

## Non Functional Retroperitoneal Paraganglioma Presenting with Extensive Skeletal Disease: A Case Report and Review of Literature

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**Abstract:** Paragangliomas (PGs) are very rare extra adrenal tumors derived from neuroendocrine cells of autonomic nervous system which rarely present as retroperitoneal mass. The Extra-adrenal paragangliomas are reported to account for only 10-15% of all paragangliomas. They may present incidentally as a mass. Histological findings are used for definitive diagnosis and surgery is the treatment of choice. We report a case of a non functional retroperitoneal PG with extensive skeletal metastasis in a 22-year-old gentleman treated with palliative chemotherapy.

**Keywords:** Paragangliomas, neuroendocrine, skeletal metastasis.

### INTRODUCTION

Paragangliomas (PGs) are uncommon tumors; arise from the neural crest tissue that develops into sympathetic and parasympathetic paraganglia. Paraganglioma of adrenal medulla is called as pheochromocytoma, while those located outside of adrenal gland are called as extra-adrenal paragangliomas. On the basis of their ability to secrete hormones/peptides, paragangliomas are classified as functioning and nonfunctioning. The functional paragangliomas secrete nor epinephrine and normetanephrine with symptoms such as fluctuating or episodic hypertension, headache, and sweating. The nonfunctioning paragangliomas are present incidentally as a mass and are mostly asymptomatic [1]. Literature review revealed few cases of non functional PG presenting as retroperitoneal mass and extensive skeletal metastasis.

### CASE REPORT

We present a case of 22 year old male, presented with pain & lump in right lower abdomen since 6 months. He also noted left supraclavicular swelling and scalp swelling in occipital region since 1 month duration (Fig. 1). No other significant history noted. Abdominal examination revealed large palpable lump in right iliac fossa extending up to right hypochondrium with restricted mobility approximately 12 x 10 cm in dimensions with ill defined borders. Laboratory investigations and the tumor markers (AFP, LDH and Beta HCG) were within the normal range.

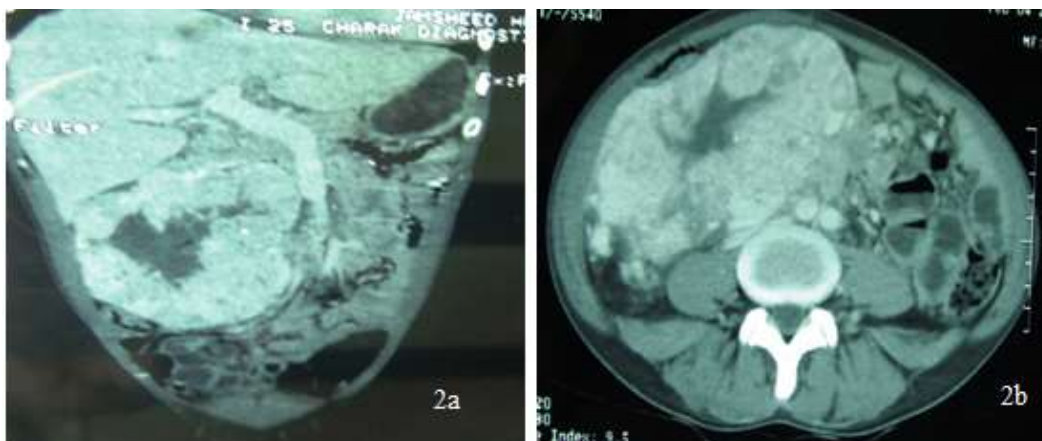
CECT scan of whole abdomen & chest revealed a large heterogeneous enhancing soft tissue mass with necrosis located medially within right perinephric space displacing renal vein anteriorly, renal artery and ureter posteriorly. The mass lesion was associated with liver metastasis (Fig. 2a & 2b). There was mediastinal adenopathy with no evidence of pulmonary metastases. Bone scan (Technetium-99m Methylene Diphosphonate) was performed which revealed multiple bony metastases with lateral displacement of left kidney due to clinically known retroperitoneal mass (Fig. 3). Incision biopsy of left supraclavicular node was done and histopathology revealed paraganglioma with tumor cells (Fig. 4a) expressing positivity for synaptophysin, chromogranin (Fig. 4b) and S-100. His vanillylmandelic acid level was mildly raised (30 mg; Normal <13.6 mg) Biochemical analysis revealed normal serum cortisol (17mcg/Dl, normal [4-22 mcg/dL]), serum aldosterone (1 ng/Dl, normal [1-13 ng/dL]) and plasma rennin activity (3.13 ng/mL/h, normal [0.25-5.82 ng/ml/h]). In view of multiple bony metastases, I-131 labelled Meta Iodo Benzyl Guanidine (MIBG) therapy was considered and a diagnostic whole body scan with 1 mCi of 131I-MIBG was performed to assess extent and avidity of functioning metastases for MIBG. This scan showed widespread MIBG-avid bony metastases involving predominantly axial skeleton (Fig. 5a & 5b). His serum Chromogranin-A (CgA) level was 592 ng/mL (Normal: 1.9-15 ng/mL). His 24 hour urinary metanephrine was 151 mcg (normal range is 25-222 mcg/24hr), normetanephrine was 620mcg (normal range is 40-412 mcg/24hr) and total metanephrines was 771 (normal

