CT and MR Imaging of Progressive Dural Involvement by Nephrogenic Systemic Fibrosis

Nephrogenic systemic fibrosis (NSF), previously referred to as nephrogenic fibrosing dermopathy, is a relatively new, rare systemic condition first described in the literature in 2000. NSF occurs only in people with renal disease, with more than 215 documented cases in the NSF Registry at Yale University. All but a few of the known cases have been associated with intravenous administration of gadolinium-based contrast material. Manifestations of the disease are primarily cutaneous, but multiorgan system involvement has been described. Dural involvement has been described at autopsy; however, the depiction of the progressive imaging changes has not been documented. We report a patient with progressive dural calcification, thickening, and enhancement as a manifestation of NSF.

Case Report
A 52-year-old woman with a medical history of end stage renal disease secondary to adult polycystic kidney disease required hemodialysis starting in 2004. She had 8 gadolinium-enhanced MR imaging studies of the brain and abdomen between September 2004 and November 2006, all performed with gadodiamide. The diagnosis of NSF is made by a combination of clinical, laboratory, and histopathologic findings. Prognosis is variable, but NSF may be fatal. Treatment leading to improved renal function may lead to relief of symptoms and may halt progression.

Nephrogenic systemic fibrosis was first reported in 2006. Both spinal and intracranial dural involvement have been described. Microscopic examination demonstrates fibrosis with a spindle-cell proliferation, areas of calcification, collections of CD68-positive mononuclear cells, and occasional multinucleated giant cells. The histologic changes in the dura mater of our patient were similar to those previously reported. 

The differential diagnosis for dural thickening and enhancement is extensive and includes sarcoidosis, tuberculosis, Wegener granulomatosis, intracranial hypotension, lymphoma, and metastatic disease. The differential diagnosis for dural calcifications includes physiologic calcifications, previous hemorrhage, previous infection, pseudoxanthoma elasti-
cum, hyperparathyroidism, basal cell nevus syndrome, and idiopathic calcification.\textsuperscript{13}

Virtually all patients with endstage renal disease have resultant hyperparathyroidism, and “metastatic calcification” may develop, with calcium deposition in the soft tissues and dura mater. However, dural thickening and enhancement are not typically seen in these patients. The patient in this case report had serum calcium values within normal limits and did not have other foci of soft tissue calcification. Her serum phosphorus levels ranged from low to elevated but were predominantly within normal limits. Although hyperparathyroidism cannot be excluded as the cause of the dural calcification because of the dural thickening and enhancement, the lack of other evidence of metastatic calcification, and her normal serum calcium levels, it is believed that the dural disease most likely represents a manifestation of NSF.

We suggest that dural calcification, thickening, and enhancement in patients with renal disease and a history of exposure to gadolinium may represent an early manifestation of NSF. Neuroradiologists must recognize these findings as a part of the spectrum of disease in NSF and may be the first to suggest the diagnosis if these radiologic findings precede cutaneous manifestations.
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