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MR of Leptomeningeal Spinal and Posterior Fossa Amyloid

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Summary: We report an unusual cause of leptomeningeal MR enhancement, amyloid, along the surfaces of the spinal cord and brain stem and in the spinal subarachnoid space, with sacral intradural and epidural deposition. Type I familial amyloid polyneuropathy may cause amyloid deposition along the leptomeninges of the spinal cord and brain in addition to the visceral organs and the peripheral somatic and autonomic nerves.

Leptomeningeal enhancement has been described on MR images of the brain and spinal canal in patients with metastatic lesions from non-CNS primary tumors, such as breast and lung carcinoma, lymphoma, and leukemia; in drop metastases from intracranial medulloblastoma, ependymoma, glioblastoma multiforme, and pineal tumors; in infectious meningitis; and in granulomatous disorders, such as sarcoidosis and tuberculosis (1-3). The purpose of this article is to report the MR finding of intradural leptomeningeal enhancement caused by amyloid protein deposition in type I familial amyloid polyneuropathy (FAP), one form of hereditary generalized amyloidosis.

Case Report

A 35-year-old woman had a 4-year history of bowel and bladder symptoms, including urinary retention and frequency, constipation, lower back pain, and numbness and tingling extending to the buttocks and posterior thighs. Physical examination revealed Adie's tonic pupils, absent Achilles reflexes, decreased sensation to pin prick in the left S-2 and S-3 dermatome, and decreased vibration along the right lateral foot. The patient's family history included the early death (at age 45) of her mother. Her maternal grandmother, maternal uncle, and his son (who underwent liver transplantation) all had familial amyloidosis. And, recently, the patient's male and female siblings have been found to have amyloidosis.

An MR examination performed at an outside institution (not available) showed a mass in the sacral canal. Two months after presentation, a partial surgical resection of this mass from the sacral intrathecal sac and epidural space was performed. The pathologic specimen consisted of pale gray-white firm tissue. Histologic examination showed pale eosinophilic amor-

phous avascular material embedded in a background of fibrofatty tissue, which was positive for amyloid, stained red with Congo red dye, and showed green birefringence under polarized light (Fig 1). The amyloid stained strongly with antibodies against transthyretin (immunoperoxidase method), confirming the nature of the amyloid as AA, which is reported in familial forms of amyloidosis. Electron microscopic examination confirmed the fibrillar nature of the amyloid. Laboratory tests did not reveal any monoclonal gammopathy.

Five months after surgery, the patient's earlier symptoms persisted, and jabbing pains involving the right foot and posterior calves appeared. More prominent were transient episodes of right-sided numbness, beginning in her face and eventually, over 30 to 45 minutes, extending over the right side of her body. Occasionally, these episodes were followed by headache; at other times, only sensory symptoms, dysarthria/aphasia or right-sided weakness, occurred. Physical examination now also showed decreased sensation over S1-5 dermatomes, decreased position sense in both feet, and orthostatic blood pressure changes. MR imaging of the brain and whole spine was performed with and without infusion of gadopentetate dimeglumine. Contrast-enhanced MR images revealed leptomeningeal enhancement in the brain stem and cerebellum and in the cervical, thoracic, and lumbar spine; sacral intradural and epidural enhancing deposits were present along a 4-cm length of S-1; and smooth, scalloped bone erosion of the right sacral foramen was seen adjacent to the amyloid deposit (Fig 1B-E).

Discussion

Amyloidosis comprises a group of diseases in which protein tissue deposits have common morphologic, structural, and staining properties but variable protein composition (4, 5). All amyloid fibrils are arranged in a β -pleated sheet that produces the characteristic staining and optical features of amyloid. All amyloids stain red with Congo red dye, and the deposits exhibit green birefringence when viewed under polarized light.

