HEMODECTOMAS or paragangliomas are tumors that arise from the autonomic nervous system–associated paraganglia. Paraganglia are groups of cytochemically and morphologically similar cells derived from the neural crest. Most chemodectomas arise from the carotid bodies and glomus jugulare but other sites include aortic bodies, adrenal medullae, and retroperitoneum. They are usually considered to be benign lesions but up to 10% can metastasize to lungs, liver, or bones.11 There have been several reported cases of vertebral metastases.1,3,5,13,14 Most patients have been treated with a combination of surgical decompression and adjuvant radiotherapy. The use of radiolabeled octreotide scintigraphy to demonstrate somatostatin receptors in chemodectomas is of proven value;9,12 however, the therapeutic use of octreotide to treat vertebral metastatic chemodectoma is poorly documented.

Case Report

Examination. This 32-year-old woman presented with severe thoracic back pain and paraplegia following a fall 2 weeks previously. In addition she described a 2-year history of progressive back pain and difficulty walking. She had undergone excision of a left carotid body tumor 14 years earlier. Clinically, she had a severe spastic paraparesis (Grade 1/5 power proximally and 0/5 power distally), with sensory loss below the T-10 level.

Neuroimaging and Histological Findings. Magnetic resonance imaging of spine demonstrated metastatic infiltration of the T1–2 and T-9 vertebral bodies, with cord compression being most severe at T-9. Smaller targetlike lesions were also demonstrated throughout the spine (Fig. 1).

A computerized tomography–guided biopsy of the T-9 vertebra was performed. Histological examination revealed nests of polygonal tumor cells with rounded nuclei and granular cytoplasm consistent with metastatic chemodectoma. Immunohistochemical analysis showed strong reactivity with synaptophysin, chromogranin A, neuron-specific enolase, and neurofilament protein. Octreotide-enhanced scintigraphy revealed increased uptake and the presence of somatostatin receptors at T1–2, T-9, and the sacrum (Fig. 2).

Spinal angiography, which was performed as a precuror to potential surgery, demonstrated the vertebral metastases to be extremely vascular (Fig. 3). In addition, multiple small lesions were seen throughout the cervicothoracic spine, which had not been revealed by the MR imaging studies.

Treatment and Posttreatment Course. In view of the vascularity of the lesions, her widespread disease, established severe paraparesis, and the patient’s wishes surgery was not performed.

The patient underwent external-beam radiotherapy (30 Gy in 10 fractions) to her cervicothoracic, lower thoracic, and lumbosacral spine over a 12-day period. Following treatment, she had Grade 2/5 power proximally and Grade 0/5 power distally. Sensory loss remained unchanged. Her

Abbreviation used in this paper: MR = magnetic resonance.
Thoracic pain significantly improved. Subcutaneous injections of octreotide (100 μg) were administered three times daily. This dose was reduced to 100 μg two times daily because the patient experienced persistent gastrointestinal side effects such as abdominal pain, nausea, and diarrhea. She continued on this regimen for 18 months before finally stopping treatment of her own accord because of intermittent nausea. Three years later, her lower limbs have improved to Grade 4/5 power proximally and distally, her back pain is minimal, and she does not have any sensory deficit and can walk with assistance. Serial MR imaging over the 3-year period has not demonstrated evidence of any disease progression.

Discussion
The natural history of metastatic chemodectomas is unpredictable. Clinical manifestations of metastatic disease may take up to 17 years to present after initial resection, despite the tumor appearing histologically benign. Most metastatic tumors are slow growing but aggressive behavior has been described, with patient survival ranging from 6 months to 9 years. Imaging modalities for vertebral metastatic disease include plain radiography, computerized tomography, and MR imaging; however, these primarily depict anatomy and can miss multiple tumors and metastases at sites where disease is not clinically suspected. Iodine-123 metaiodobenzylguanidine (an analog of noradrenaline) scintigraphy has been used with a sensitivity of 40 to 54% in patients with chemodectomas. In this case report we emphasize the importance of two additional modalities: octreotide scintigraphy and spinal angiography. The presence of somatostatin receptors in chemodectomas has led to the application of somatostatin receptor analog scintigraphy in conjunction with either 111In pentreotide or 123I tyrosine octreotide; the sensitivity has ranged from 86 to 100%. We used octreotide scintigraphy to confirm receptor positivity prior to administering therapeutic oc-

![Fig. 1. Sagittal T2-weighted images of cervicothoracic (upper) and lumbar (lower) spine revealing two radiological patterns of disease: diffuse reduced signal at T1–2 and T-9 and target lesions of round areas of reduced signal surrounded by high signal within other vertebral bodies.](image1)

![Fig. 2. Iodine-123 tyrosine octreotide scintigraphy revealing increased uptake in lower cervical and lower thoracic spine and sacrum.](image2)
treotide. Spinal angiography, in our case performed for the purposes of planning surgery, suggested that the metastatic disease was more extensive than that demonstrated on MR imaging. It also revealed the extent of vascularity of the vertebral metastases and was important in our decision that the risks involved with surgery outweighed the benefits. These spinal angiographic appearances have not been described before.

Treatment for metastatic chemodectomas has generally been external-beam radiotherapy with or without surgical decompression.\(^1,3,11\) Radiotherapy is effective in achieving clinical control for both primary disease and metastases.\(^1,3,11,14\) Surgical decompression may involve corpectomy and stabilization.\(^1,3,3,13,14\) The response to chemotherapy is, in general, poor.\(^1,18\) Radionuclide therapy with high doses of \(^123\)I metaiodobenzylguanidine has been successfully used for temporary palliative treatment of progressive disease.\(^1,12\)

Octreotide, a somatostatin analog, has been used for the treatment of a variety of neuroendocrine tumors including pituitary adenomas, carcinoids, and gastrointestinal endocrine tumors.\(^9,18\) Its main therapeutic action in such tumors is to inhibit hormonal hypersecretion and to control symptoms. The presence or absence of somatostatin receptors and receptor density can be used to predict the success of octreotide therapy;\(^10\) however, experience with octreotide for treatment of chemodectomas is limited. Muros, et al.,\(^12\) reported a decrease in tumor size in two patients with glomus tumors after treatment with octreotide (200 pg administered subcutaneously three times daily for the first 3 days and then 500 pg three times daily). Kau, et al.,\(^6\) showed a growth inhibitory effect with octreotide therapy for inoperable chemodectomas of the head and neck. Tenenbaum, et al.,\(^16\) describe a transient response in a patient with bone metastases whose tumor recurred following other treatment modalities, including external-beam radiotherapy and chemotherapy. In the case we describe, we believe that the combination of external-beam radiotherapy and octreotide administration has resulted in improvement of neurological function and radiological stabilization of disease. Novel applications include radionuclide therapy with high doses of \(^111\)In- or \(^90\)Y-labeled octreotide analogs. Slooter, et al.,\(^15\) have used this technique for a variety of neuroendocrine tumors with improvement in outcome. Two of their patients had glomus tumors: one remained stable and the other showed a reduction in tumor size. Side effects were minor except for transient decline in platelet count and white cell count.

Conclusions

We suggest that if somatostatin receptors can be demonstrated by radionuclide imaging, adjuvant therapy for vertebral metastatic chemodectoma should include long-term octreotide administration. In addition, spinal angiography is an invaluable adjunct to conventional MR imaging scanning in such cases.

References

Vertebral metastatic chemodectoma


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