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Radiographic findings in Moebius and Moebius-like syndromes.

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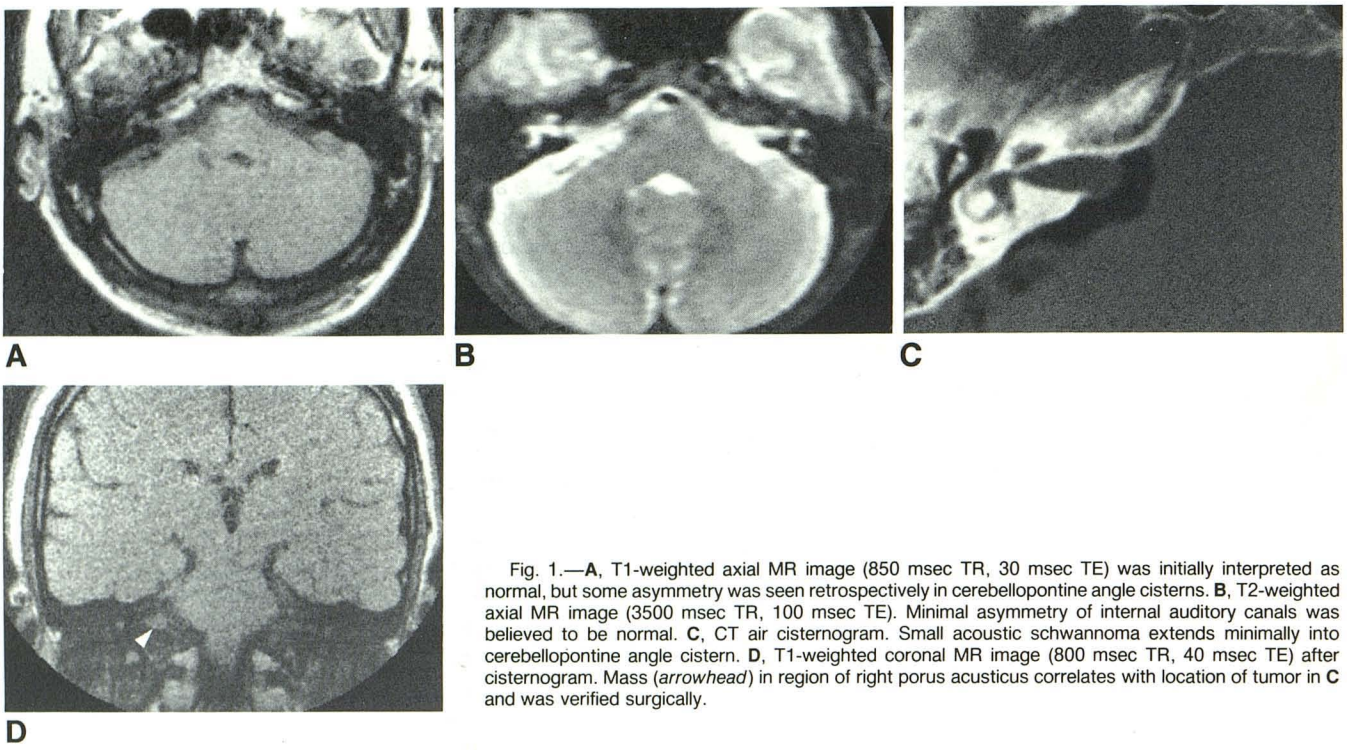

Fig. 1.—**A**, T1-weighted axial MR image (850 msec TR, 30 msec TE) was initially interpreted as normal, but some asymmetry was seen retrospectively in cerebellopontine angle cisterns. **B**, T2-weighted axial MR image (3500 msec TR, 100 msec TE). Minimal asymmetry of internal auditory canals was believed to be normal. **C**, CT air cisternogram. Small acoustic schwannoma extends minimally into cerebellopontine angle cistern. **D**, T1-weighted coronal MR image (800 msec TR, 40 msec TE) after cisternogram. Mass (*arrowhead*) in region of right porus acusticus correlates with location of tumor in **C** and was verified surgically.

canal in this case was obliquely situated with respect to the horizontal plane (fig. 1C) and, therefore, was scanned more directly in the coronal than in the axial plane. In addition, the increased spatial resolution obtained with a 256 × 256 matrix is important in detecting a small lesion.

In conclusion, we believe that in the evaluation of sensorineural hearing loss by MR, both coronal and axial thin sections (5 mm or less) are essential. In addition, optimum imaging parameters must be used, particularly a strongly T1-weighted sequence. There is little doubt that with further improvement in MR technology, especially surface-coil imaging, it will become the imaging method of choice. For the present, CT air cisternography remains the gold standard in the evaluation of possible acoustic neurinomas.

