

Chemodectoma Presenting with Dorsal Vertebral Metastasis

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Abstract: Tumors of the paraganglionic system include pheochromocytomas, carotid body tumors (chemodectoma), jugulotympanic paragangliomas (glomus jugulare tumors), and the rare aorticopulmonary and aortic sympathetic paragangliomas. Paragangliomas are generally well-differentiated tumors, with only a minority (7% to 10%) behaving in a malignant fashion by causing distant spread. The case report of a 37-year-old male patient, with a medical history of glomus jugulare tumor resected 1 year ago, attending with a spinal column metastasis is presented. Surgical resection of the metastases, combined with radiotherapy, was performed on the patient. Paragangliomas have the potential to be malignant and although rare, the spinal column is one of the targets of spread.

Key Words: glomus jugulare tumor, paraganglioma, spinal metastasis

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Paragangliomas are rare tumors that arise from the autonomic nervous system associated with paraganglia. They are mostly benign tumors but occasionally metastasize. Infrequently, they metastasize to the spinal cord. Isolated metastatic involvement of the vertebral body is extremely rare. We present a case of metastatic nonchromaffin paraganglioma (glomus jugulare tumor) with 3 levels of dorsal vertebral column involvement.^{1,2}

CASE REPORT

In 2000, a 37-year-old laborer presented with a palpable, painless, and enlarging mass of the right sternocleidomastoid muscle. Investigation with a magnetic resonance imaging scan demonstrated a glomus jugulare tumor, which was resected in 2003 after an embolization procedure. The pathologic study revealed that it was a nonchromaffin paraganglioma, namely glomus caroticum tumor, and the patient was believed to be cured.

In October 2004, the patient was admitted to our oncology department with severe dorsal pain. He complained

of a disturbance of gait and lower extremity weakness, which started on the right side and progressed to the left in 2 weeks. At admission, his physical examination revealed a marked spasticity of the lower limbs with hyperreflexia, bilateral ankle clonus, and Babinski's sign. Sensation to pinprick was absent below the level of T₁₂ and sphincter function was intact. Dorsal roentgenograms showed a burst fracture of T₉ and a dorsal magnetic resonance imaging revealed diffuse spinal cord edema at the 6th, 9th, and 10th vertebral levels, characterized by hyperintensity on T₂-weighted images (Figs. 1A, B).

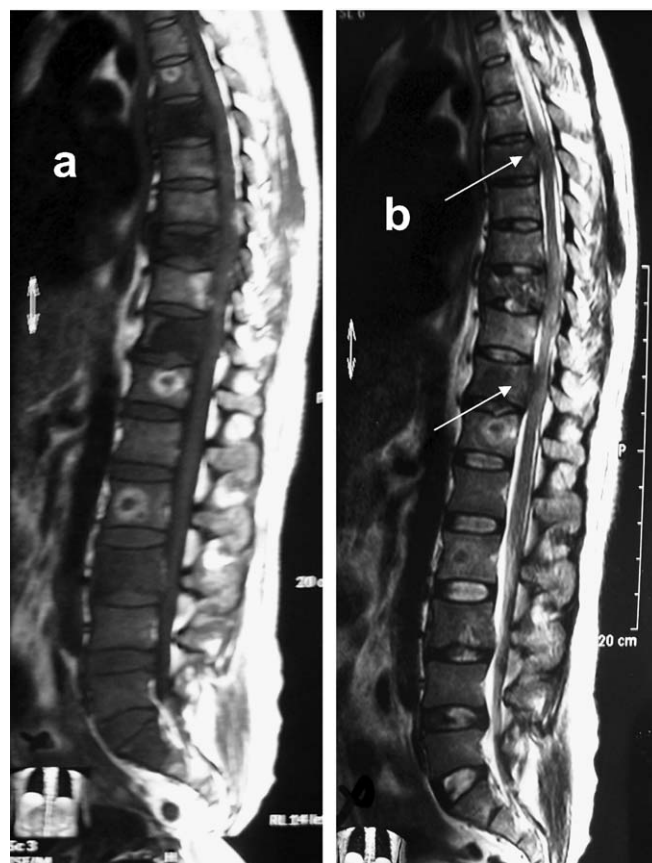


FIGURE 1. T1 (A) and T2-weighted (B) magnetic resonance images of the lumbar and thoracic spine demonstrating multiple metastatic foci in the dorsal and lumbar vertebrae. There is also 50% height loss of the ninth vertebral body and prominent spinal cord compression. Moreover, spinal cord compression is noted at the 6th and 11th vertebral body levels (arrows).

