CASE REPORT

SPINAL CORD COMPRESSION BY METASTATIC THORACIC SPINE PARAGANGLIOMA

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Paragangliomas are rare neuroendocrine tumours most commonly located in the adrenal glands. The overall incidence of paragangliomas is 0.8 per 100,000 persons, but the incidence of malignant paraganglioma was found to be 93 cases out of 400 million persons in United States. We present a case of 50 year old male who came to the hospital with back pain and progressive bilateral lower limb weakness for the past 6 months. Imaging studies revealed enhancing lesions on dorsal spines. Bone scintigraphy showed increased tracer uptake at multiple sites. Bone biopsy and immune-histochemical staining proved metastatic paraganglioma. After a thorough literature search only few cases of metastatic spine paraganglioma causing spinal cord compression have been reported to date.

Keywords: Paraganglioma; spinal cord compression
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INTRODUCTION

Paraganglioma are rare neuro-endocrine tumours that arise mainly from small organs consisting cells derived mainly from neural crest cells, extra adrenal autonomic paraganglioma and cells having the capability to secrete catecholamines.[1] Paragangliomas can be technically classified as sympathetic and parasympathetic. The sympathetic paragangliomas have got intrinsic capacity to secrete catecholamines and usually present with episodic flushing, hypertension, tachycardia, headache and sweating etc. In contrast majority of parasympathetic paragangliomas are non-functional. Majority of paragangliomas are benign with only few metastasizing to distant sites like spine.

We present a case of a metastatic spine paraganglioma in a 50 year old male who presented initially with back pain. All findings including were consistent with metastatic thoracic spine paraganglioma.

CASE REPORT

A 50 year old male presented with complaints of lower back pain for the last 6 months. The pain started gradually, radiating to both legs. The pain was associated with weakness of both lower limbs. The patient denies any urinary or bowel incontinence and weight loss, numbness, tingling, cough, night sweats or trauma to his back. On physical examination strength in lower limbs was decreased, the reflexes were brisk bilaterally and sensations were decreased up to the level of D4. His labs did not reveal any significant abnormality. MRI dorsal spine showed multiple enhancing lesions within the bodies of the vertebrae and the collapse of D3 and D4 vertebrae (Figure-1). Bone scintigraphy showed increased tracer uptake at multiple levels of the skeleton, including D3 and D4 vertebrae and left greater trochanter suggesting bony metastasis. Bone biopsy revealed cellular lesions in nests and clusters. Individual cells had eosinophillic cytoplasm and nuclear pleomorphism. Intervening stroma also showed vessels of varying sizes. Some fibrosis was also noted. Immunehistochemical staining turned out positive for Vimentin, S-100 and CD-56. These findings are consistent with metastatic paraganglionoma. The primary tumour could not be identified due to financial restraints of patient and lack of availability of advanced diagnostic modalities for paraganglioma. The patient was referred to oncology for palliative chemo and radiotherapy. Due to financial constraints the patient could not be followed appropriately and his spine lesions could not be excised.

DISCUSSION

Paragangliomas are rare neuroendocrine tumours derived from embryonic neural crest cells, mainly found in the autonomic ganglia of thorax, abdomen and pelvis. The sympathetic paragangliomas are usually located outside the head and neck and majority of them secrete catecholamines.[1] Most of the extra-adrenal paragangliomas are located in abdomen.[2] The annual incidence of paragangliomas is 0.8 per 100,000 persons.[3] The estimated incidence of malignant paraganglioma was found to be 93 cases out of 400 million persons.[4] Spine paragangliomas with spinal cord compression as in our case is reported in only a few cases.[5] The majority of paragangliomasare sporadic and about one third is caused by a germline mutation making them a part of important inherited tumour syndrome.[3] Several genes have been implicated in the pathogenesis of

paragangliomas including KIF1Bβ, EGLN1/PHD2, SDHAF2, TMEM127, SDHA, and MAX.[4] Histologically, it is composed of polygonal epithelial cells in nests and trabecular fashion. Nuclear atypia, mitotic figures and local invasion can be present but they are not the signs of malignancy. The malignant potential of these tumours cannot be identified, unless there is metastatic disease and no specific predictor has been studied for malignant paragangliomas.[7] Some studies have proven that malignant paraganglioma is more common in individuals with the SDHB mutation than in any other mutation or sporadic lesions.[8]