range is 94-604 mcg/24 hr). Patient was started on palliative chemotherapy (Cisplatin, Etoposide and Zoledronic acid). After receiving 4 cycles of chemotherapy, patient had clinical and radiologic

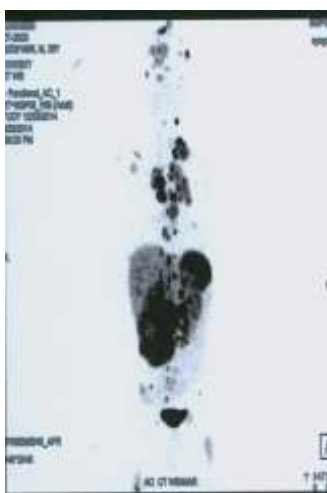
response and chemotherapy was continued further for two more cycles. Patient lost follow up after 6 months of chemotherapy.



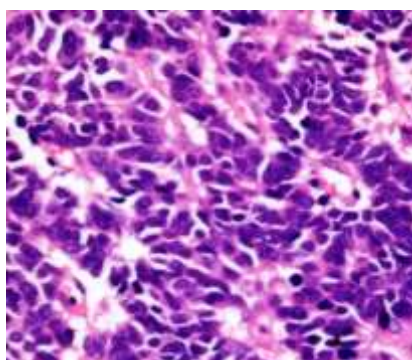
**Fig. 1:** Clinical photograph showing scalp swelling in occipital region



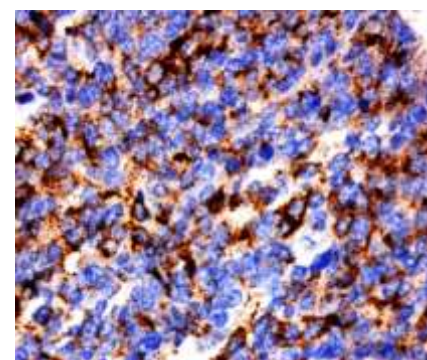
**Fig. 2a & 2b:** CECT scan revealed a large heterogeneous enhancing soft tissue mass with necrosis located medially within right paraneurial region with evidence of liver metastasis



**Fig. 3:** Bone scan revealed multiple bony metastases



**Fig. 4a:** Hematoxylin-and-eosin section showed sheets of small round cells with prominent nucleoli, and coarse "salt and pepper" chromatin



