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Neurofibromatosis Type 1 and Arnold-Chiari Malformation

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To the Editor:

Neurofibromatosis type 1 (NF-1) or von Recklinghausen disease is the most common neurocutaneous syndrome. It is characterized by the appearance of various cutaneous stigmata, neurological manifestations, and an increased susceptibility to develop tumors.¹ Although it is frequently

associated with a wide variety of central nervous system (CNS) dysplasias, the association with Arnold-Chiari malformation type I is unusual.

A 60-year-old woman with a past history of systemic hypertension, hiatus hernia, and iron-deficiency anemia, was seen in our outpatient clinic for lesions on her neck that had been present the years and that caused discomfort due to friction. On physical examination of the skin, multiple soft fibromas were observed in the cervical region; however, a large number of hyperpigmented macules with a homogeneous, light brown color and well-defined borders were also observed, mainly on the trunk, though also at the root of the limbs, and 9 of them were over 15 mm in diameter, and there were also groups of hyperchromic macules between 2 and 10 mm in diameter in both axillas, clinically consistent with lentigo simplex (Crowe sign). The patient stated that those lesions had been present since birth, and that her father had similar spots. Based on these findings, the patient was diagnosed with NF-1 and was referred to the neurology and ophthalmology departments to exclude CNS and optic nerve involvement. Ophthalmologic examination was normal, with no evidence of Lisch nodules. The patient had no neurological symptoms and neurological examination revealed generalized, symmetrical muscle hyperreflexia but no other alterations. Cerebral magnetic resonance imaging (MRI) showed herniation of the cerebellar tonsils into the upper cervical canal, below the level of the

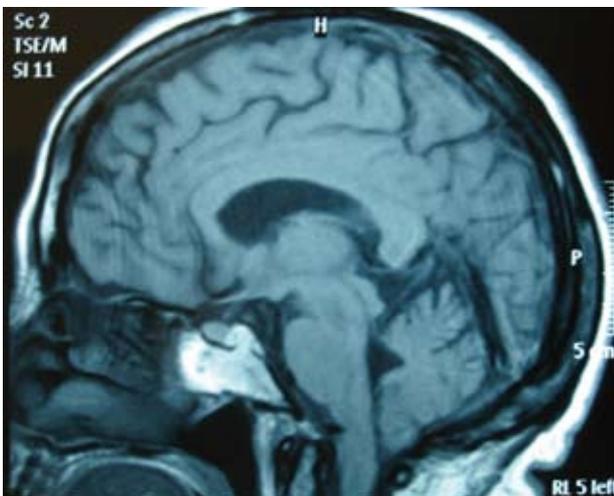


Figure 1. Sagittal cerebral magnetic resonance image: the T1-weighted image shows herniation of the cerebellar tonsils into the upper cervical canal, below the level of the occipital foramen, consistent with Arnold-Chiari malformation type I.

foramen magnum (Figure), consistent with an Arnold-Chiari malformation type I. Blood tests, abdominal ultrasound, and a bone scan were also performed, with normal results. As the Arnold-Chiari malformation type I was an incidental finding, and as the patient had no symptoms, no treatment was required.

Numerous neurological abnormalities related to NF-1 have been reported, including macrocephaly, unilateral sphenoidal dysplasia, glioma of the optic chiasm, optic nerve, diencephalon, or brainstem, meningiomas, cranial nerve schwannomas, plexiform or intraspinal neurofibromas, hamartomas, stenoses of the cerebral aqueduct, heterotopias, and hyperintense lesions in the basal ganglia, internal capsule, and cerebellum on T2-weighted MRI.² Vascular malformations or dysplasias have also been reported, some of which have been related to other neurocutaneous syndromes, such as Sturge-Weber syndrome.³

On the other hand, the association of Arnold-Chiari malformation type 1 and NF-1 is rare.^{1,2,4-10} We present the case of a patient with NF-1 and with an asymptomatic Arnold-Chiari malformation type I diagnosed as an incidental finding on performing cerebral MRI. The diagnosis of NF-1 was made in accordance with current criteria.¹¹ Although the Arnold-Chiari malformation type 1 is usually associated with other abnormalities such as basilar impression, occipitalization of the atlas, scoliosis, or spina bifida, in our case the patient only presented herniation of the cerebellar tonsils and of the medial part of the inferior lobe of the cerebellum into the cervical canal.

