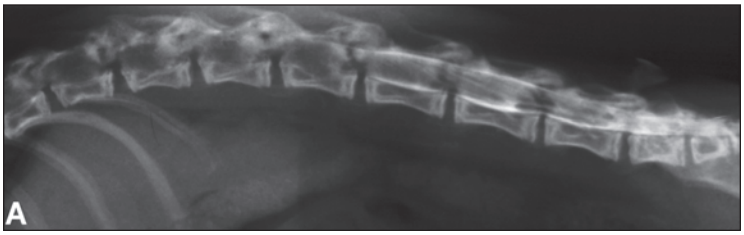
In cooperation with

What Is Your Diagnosis?



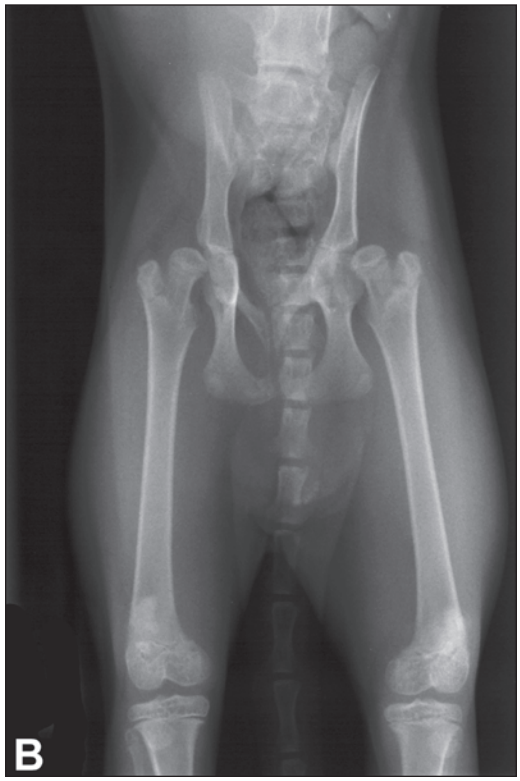


Figure 1—Lateral myelographic view of the lumbar portion of the vertebral column (A) and ventrodorsal radiographic view of the pelvis (B) of a 3-month-old cat evaluated for progressive hind limb paresis and ataxia of approximately 1 month's duration.

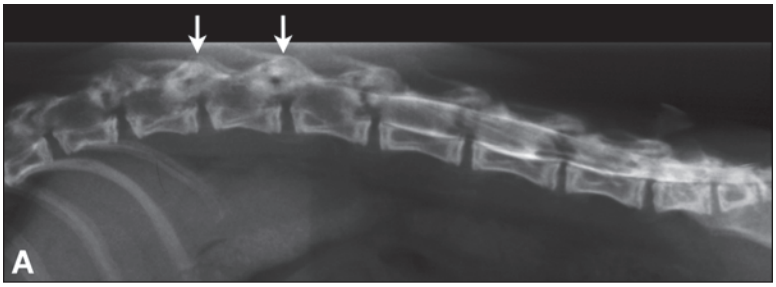
History

A 3-month-old sexually intact female Siamese cross was referred for evaluation of progressive weakness in the hind limbs of approximately 1 month's duration. History prior to referral included failure to thrive and small size as compared with littermates. Test results for FeLV and FIV infection were negative, and the cat had been vaccinated against FeLV feline viral rhinotracheitis virus, calicivirus, and panleukopenia virus. The cat had not been vaccinated against rabies virus at the time of evaluation.

Physical examination findings included a broad and flattened face, bilateral corneal opacification, poor body condition, stunted growth, signs of pain on extension of the hip joints, and generalized laxity of ligaments and muscles of both hind limbs. Abnormal findings detected on neurologic examination included ataxia, conscious proprioceptive deficits, and hyperreflexive to normal withdrawal and patellar reflexes in both hind limbs. Hyperesthesia was detected on palpation of the lumbar portion of the vertebral column. Survey radiography of the thoracic portion of the vertebral column revealed degenerative joint disease. Radiographs of the pelvis and lumbar portion of the vertebral column after a myelogram was performed were obtained (**Figure 1**).

Determine whether additional imaging studies are required, or make your diagnosis from Figure 1—then turn the page ⏵

This report was submitted by Arathi Vinayak, DVM; Alan R. Cross, DVM, DACVS; and Susan Newell, DVM, MS, DACVR; from the Department of Small Animal Clinical Sciences (Vinayak), College of Veterinary Medicine, University of Tennessee, Knoxville, TN 37996; and Georgia Veterinary Specialists (Cross, Newell), Atlanta, GA 30328. Dr. Vinayak's present address is Department of Small Animal Medicine and Surgery, College of Veterinary Medicine, Texas A&M University, College Station, TX 77843.
The authors thank Dr. George Henry for assistance.
Address correspondence to Dr. Vinayak.