The clinical classification of amyloidosis includes primary (de novo), secondary (a complication of a previously existing disorder), familial, and isolated types (5). Familial amyloidosis may occur in patients with familial Mediterranean fever or it may arise from transthyretin mutations. Approximately 30 such mutations have been described. Clinically, patients are

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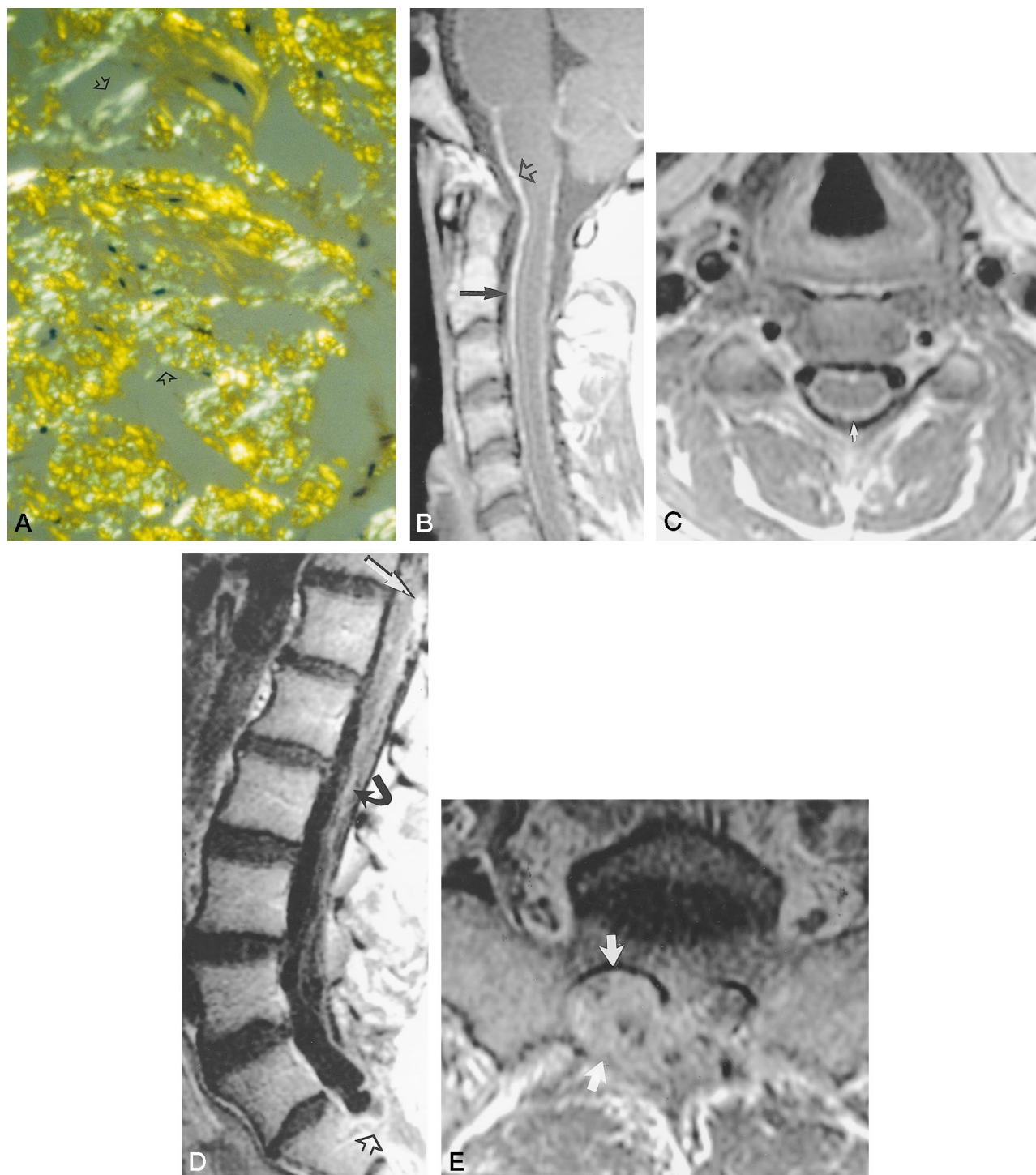


FIG 1. 35-year-old woman with type I familial amyloid polyneuropathy.

A, Apple green birefringence (arrows) is seen on tissue section stained with Congo red and viewed under polarized light (original magnification $\times 400$).

Contrast-enhanced sagittal T1-weighted (500/11/3 [TR/TE/excitations]) (B) and axial T1-weighted (500/13/2) (C) cervical MR images show linear pial leptomeningeal enhancement (closed arrow) along the surfaces of the cervical spinal cord, medulla (open arrow), pons, and cerebellum.

D, Contrast-enhanced sagittal T1-weighted (550/11/2) lumbar MR image shows nodular and linear pial and arachnoidal leptomeningeal enhancement dorsal to the conus medullaris (closed straight arrow), enhancing nerve roots of the cauda equina (curved arrow), and enhancement within the sacral canal (open arrow).