The table summarizes the cases of Arnold-Chiari malformation type 1 associated with NF-1 reported in the literature. The prevalence of the Arnold-Chiari malformation type 1 is of 1 in 3700² and that of NF-1 is 1 in 4500 to 1 in 6700,¹¹ and the probability of having both conditions is therefore very low (1:16 650 000 to 1:24 790 000).² However, it must be remembered that Arnold-Chiari malformation type 1 can be asymptomatic, being detected as an incidental finding in neuroimaging studies, as in the case described here; the routine use of cranial-cervical magnetic resonance imaging in all patients with NF-1, even in the absence of symptoms or signs of neurological disease, will therefore probably reveal a higher frequency of this association. On this subject, Tubbs et al¹² demonstrated that up to 8.6% of patients with NF-1 (17 of a series of 198 patients) presented an asymptomatic Arnold-Chiari malformation type I.

The frequent detection of lesions affecting the CNS in patients with NF-1, including spina bifida, hydrocephalus, and meningocele, as well as the frequent presence of bone lesions such as scoliosis, macrocephaly, or sphenoidal dysplasia, suggests that this association is actually more than incidental. The gene mutation (17q11.2) found in NF-1 facilitates the abnormal proliferation of tissues and

Table. Cases or Case Series of Arnold-Chiari Malformation Type I Associated With Neurofibromatosis Type 1 Reported in the Literature

Authors	Year	Cases	Symptoms
Herrero A et al ¹⁰	2007	Woman, 23 y	Headache
Hara M and Arakawa M ¹	2005	Woman, 29 y	Gait disorder, sensory and urinary disturbances
Tubbs RS et al ¹²	2004	Series of 198 cases of Chiari I	8.6% of cases with asymptomatic NF-1
Chakravarty A et al ⁹	2002	Woman, 22 y	Optic nerve glioma, scoliosis, syringomyelia
Guistini S. et al ⁸	2002	2 cases	Asymptomatic (case 1) and hydrocephalus (case 2)
Batissela PA et al ²	1996	Boy, 11 y	Headache
Dooley J et al ⁷	1993	Boy, 16 y	Asymptomatic
Tominga T et al ⁶	1991	1 case	Headache, hydrocephalus
Affifi AK et al ⁵	1988	2 cases	Hydrocephalus (both)
Parkinson D and Hay R ⁴	1986	1 case	Rhinorrhoea, fistula

the subsequent appearance of ectodermal and mesodermal dysplasias and various tumors. The pathogenesis of Arnold-Chiari malformation type 1 appears to be related to hypoplasia of the posterior fossa, leading to herniation of the cerebellum through the foramen magnum. In this context, congenital central nervous system dysgenesis could represent a common pathogenic mechanism for both conditions.¹³

In summary, and in view of what has been discussed, we consider that Arnold-Chiari malformation type 1 should probably be considered as one of the CNS dysplasias to be excluded in all patients with NF-1. In our opinion, cerebral MRI should therefore be performed in all patients with NF-1.

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Conflicts of Interest

The authors declare no conflicts of interest.

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Hair Follicle Nevus: A Case Report and Review of the Literature

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To the Editor:

The hair follicle nevus is a very rare hamartoma that is usually congenital or appears in the first years of life, and presents as a papule, plaque, or nodule on the face.¹⁻⁶



Figure 1. Clinical image of the lesion. Velvety plaque on the lower eyelid of the right eye.

We present the case of a 16-year-old girl with no past history of interest, who was seen for a lesion on the lower eyelid of the right eye and that had been present since birth. The lesion was a homogeneous, skin-colored, velvety plaque of approximately 1 cm × 0.5 cm, with poorly defined borders, and with no orifices or comedones on its surface (Figure 1). The lesion was asymptomatic and had always been stable, with no sudden changes in size, shape, or appearance. A 4-mm punch-biopsy was taken from the center of the lesion, revealing a tumor with follicular differentiation. Serial sections were performed of the whole biopsy, observing a proliferation of mature hair follicles in similar stages of differentiation in the upper regions of the reticular dermis, surrounded by a highly cellular stroma (Figure 2). The connective tissue sheath of all the follicles presented marked fibrous thickening (Figure 3). No central cystic cavity was found in any part of the sample. The diagnosis of hair follicle nevus was made on the basis of the clinical features and the histological findings. As the lesion was asymptomatic and did not trouble the patient from a cosmetic point of