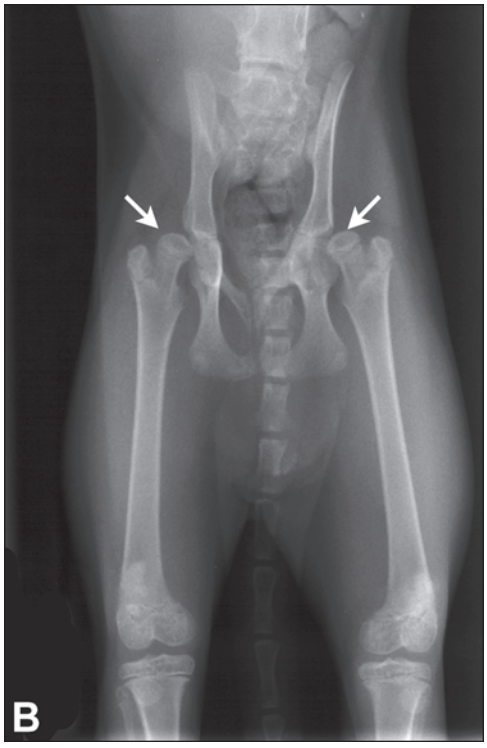


Figure 2—Same views as Figure 1. Notice degenerative joint disease and bony proliferation of articular facets (arrows) of the lumbar portion of the vertebral column with subsequent compression of the spinal cord (A) and subluxation to near luxation of the hip joints with flattened femoral heads and shallow acetabula of the pelvis (B; arrows).

Radiographic Findings and Interpretation

Misshapen vertebral bodies, degenerative joint disease, and remodeling with new bone formation in the articular facets along the entire lumbar portion of the vertebral column are evident. Bilateral severe subluxation to near luxation of the hip joints with shallow acetabula and flattened femoral heads indicate remodeling (**Figure 2**). Radiographic findings were consistent with a diagnosis of **mucopolysaccharidosis (MPS)** type VI.

Comments

Mucopolysaccharidosis type VI is an inherited autosomal recessive lysosomal storage disease recognized in cats of Siamese ancestry.[1] Mucopolysac-charidoses are a family of 11 distinct lysosomal enzyme deficiencies resulting in the accumulation of partially degraded **glycosaminoglycans (GAGs)** within the patient's lysosomes, and MPS type VI is characterized by a deficiency in arylsulfatase B activity causing lack of degradation and subsequent accumulation of dermatan sulfate containing GAGs. This product is stored in tissues of many organs and is excreted in urine. Diseases of similar radiographic appearance include congenital hypothyroidism, epiphyseal dysplasia, hypervitaminosis A, and hyperparathyroidism. Therefore, radiographic findings in conjunction with clinical signs and Siamese ancestry are suggestive but not definitive for MPS type VI. A definitive diagnosis is made by detection of excess dermatan sulfate in urine via a toluidine blue spot test, metachromatic granules in the cytoplasm of 90% to 100% of neutrophils in toluidine blue-stained blood smears, and marked increase in activity of arylsulfatase B in peripheral leukocytes.[2] A positive toluidine spot test result for dermatan sulfate in urine and detection of metachromatic granules in neutrophils were confirmed in the cat of this report.

Clinical features in this disease usually become apparent by 8 weeks of age and include corneal opacification, facial dysmorphism, dwarfism, osteopenia, bilateral coxofemoral subluxation, severe degenerative joint disease, and spinal cord compression with resultant hind limb paresis.[1,2] Neurologic abnormalities in 25% of cats with MPS VI are not caused by abnormalities in the spinal cord itself but rather are caused by proliferation of bony tissue causing compression of the spinal cord.[2] Computed tomography of the lumbar portion of the vertebral column of the cat in this report revealed bony proliferation of the articular facets and subsequent spinal cord compression. Abnormalities detected on radiographs of the vertebral column include fusion of the cervical, thoracic, and lumbar vertebrae and bony proliferation detected mostly in the thoracolumbar portion of the vertebral column, which causes upper motor neuron hind limb paresis in affected cats. However, radiographic changes cannot be used to predict severity and progression of clinical signs. A decompressive dorsal laminectomy can be attempted to relieve spinal cord compression; however, in 1 study,[2] clinical signs progressed from paraparesis prior to surgery to complete hind limb paralysis within 1 week of surgery. Results of a study[3] on recombinant human *N*-acetylgalactosamine-4-sulfatase enzyme supplementation for the treatment of MPS type VI indicate greater improvement in physical, neurologic, and skeletal condition in cats experimentally treated at birth, compared with cats treated at a later age. Clinically, however, MPS type VI is diagnosed in most cats after worsening in clinical signs and development of considerable lesions.

1. Konde LJ, Thrall MA, Gasper P, et al. Radiographically visualized skeletal changes associated with mucopolysaccharidosis VI in cats. *Vet Clin North Am Small Anim Pract* 2000;30:281–302.

2. Haskins ME, Bingel SA, Northington JW, et al. Spinal cord compression and hindlimb paresis in cats with mucopolysaccharidosis VI. *J Am Vet Med Assoc* 1983;182:983–985.

3. Auclair D, Hopwood JJ, Brooks DA, et al. Replacement therapy in mucopolysaccharidosis type VI: advantages of early onset of therapy. *Mol Genet Metab* 2003;78:163–174.