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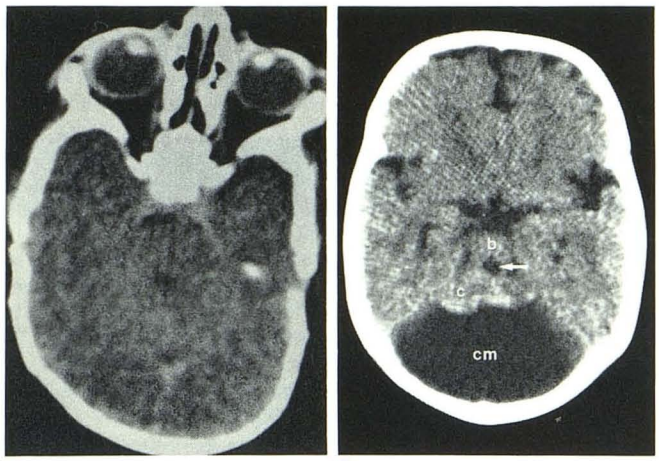
Radiographic Findings in Moebius and Moebius-like Syndromes

Moebius syndrome is a congenital nonprogressive neuromuscular disorder characterized by bilateral facial and lateral rectus paralysis. Radiographic studies are useful in evaluating the syndrome, especially in documenting any associated malformations that might indicate that the Moebius sequence is part of a more extensive central nervous system process that may not be clinically evident during the neonatal period. We are reporting the clinical and radiographic findings in a case of classic Moebius syndrome and in a case of Moebius-like syndrome.

Case Reports

Case 1

This male infant born of an uncomplicated term pregnancy and vaginal delivery had medial deviation of both eyes, absent facial



1 2

Fig. 1.—Case 1. Axial CT image through lenses of globes. Mild inward deviation of both eyes suggests bilateral sixth nerve palsies. Brain structures are normal except for minimal flattening of left cerebral hemisphere because of skull deformity.

Fig. 2.—Case 2. Axial CT scan through posterior fossa. Large cerebrospinal-fluid-density space (cm). Cerebellar hemispheres (c) and brainstem (b) are hypoplastic and fourth ventricle (*arrow*) is rostrally located.

movement, poor suck, and absence of the right nipple, pectoralis major muscle, and the digits. The neurologic examination was remarkable only for bilateral fifth, sixth, and seventh cranial nerve palsies. The diagnosis of Moebius syndrome associated with the Poland sequence was made. Computed tomography (CT) of the head showed deviation of both eyes suggesting bilateral sixth nerve palsies (fig. 1), and no intracranial abnormality. At the age of 1 month, the child died of aspiration pneumonia.

Case 2

This female infant was a 2620 g product of a 37-week-gestation pregnancy. At birth, absent facial movement was noted, and on examination bilateral seventh nerve palsies and mild generalized hypotonia were evident. Cranial sonography suggested a small cerebellum with an enlarged cisterna magna. Cranial CT showed that most of the posterior fossa was filled with cerebrospinal-fluid density with only a small part of the cerebellum being present behind a somewhat deformed and rostrally located fourth ventricle (fig. 2). The brainstem was also small. An air cisternogram via lumbar puncture showed immediate filling of a large cisterna magna and no ventricular filling. She developed bilateral sixth nerve palsy and at 18 months of age was noted to be hyperreflexic and mildly ataxic. A repeat head CT scan was unchanged.

Discussion

Moebius syndrome classically presents with congenital nonprogressive bilateral oculofacial diplegia, although the paralysis can be unilateral and incomplete and may involve other cranial nerves, such as in our first case. Moebius syndrome may also be associated with micrognathia, hypoplasia of the tongue, branchial cleft musculature defects, and the Poland sequence (absent pectoralis muscle, syndactly, and limb reduction defects) [1–3]. Most patients with Moebius syndrome will have a normal CT head scan except for medial deviation or inward gaze of the eyes as a result of the lateral rectus paralysis, as seen in our first case. CT and cranial sonography are particularly valuable in identifying any other concomitant brain malformations that can occur in a patient who presents with a Moebius syndrome such as in our second case. A dysplastic, small brainstem and cerebellum resembling the Dandy-Walker variant malformation as in our case 2 was described in one other report [4]. The radiographic evaluation of patients with Moebius syndrome, which is most often normal, helps to exclude possible treatable or progressive disorders such as infarction, hemorrhage, trauma, infection, hydrocephalus, or tumor that could mimic a Moebius-like syndrome. We believe careful radiologic workup with particular attention to the brainstem, cerebellum, base of the skull, and facial musculature, with appropriate radiographic evaluation of the extremities when indicated, is warranted in Moebius syndrome. High-quality CT scanning and sonography are two noninvasive complementary methods ideal for the initial evaluation of these patients.

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