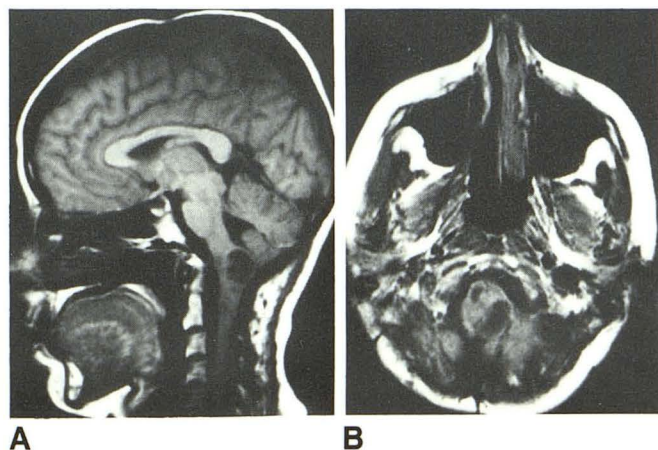


Fig. 1.—A and B, T1-weighted sagittal (A) and axial (B) brain scans of older sister show low-signal cystic lesions arising from anterior and posterior aspects of brainstem.

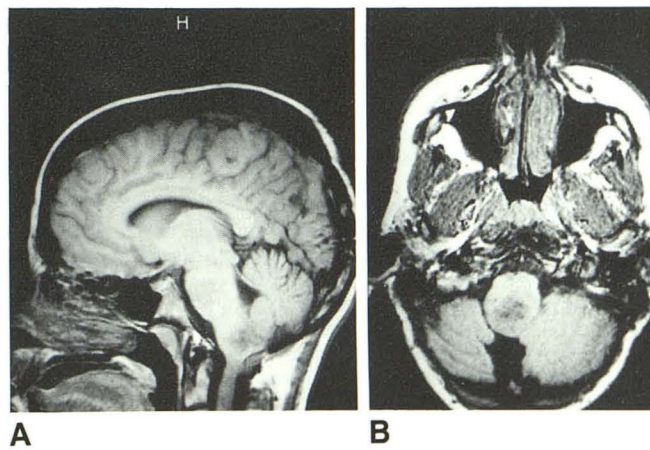


Fig. 2.—A and B, T1-weighted sagittal (A) and axial (B) brain scans of younger sister show an ill-defined, expansile lesion within brainstem.

a superior diagnostic and screening examination in patients with neurofibromatosis.

Tony Kim
Robert J. Foust
Saeid Mojtahedi
*University of Chicago
Chicago, IL 60637*

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