CASE REPORT

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Malignant Retroperitoneal Paraganglioma with T11 Metastasis and Compression of the Spinal Cord

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ABSTRACT

A 38 years old male worker presented with an advanced left side retroperitoneal mass, with metastatic involvement of 11th Thoracic Vertebra (T11) and resultant paraparesia and paresthesia. The histopathologic examination and immune-histochemical analysis confirmed the diagnosis of paraganglioma. Decompression laminectomy and fixation of involved vertebra resulted to considerable symptomatic relief. Due to inability of resection at presentation, chemotherapy with palliative intent performed, and resulted to mild response. Palliative debulking performed after chemotherapy and resulted to better quality of patient's life during next 8 months. Remaining mass at pelvic area increased in size and patient received (131) I-MIBG therapy, 8 months post-operatively. A second debulking surgery, as well as other palliative measures will be considered for future.

Keywords: ***Retroperitoneal paraganglioma, Metastasis, Spinal cord, (131) I-MIBG therapy.***

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Background

Retroperitoneal paragangliomas, derived from the extra-adrenal paraganglia of the autonomic nervous system, a widely dispersed collection of specialized neural crest cells,[1] are rare, usually occur in adults, and most are benign in nature. In children 15 to 26 percent of paraganlionomas are extra adrenal. The most aggressive paraganglioma is intra-abdominal extra-adrenal one.[2] Imaging studies, including computed tomography (CT) and magnetic resonance imaging (MRI) are necessary for diagnosis. The gold standard for accurate diagnosis is histopathologic methods. Metastases to vertebrae, with or without extradural extension and destruction or compression of the spinal cord are rare.[3-4] Surgical resection of primary tumors and metastatic lesions, remains the mainstay of treatment. Response to radiation and/or chemotherapy is poor.[5]

Case Report

A 38-year-old male with a recent history of deep vein thrombophlebitis, presented with weight loss, vague abdominal pain and progressive neurological deficit of lower extremities of about 8 months duration. He had no history of hypertension and there was no symptom related to excess catecholamine release. Examination revealed a huge abdominal mass and bilateral paraparesis and lower extremities edema. Otherwise physical examination was unremarkable.

Abdominal Ultrasonography revealed a large left retroperitoneal hypoechoic solid mass, with 90 × 190 mm dimension. Computed tomography showed huge, ill-defined, hypervascular, left retroperitoneal mass of heterogeneous density, extending from pancreatic level to pelvic cavity (**figure 1**). According to the clinical presentation and CT findings, advanced retroperitoneal mass, probably paraganglioma was suspected. 131I-MIBG SPECT showed inhomogeneous tracer accumulation in the huge retroperitoneal mass (**Figure 2a**). Metastatic involvement of T11 and ischium was confirmed at bone scintigraphy (**Figure 2b**). Surgical intervention at an emergency basis performed, during which Decompression laminectomy, internal fixation and biopsy of retroperitoneal mass and metastatic tumor performed (**Figure 3**).

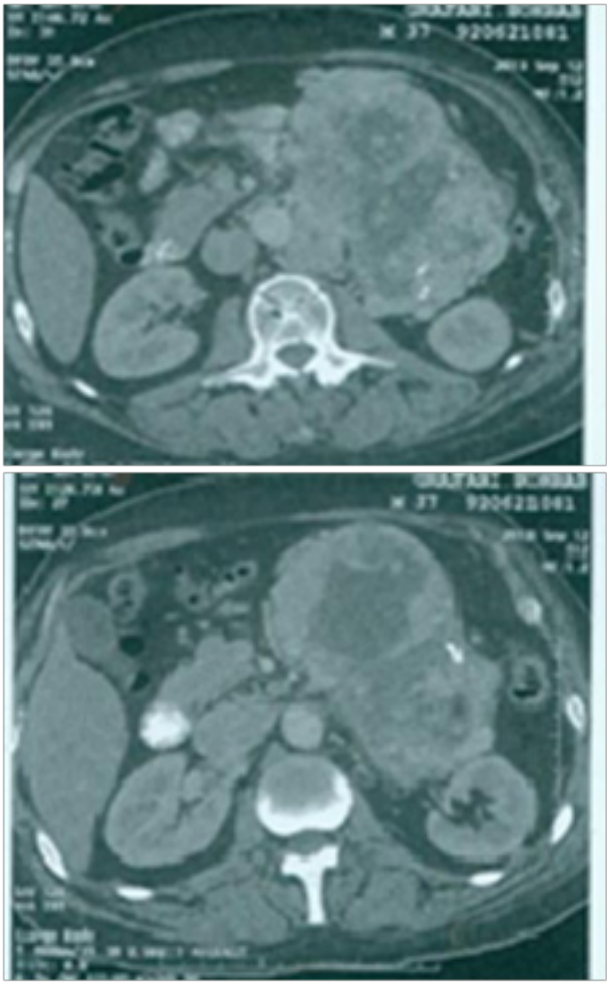

Figure 1: Huge ill-defined heterogeneous enhancing hypervascular retroperitoneal mass, containing calcifications

The neurological outcome of surgery was dramatic, and patient returns to nearly normal physical activity. Post-operative image is shown on **Figure 4**.