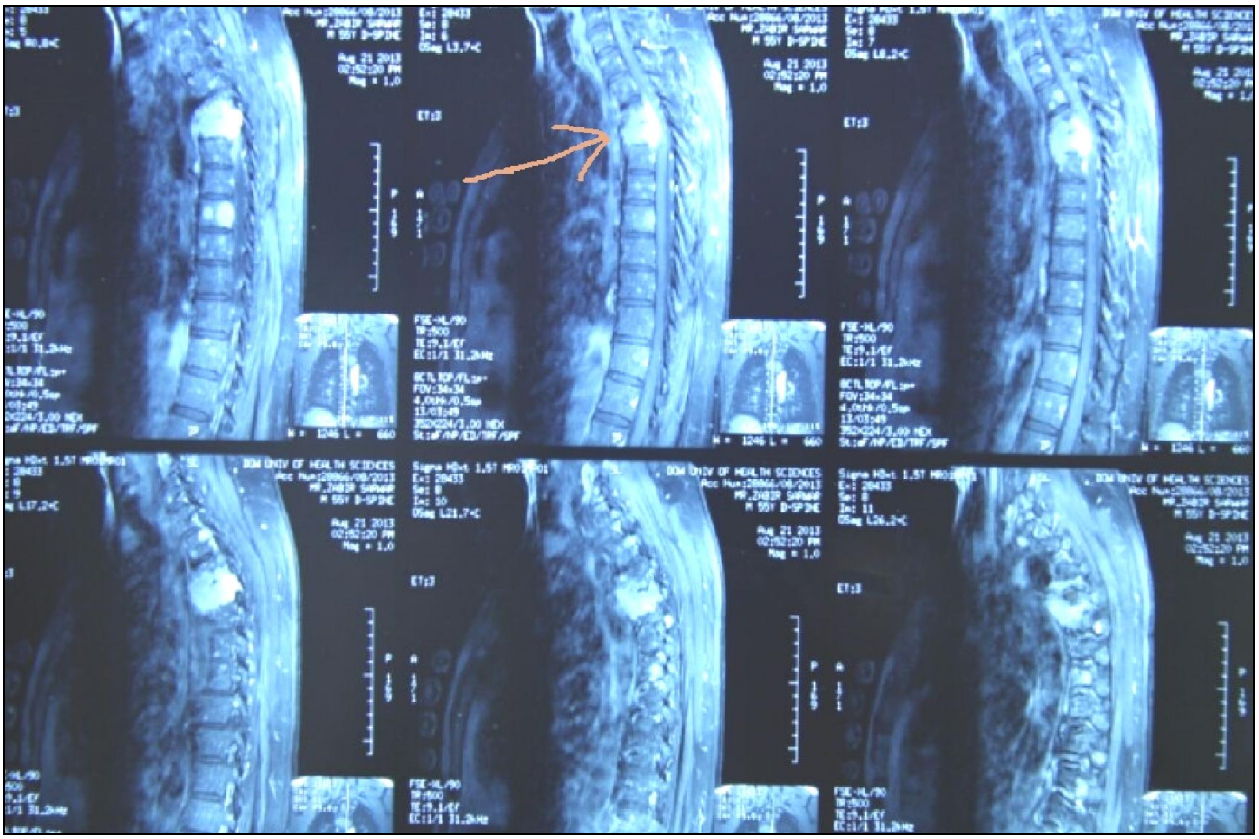

Figure-1: MRI Dorsal Spine with Contrast. Caption: MRI Dorsal Spine showing enhancing lesions at D3 & D4 levels.

The classic triad of pheochromocytoma/ paraganglioma is headache, sweating and palpitation.[9] The diagnosis of secretory tumours can be established by measuring blood and urinary metanephrine, normetanephrine and chromogranin-A levels. Positive results should be followed by imaging studies to locate the tumour. CT scans and MRI both can be of value in adrenal tumours but MRIs are more useful in non-adrenal tumours.[9] Functional imaging with ^{123}I- or ^{131}I-metaiodobenzylguanidine (MIBG) scintigraphy and FDG-PET scanning can detect those tumours which cannot be diagnosed on traditional CT/MRI.[8] The diagnosis of malignant paraganglioma is very difficult because the general histologic features are found in benign lesions as well. Therefor diagnosis of malignancy is established by the presence of metastasis at sites that are devoid of chromaffin tissue.[10] These modalities can also be used to localize distant metastasis. Due to financial constraints of the patient and lack of availability of advanced diagnostic modalities the primary tumour couldn't be identified.

The treatment of spine paraganglioma is surgical resection followed by external beam radiotherapy. The symptoms of catecholamine excess can be managed by administering alpha and beta blockers. Radiotherapy, ablation therapy and trans-arterial chemo embolizations are other alternatives for metastatic disease. ^{131}I-mIBG has also proven benefit in symptomatic as well as hormonal improvement in metastatic neuroendocrine tumours.[11] Targeted therapy with somatostatin analogues has shown to be a novel therapy for metastatic paragangliomas in regard of

survival benefit and symptomatic response.[12] The Prognosis of these tumours is variable. It mainly depends upon location of tumour, rate of progression, extent of metastasis.[13]

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