Spinal Epidural Lipomatosis in a Human Immunodeficiency Virus–Positive Patient Receiving Steroids and Protease Inhibitor Therapy

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We describe a patient who became cushingoid as a result of receiving steroid therapy for thrombocytopenia purpura and who then developed spinal epidural lipomatosis 4 months after he started receiving ritonavir as part of his therapy for human immunodeficiency virus infection. We believe that ritonavir may have contributed to the development of epidural lipomatosis and that clinicians should be aware of this possible association.

Spinal epidural lipomatosis is a very rare condition in which excess fat accumulates in the epidural space and can result in cord and/or nerve root compression and symptoms of myelopathy. The majority of ~60 cases reported to date have resulted from exogenous Cushing's syndrome or obesity [1–3]. We report a case of spinal epidural lipomatosis that developed after treatment with a protease inhibitor was initiated in a patient who had been receiving long-term steroid therapy.

The patient was a 38-year-old HIV-positive man who had a CD4 cell count of 280 cells/mm³ and an undetectable HIV RNA level (<400 copies/mL), and who, during the course of ~2 months, had experienced increasing leg weakness, unsteady gait, and urinary incontinence. Three years previously, he had been given a diagnosis of thrombotic thrombocytopenia purpura (TTP) and HIV infection; management with plasmapheresis and long-term steroid therapy (dexamethasone, 6 mg/day, which was slowly decreased to 1–2 mg/day by the time neurologic symptoms developed) resulted in good control of the TTP. Treatment with lamivudine and zidovudine was begun 3 months after the HIV infection was diagnosed. Approximately 1 year after initiation of steroid treatment, the patient developed a cushingoid appearance that included enlarged supraclavicular fat pads and diabetes mellitus that required insulin treatment. Ritonavir, which was the first protease inhibitor given to this patient, was added to treatment with zidovudine and lamivudine 4 months before symptoms of myelopathy developed.

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Figure 1. Sagittal, T1-weighted image of the spine. Increased signal intensity is seen in the epidural space along the posterior aspect of the thoracic cord (bottom arrow). Also indicated is nuchal accumulation of fat (known as a “buffalo hump”, top arrow).
MRI revealed epidural lipomatosis at the L5-S1 level, effacing the thecal sac, and also at T1-12, compressing and displacing the thoracic spinal cord with near obliteration of the subarachnoid space (figure 1). Pronounced mediastinal and peri-aortic lipomatosis was also noted. The cholesterol level, which had been normal (193 mg/dL) 10 months before initiation of protease inhibitor therapy, was elevated to 347 mg/dL, and the triglyceride level was elevated to 464 mg/dL (reference range, 35 mg/dL to 260 mg/dL) after 1 year of administration of ritonavir. It is unfortunate that lipid levels were not measured at intervals between these 2 times.

Five months after the onset of neurologic symptoms, the patient underwent thoracic laminectomy with cord decompression, and he experienced considerable, but not complete, neurologic recovery. Antiretroviral therapy administered at this time consisted of ritonavir, saquinavir, and lamivudine.

HIV lipodystrophy associated with protease inhibitor therapy may be characterized by an accumulation of fat in the abdominal viscera, supraclavicular fossae, women’s breasts, and dorsocervical tissue [4–7]. These changes are also characteristic of the fatty redistribution seen in patients with exogenous or endogenous Cushing’s syndrome. Whether HIV lipodystrophy may also cause epidural lipomatosis, as is described with Cushing’s syndrome, is unknown and has not been reported. Our patient was cushingoid as a result of receiving long-term exogenous steroid therapy, but he did not have symptoms of myelopathy until 4 months after initiation of ritonavir therapy. We believe that it is possible that an effect that was additive to that of the steroids contributed to our patient’s epidural lipomatosis and that clinicians should be aware of the possible association.

References