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CASE REPORT

Metastatic Paraganglioma: Recurrence in Uncommon Sites

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Abstract

Paragangliomas are rare neuroendocrine tumors, and mediastinal involvement is exceptionally uncommon. We describe a case of metastatic mediastinal paraganglioma in a 54-year-old woman, occurring 26 years after resection of a retroperitoneal paraganglioma. CT imaging revealed both a posterior mediastinal mass and a liver lesion. FNAC and biopsy from both sites demonstrated features typical of paraganglioma, including zellballen architecture and salt-and-pepper chromatin. The tumor was non-functional. This case highlights the potential for very late metastasis and underscores the need for long-term surveillance, integrated diagnostic evaluation, and genetic testing in paraganglioma management.

Key Words

Paraganglioma, Mediastinum, Liver Metastasis, FNAC, Neuroendocrine Tumor

Introduction

Paragangliomas are uncommon neuroendocrine tumors arising from extra-adrenal paraganglia derived from neural crest cells[1]. Their behavior varies widely depending on site and functional status. While head and neck paragangliomas are often benign and non-functional, thoracic and abdominal paragangliomas demonstrate a greater malignant potential.

Mediastinal paragangliomas represent <2% of all mediastinal tumors and may remain undetected until they become large or metastasize[2]. Malignancy in paragangliomas is defined by the presence of metastasis, as histological features alone cannot reliably predict aggressive behavior. Functional tumors produce catecholamines, whereas non-functional tumors may present with nonspecific symptoms or be detected incidentally[3].

We present a rare case of metastatic mediastinal paraganglioma with synchronous liver involvement, occurring more than two decades after excision of a retroperitoneal primary tumor, highlighting its clinical presentation, radiological findings, histopathological features, and therapeutic approach.

Case Report

A 54-year-old female came to the Oncology OPD with complaints of left-sided abdominal pain. The patient was a known operated case of retroperitoneal extra-adrenal paraganglioma 26 years ago. The patient was not hypertensive and had no history related to neuroendocrine hormone over-secretion. On CT abdomen, a hyperenhancing mass lesion with central cystic areas of necrosis in the left paravertebral region of the posterior mediastinum extending from D8–D10 vertebral levels measured 52 × 46 × 62 mm. It also showed a peripherally enhancing centrally hypodense lesion in segment VIII/VII of the liver (homogenous and significant enhancement with perilesional halo in arterial and portal venous phase) measuring 25 × 23 mm (Figure 1, 2).

The patient was sent to the department of pathology

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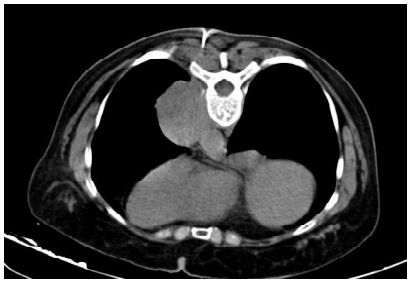

Figure 1: On CTthorax hyper enhancing mass lesion with central cystic areas necrosis in left paravertebral region of posterior mediastinum.

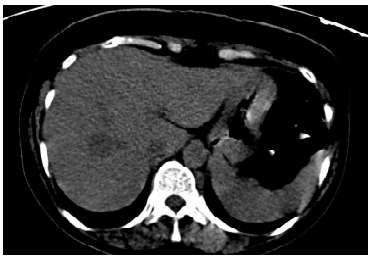

Figure 2: On CTabdomen peripherally enhancing centrally hypodense lesion in segment VIII/VII of liver.

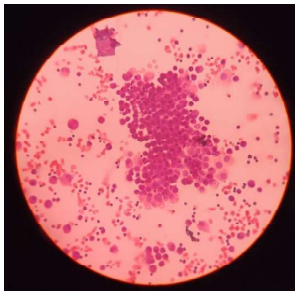

Figure 3: FNAC from mediastinal mass showed neoplastic cells with pleomorphic nuclei with granular chromatin (400X H&E).

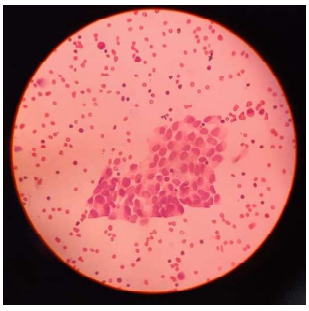

Figure 4: FNAC from liver mass show neoplastic cells with pleomorphic nuclei with granular chromatin (400X H&E).