**Fig. 4b:** Tumor cells stained positive for chromogranin

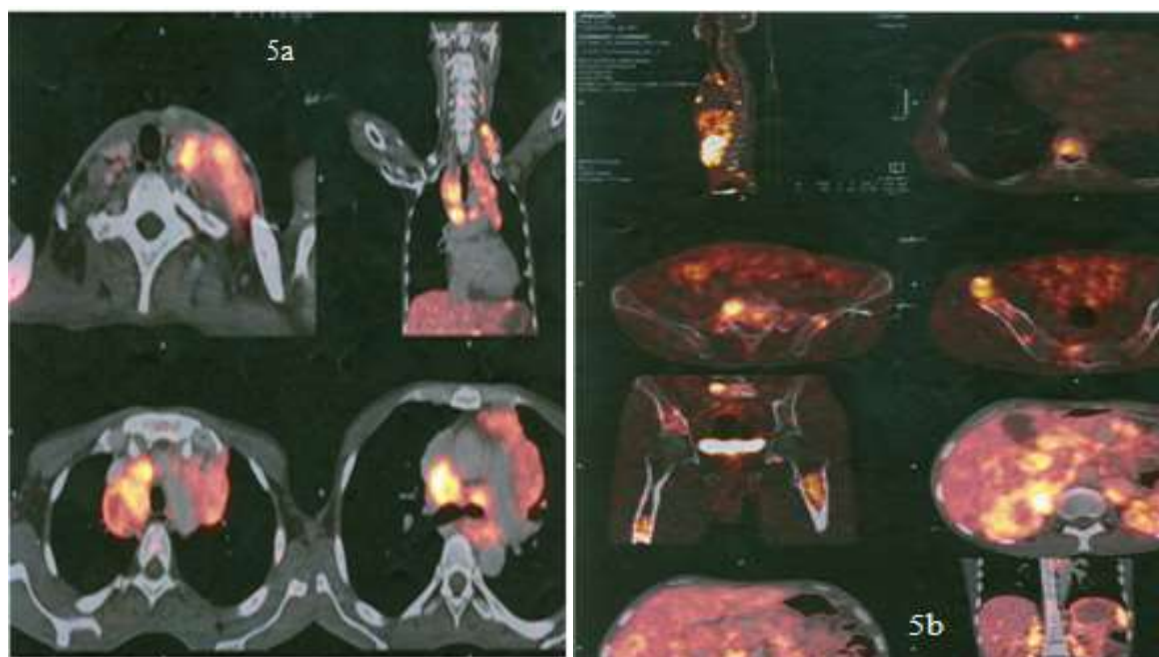


Fig. 5a & 5b: 131I-MIBG scan showed widespread MIBG-avid bony metastases involving predominantly axial skeleton with uptake in mediastinal and supraclavicular region

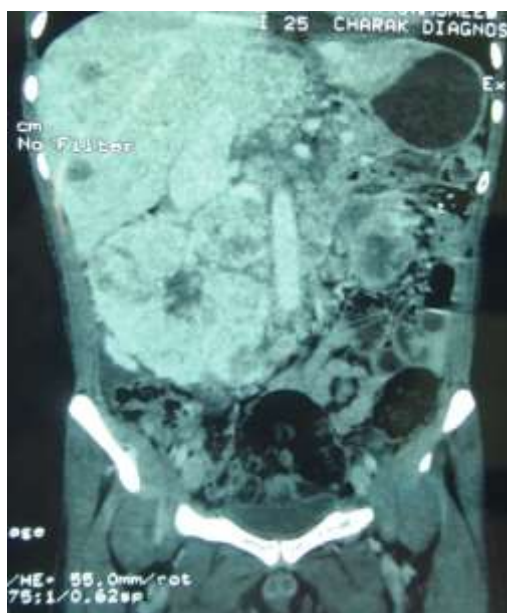


Fig. 6: Post chemotherapy CT scan showing slight reduction in tumor size (partial response)

## DISCUSSION

Extra-adrenal paragangliomas have been reported to account for 10-15% of all adult paraganglioma [1] and can develop anywhere along the midline of retroperitoneum [2]. Most common age of presentation is between 30 and 45 years with slightly male predominance [2, 3].

Genetic mutation in the succinate dehydrogenase B unit (SDHB) and succinate dehydrogenase D unit (SDHD) have been found to be associated with increased risk for extra-adrenal

paragangliomas [4]. The association between extra-adrenal paraganglioma, gastrointestinal stromal tumor and pulmonary chondroma is known as the Carney's triad [5]. Abdominal paragangliomas accounts for 85% of all extra adrenal paragangliomas. They are mostly retroperitoneal in location. The most common site for retroperitoneal paragangliomas is organ of Zuckerkandl. Paragangliomas are called as chemodectomas when they arise from jugulotympanic body and when they originate from the carotid body; they are called as carotid body tumors [6]. Majority of nonfunctional paragangliomas are found incidentally or they are



present with backache or palpable mass with symptoms of surrounding organ compression [1, 7]. Non functional paraganglioma may present with extensive metastasis and has aggressive course and poor outcome.

Magnetic resonance imaging (MRI) is recommended as the first imaging modality for the evaluation of extra-adrenal paragangliomas because of its superior tissue characterization and absence of radiation exposure [5]. Computerized tomography (CT) scan has been reported to have identification sensitivity of around 90% [8]. Meta iodo benzyl guanidine (MIBG) scintigraphy has lacks sensitivity for extra-adrenal paragangliomas. Positron emission tomography scan is more sensitivity when compared to MIBG scintigraphy [9].

Extra-adrenal paragangliomas have a malignant potential [10]. Extensive local invasion and distant metastasis to liver, bone, and lymph nodes have been used as indicators for malignancy [1]. Parameters like tumor size >10cm, tumor weight >80 gm, presence of necrosis, younger age carries increased risk of malignancy. In the present case, tumor size >5 cm, young age, presence of tumor necrosis and evidence of distant lesions were in favor of malignant Paraganglioma.

Surgery with complete removal of mass by laparoscopy or traditional laparotomy is the treatment of choice. Adjuvant radiotherapy is needed in the patient with metastatic disease [1, 10]. Owing to its malignant potential and higher recurrence rate, lifelong follow up is usually recommended [1, 9].

In the present, non functional paraganglioma of retroperitoneum presented with extensive metastasis was treated with palliative chemotherapy, as patient was not affordable for radionuclide therapy. Patient had partial response to chemotherapy with disease control duration of 6 months.

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