E, Contrast-enhanced axial T1-weighted (466/10/2) MR image shows enhancing epidural and intradural mass filling the sacral canal (bottom arrow) and encasing the lower signal intensity nerve roots. Note smooth, scalloped bone erosion of the right sacral foramen (top arrow).

susceptible to neuropathic, cardiopathic, or nephropathic complications. Type I familial hereditary generalized amyloidosis is inherited as an autosomal dominant gene mutation with a single amino acid substitution of methionine for valine at position 30 in transthyretin.

Transthyretin, previously called prealbumin because it migrates ahead of albumin in standard electrophoretic separations, is a serum carrier of thyroid hormones and vitamin A. The mutant transthyretin is deposited as extracellular twisted β -pleated sheet fibrils in peripheral somatic and autonomic nerves and visceral organs; it causes autonomic and peripheral somatic disorders, and the disease is ultimately fatal (5). The peripheral neuropathy is generally distal, symmetric, and progressive, initially sensory rather than motor, with dysesthetic numbness, reduced pain, and temperature sensation, involving the lower extremities more often than the upper extremities (4). Progressive autonomic dysfunction includes light-headedness, syncope, orthostatic hypotension, neurogenic bladder, impotence, and gastrointestinal disturbance.

Cerebrovascular amyloidosis is characterized by amyloid deposits in the media and intima of arteries and arterioles of the leptomeninges and cortices. It is seen in Alzheimer disease, Down syndrome, and in some elderly persons without dementia. It may be a cause of spontaneous cerebral hemorrhage in normotensive elderly persons and of sporadic cerebral amyloid angiopathy. It is also seen in Icelandic- or Dutch-type hereditary cerebral hemorrhage and amyloidosis (6). The cerebrovascular system is rarely involved in systemic amyloidosis in which there is diffuse amyloid deposition in various organs. Nonfamilial primary systemic amyloidosis rarely involves the CNS but has been known to involve the pituitary and choroid plexus.

Type I FAP can involve the CNS as well as the peripheral somatic and autonomic nerves. In the CNS of patients with type I FAP, the leptomeningeal vessels are the principal site of amyloid deposition, as shown in two recent pathologic studies of 18 patients with systemic amyloidosis, 12 of whom had type I FAP (6, 7). In these studies, amyloid CNS deposits were found in the leptomeningeal vessels and the pia-arachnoid membranes, predominantly involving the arteries and arterioles of the subarachnoid cerebral space.

Our case of type I FAP establishes the corresponding MR findings of diffuse, linear, enhancing leptomeningeal amyloid deposition along the pial surfaces of the spinal cord and brain stem, arachnoid nodular enhancement dorsal to the conus medullaris, linear nerve root enhancement of the cauda equina, and sacral intrathecal and epidural amyloid deposits. The smooth, scalloped bone erosion of the sacral canal could have resulted from a prior Tarlov cyst that was

subsequently compressed by epidural amyloid or, alternatively, the bone could have been eroded by the amyloid deposit itself. This bony scalloping differs from the direct bony involvement with amyloid that is most commonly associated with multiple myeloma or other plasma cell dyscrasia (8). Primary solitary amyloidoma of bone is a rare localized amyloid deposit without plasma cell dyscrasia or abnormal serum proteins, reported in seven cases of vertebral body lesions, six thoracic and one cervical. This lesion is usually osteolytic, but it was calcified in one case and displayed paraspinal extension that caused cord compression in two cases (9, 10).

Enhancement of amyloid deposits has been previously reported in blood vessel walls in a primary amyloidoma of the brain (11). Potential explanations for contrast enhancement in leptomeningeal pial and arachnoid amyloid may include an inflammatory reaction to the amyloid deposits with extracellular enhancement outside the blood-cord or blood-brain barrier, lack of wash-out of contrast due to altered vascularity or vascular stasis within the abnormal leptomeningeal vessels or tissues, or enhancement of edematous tissue. Contrast agent itself has no protein-binding capacity.

Conclusion

This case illustrates a rare cause of leptomeningeal MR enhancement due to amyloid deposition along the pia of the spinal cord, in the spinal and cerebral subarachnoid space, and in the sacral intradural and epidural spaces.

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