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The patient was operated via a posterior approach, and T₉ and T₁₀ laminectomy was performed. The posterior elements were invaded by the highly vascular tumor, which seemed reddish brown, soft, and friable, and bled easily. All the accessible tumor tissue was resected and the dural sac was observed to reexpand. The procedure lasted 120 minutes, with a blood loss of 900 mL and without any preoperative complications. At the postoperative period, his neurologic signs did not resolve.

The biopsy specimen was submitted for pathologic analysis. The pathologic evaluation demonstrated that the trabecular bone was invaded by tumor cells with accompanying vascular components. The tumor cell islets were surrounded by hyalinized cellular nests and were mucine negative. The immunohistochemical analysis demonstrated that the tumor cells showed a positive reaction to the CD31 study. The pathologic study verified that these lesions were the metastasis of chemodectoma.

Then, the patient received external beam radiotherapy (30 Gy over 10 fractions) to the spinal column between the 5th and 12th dorsal vertebrae. No adjuvant chemotherapy was administered and he was discharged.

DISCUSSION

Tumors of the paraganglionic system include pheochromocytomas, carotid body tumors (chemodectoma), jugulotympanic paragangliomas (glomus jugulare tumors), and the rare aorticopulmonary and aortic sympathetic paragangliomas.

The tumors are generally well-differentiated, with only a minority (7% to 10%) behaving in a malignant fashion, and giving rise to distant metastasis. They are designated as functional or nonfunctional according to whether they secrete catecholamines. Spinal involvement is infrequent, and spinal paragangliomas may be primary or metastatic.^{1,3,4}

It is generally accepted that paragangliomas are most often benign, but occasionally metastasize.^{2,5-8}

Primary spinal canal paragangliomas are rare, with approximately 80 cases reported in the literature. The vast majority of these tumors are intradural, in the cauda equina; most of them are ganglionic paragangliomas. It has been suggested that primary spinal paragangliomas arise from the sympathetic chain.^{1,9} The overall incidence of extra-adrenal paragangliomas is unknown, and in an autopsy study of 19,610 patients, it was found to be 0.01%.⁸

The incidence of metastasis of paragangliomas ranges between 2% and 50% and malignancy for carotid body tumors is 2.5% and 12% for familial and sporadic groups, respectively. There is little correlation between histologic appearance and malignancy potential. Benign-appearing tumors may behave in an aggressive fashion. Thus, the metastatic potential of paragangliomas may be related to their site of origin. One large series reported an incidence of 13% metastases from all sites but up to 31% when only considering extra-adrenal sites. The very rare extra-adrenal retroperitoneal paragangliomas seem to behave even more aggressively with metastatic rates of up to 50%. Many writers have reported that these tumors

have distant sites for metastasis such as the lymph nodes, lungs, liver, bones, and the spine.^{2,10}

The nature of the skeletal metastasis has been reported as osteolytic or osteoblastic. It is interesting to note that among all extra-adrenal chemodectomas, carotid body tumors have the highest propensity for metastatic spread to the vertebral column.¹

Although it is generally believed that 10% of these tumors metastasize, 1 review described a 50% rate of metastasis with regard to extra-adrenal retroperitoneal paragangliomas.¹¹ In most cases, paragangliomas metastasize through direct spread or by invasion of local lymph nodes and are frequently found in the lungs, mediastinum, and skeleton. Spinal involvement is uncommon and is generally intradural at the level of the cauda equina. Intradural thoracic and cervical paragangliomas are less common. Filum terminale involvement is rare. Several writers have reported vertebral body metastases. Epidural compression of the spinal cord by paraganglioma metastasis in the thoracic spine has been reported in a few articles.^{1,12}

Paragangliomas often exhibit a prolonged time interval to the development of metastasis or recurrence. Recurrences and/or distant metastasis have been recorded to occur 0 to 20 years after the initial diagnosis.

The median time for recurrence or metastasis after primary resection is approximately 6 years.² In our patient, distant metastasis occurred 4 years after the diagnosis of the glomus caroticum tumor.

Radiation therapy is reported to be effective in the treatment of paragangliomas and radiation therapy in conjunction with surgery has been found to be more effective than surgery alone in the treatment of large, incompletely resected tumors, because these are radiosensitive tumors and irradiation at a stage of lesser tumor burden should be more effective.² No consistent chemotherapy has been given for metastatic chemodectoma. Results with chemotherapy, including 5-fluorouracil and cisplatin, are disappointing. In general, surgical resection of isolated metastasis has proven to be beneficial.^{13,14}

The treatment of choice for metastatic paragangliomas with spinal cord compression is usually decompressive surgery and external beam radiotherapy.

Paragangliomas are usually considered benign lesions; however, the findings of malignancy in approximately 6.5% of these lesions and that of local tumor recurrence in 12% even after 30 years mandates that these tumors be followed up and monitored over the long term.¹²

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