The case was discussed at weekly Cancer Institute general tumor board, and due to unresectability of original tumor, chemotherapy with palliative intent suggested. A combination chemotherapy (cyclophosphamide 750 mg/m 2 and vincristine 2 mg/m 2, and dacarbazine (DTIC) 600 mg/day, for 2 days of a 28-day cycle) prescribed. After 4 courses the patient's pain improved, but he suffered considerable nausea. Retroperitoneal mass showed no considerable change. 6 weeks after chemotherapy, the patient underwent a palliative surgical resection, during which considerable debulking of the retroperitoneal mass performed, and resulted a better quality

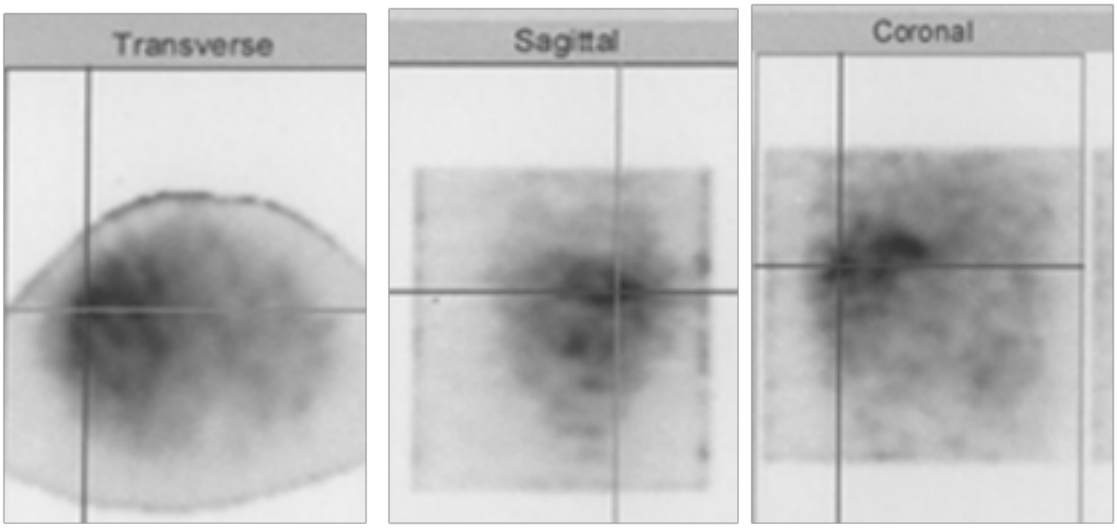

Figure 2a: Inhomogeneous localization of 131I-MIBG in the retroperitoneal mass

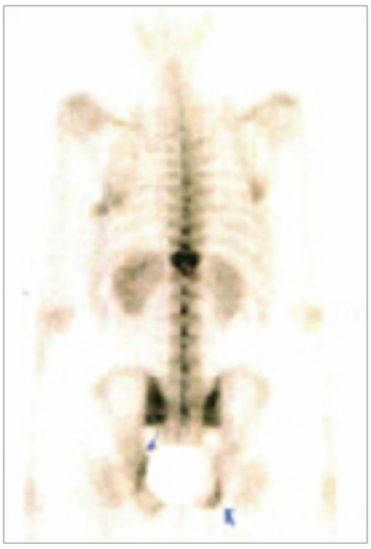

Figure 2b: Metastatic involvement of T11, extending to spinal cord canal

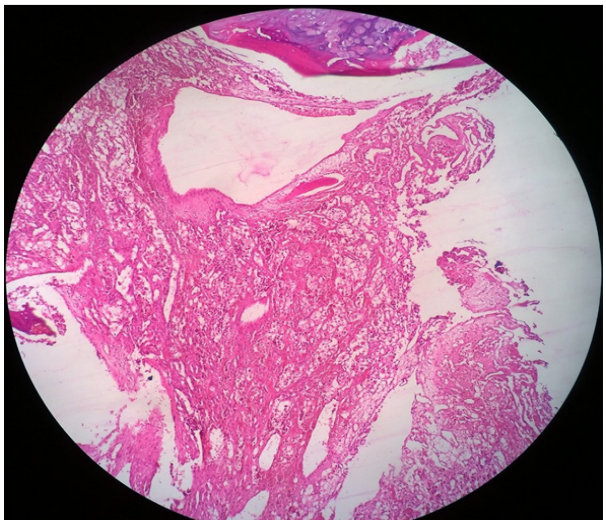

Figure 3: Tumoral cells invading bone trabeccula

of life during next 8 months. He refused continuing chemotherapy. Remaining mass at pelvic area increased in size and patient received (131) I-MIBG therapy, 8 months post-operatively. A second debulking surgery, as well as other palliative measures will be considered for future.

Discussion

Paragangliomas, or extra-adrenal pheochromocytomas, are rare, and may have the same biological, clinical, and radiological manifestations of pheochromocytomas. These tumors, most commonly present in the organ of Zuckerkandl at the aortic bifurcation. Non-functioning retroperitoneal paragangliomas are usually asymptomatic at early stages, and produce symptoms of compression to adjacent organs, when reach to considerable sizes. Pain is present in 50% of cases, and other typical symptoms are nausea, vomiting and abdominal distention,[6] as experienced in this case. Complete surgical resection, remains the mainstay of treatment of extra-adrenal retroperitoneal paragangliomas.[7] Extra-adrenal retroperitoneal paragangliomas tend to be more aggressive, comparing