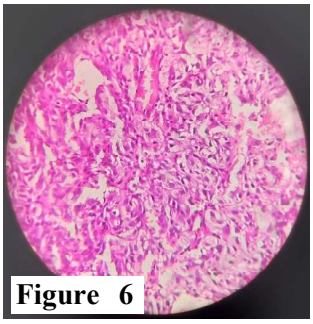

Figure 5, 6 : Microscopy - Nested zell-ballen type of arrangement and salt and pepper type of chromatin (100X, 400X H&E).

for CT-guided FNAC from both the mediastinal and liver masses. FNAC of the masses was performed using a 23G needle attached to a 10 ml syringe. Aspirate was obtained and smears with material were fixed in 95% isopropyl alcohol and were stained with H and E stain.

On microscopic examination, FNAC from the mediastinal mass showed atypical cells arranged in loosely cohesive sheets, clusters, and scattered singly. Individual neoplastic cells were round to oval, having round to oval mildly pleomorphic nuclei with granular chromatin, indistinct nucleoli, and a moderate to ample amount of eosinophilic cytoplasm. Few binucleated and trinucleated cells were noted. The background showed lymphocytes, foamy macrophages in fluid material mixed with RBCs (Figure 3).

On microscopic examination, FNAC from the liver mass showed moderately cellular smears showing neoplastic cells arranged in loosely cohesive sheets, clusters, and scattered singly. Individual cells were round to oval, having round to oval mild to moderately pleomorphic nuclei with granular chromatin, indistinct nucleoli, and a moderate to ample amount of eosinophilic cytoplasm. Few of the cells showed vacuolated cytoplasm. Few binucleated cells were also noted. Many bare atypical nuclei were noted. The background showed few inflammatory cells, few stromal fragments, and numerous RBCs (Figure 4).

The impression was given as positive for neoplastic cells having low to moderate nuclear atypia, with the possibility of metastatic paraganglioma of both the mediastinal and liver masses.

Core needle biopsy was done and sent for histopathological examination. On gross examination, multiple grey-brown tissue cores were sent. On

histopathological examination, the specimen showed a nested zell-ballen type of arrangement and salt-and-pepper type of chromatin. The tumor was diagnosed on histopathology as metastatic paraganglioma (Figure 5, 6).

Discussion

Paragangliomas are rare neuroendocrine tumors that arise from extra-adrenal paraganglionic tissue and show variable clinical behavior. Sympathetic paragangliomas, such as those in the thoracic and abdominal regions, carry a higher risk of malignancy and metastatic spread compared to parasympathetic tumors[4]. Mediastinal paragangliomas are particularly uncommon, and their presentation may range from functional tumors producing catecholamine excess to non-functional masses detected incidentally or due to compressive symptoms[5].

In the present case, the patient developed a non-functional mediastinal paraganglioma with hepatic metastasis decades after initial surgery, highlighting the tumor's potential for late recurrence and spread to uncommon sites. The demonstration of metastasis remains the only definitive criterion for diagnosing malignant paraganglioma.

Histologically, these tumors characteristically show a zellballen pattern, and immunohistochemistry with markers such as chromogranin A, synaptophysin, NSE, and S100 supports the diagnosis. As noted in this case, histology alone cannot predict malignant potential, and the presence of liver metastasis confirmed the diagnosis of metastatic paraganglioma[6].

Imaging plays a crucial role in detecting both local recurrence and distant metastasis. CT and MRI provide structural details, while functional imaging modalities such as 123I-MIBG, 18F-FDG PET, or 68Ga-DOTATATE PET/CT help identify multifocal or metastatic disease[7].

Management of metastatic paraganglioma is complex. While surgery is ideal for localized disease, recurrent or metastatic tumors often require systemic therapy, radionuclide-based treatments, or locoregional interventions for metastatic sites like the liver. Prognosis largely depends on disease burden and treatment response[8].

Given that up to 40% of paragangliomas have an underlying genetic mutation, especially involving the SDH gene complex, genetic testing is important for guiding surveillance and family screening[9].

This case emphasizes the importance of long-term follow-up in paraganglioma patients and demonstrates that recurrence may occur after many years and at unusual sites. Awareness of such presentations can aid in timely recognition and appropriate management.

Conclusion

This case illustrates late metastatic recurrence of paraganglioma at uncommon mediastinal and hepatic sites decades after initial surgery. It emphasizes the tumor's unpredictable behavior, need for lifelong surveillance, and importance of imaging and histopathology for diagnosis. Early detection and multidisciplinary management remain crucial for improving outcomes.

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