Visual Vignette

Submitted by
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Case Presentation: A 29-year-old woman presented with history of low back pain for more than 6 months' duration with radiation to both lower limbs. She underwent a computed tomography scan of the lower back with bone reconstruction views. An irregular lytic lesion involving the first and second segments of the sacrum was noted with destruction of the neural foramina and involvement of the pedicle of the L5 vertebrae (Fig. 1 and 2). An initial differential diagnosis of a giant-cell tumor or a metastasis from an unknown primary tumor was considered. The patient was scheduled for a computed tomography–guided biopsy. While the premedication for the procedure was being administered, the patient was noted to have high blood pressure measurements, so the procedure was abandoned. On subsequent questioning, the patient divulged additional information: she had surgery to treat an extraadrenal pheochromocytoma (located in the organ of Zuckerkandl at the bifurcation of the aorta) 12 years previously in our hospital when she initially presented with hypertension and a palpable abdominal mass. After 3 years of follow-up, she did not return for a check-up. She had been asymptomatic and normotensive over the next 9 years until the present episode of lower back pain. After her referral to the endocrinology department, I 131 *m*-iodobenzylguanidine (MIBG) scintigraphy was performed—the 72-hour post-MIBG images are shown (Fig. 3). Vanillylmandelic acid excretion was above the reference range on 3 different 24-hour urine collections. **What is the diagnosis?**

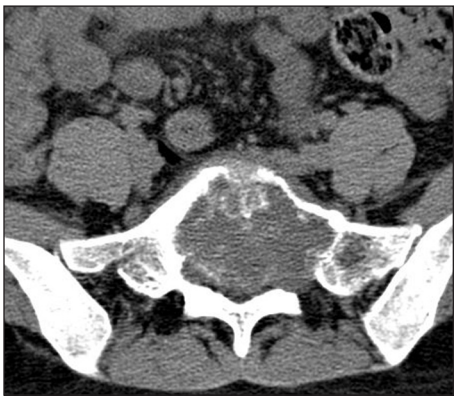

Fig. 1

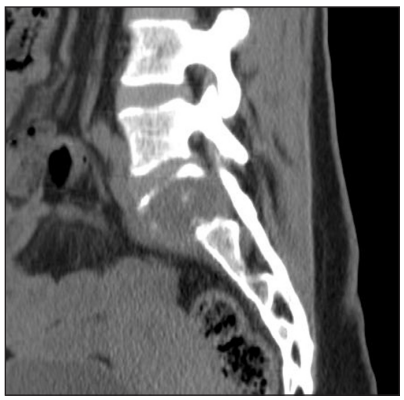

Fig. 2

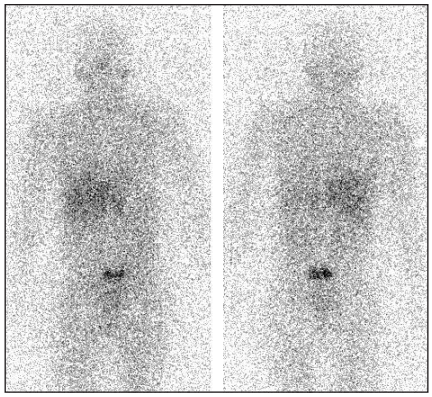

Fig. 3

Answer: Solitary sacral metastasis from a malignant paraganglioma. The MIBG scintigraphy shows intense uptake in the same region as the lytic lesion seen on computed tomography. There was no other additional focus of tracer uptake. Malignancy occurs in 2.6% to 26% of individuals with pheochromocytoma. Paragangliomas of the retroperitoneum have much higher reported rates of malignancy. The common sites of metastasis are bone, liver, and lungs (1). Malignant pheochromocytoma may recur early or late in the clinical course; the latest reported occurrence is more than 20 years after initial surgery (2). Therapy for malignant pheochromocytoma and paraganglioma includes local excision, I 131 MIBG, and external beam radiotherapy (3). In patients with rapidly progressing malignancy, chemotherapy should be used as first-line treatment. Our patient was treated with an ablative dose of 105 mCi of I 131 MIBG followed by external beam radiotherapy for local pain and α-adrenergic receptor blockade.

REFERENCES

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