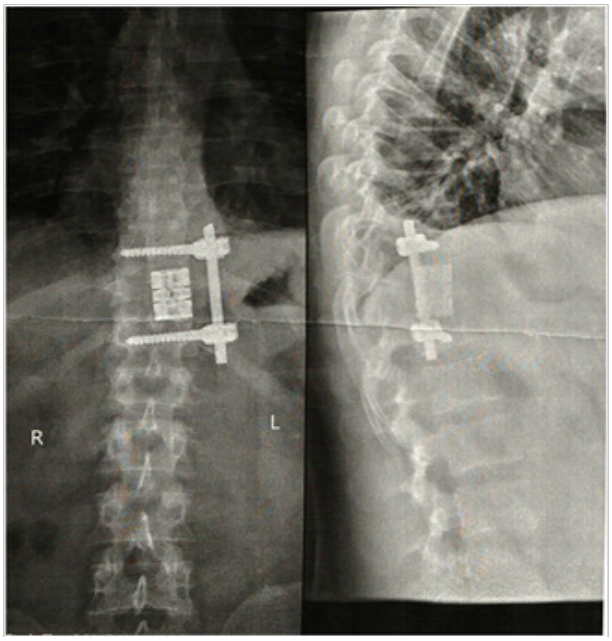

Figure 4: Post-operative view of fixated T11 vertebrae

their adrenal counterpart, and incidence of malignant transformation is higher relative to this tumor at carotid body. [7] Where nonchromaffin tissue is absent, extensive local invasion, and distant metastasis to one or more sites, are considered as malignant behavior of paragangliomas.[8] Local invasion alone, which is not always present in tumors that metastasize, is not sufficient for diagnosis of malignancy in paragangliomas.[9] Bones, lymph nodes and lungs has been reported as common sites of metastatic spread extra-adrenal retroperitoneal malignant paragangliomas.[10] Based on these criteria paraganglioma in our reported case is considered malignant. The prognosis of unresectable or metastatic tumor is poor. Metastases to vertebrae, with or without extradural extension, are rare.[11] Spinal cord compression poses the patients at dangerous condition and needs emergent treatment. External beam radiotherapy, with or without surgical decompression and fixation is considered standard for this dangerous status.[12] In order to better quality of life, other palliative measures, especially debulking of the remaining pelvic mass, are also indicated. Patient scheduled for 131I-MIBG therapy and received this treatment 8 months after surgical debulking. A recent meta-analysis showed that, stabling the disease concerning tumor volume, after 131I-MIBG therapy, could be achieved in 52% patients.[13] Current data suggest that 131I-MIBG therapy on malignant paraganglioma will result to complete response ranged from 0•00 to 0•38, partial response 0•00 to 0•83 and stable the disease 0•00 to 0•90.[13] In patients in whom there is evidence of progressive disease and treatment is palliative, such as our patient, 131I-MIBG therapy is considered a proper selection.[14]

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