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Metastasis of Malignant Pheochromocytoma to the Sphenoid Sinus: A Rare Entity

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Abstract **Case Report**

Pheochromocytomas are uncommon neuroendocrine tumors of the adrenal medulla. Malignant behavior is seen in approximately 10% of these lesions, evidenced by distant metastasis to sites without chromaffin tissue. Here we report a rare case of the sphenoid sinus metastases of an adrenal pheochromocytoma in a 80-year-old man.

Keywords: Pheochromocytoma, Metastasis, Sphenoid sinus.

INTRODUCTION

Pheochromocytomas are rare neuroendocrine tumors of the chromaffin system. Malignant behavior is seen in approximately 10% of these lesions, evidenced by distant metastasis to sites without chromaffin tissue. Metastatic pheochromocytoma is most commonly reported in bone, liver, kidney, lung, and lymph nodes. Sphenoid sinus metastases are relatively uncommon, with reported incidence varying between 1% to 8% of cancer discovered elsewhere in the body

CASE REPORT

A 80-year-old man with a history of malignant adrenal pheochromocytoma complained of numbness of the right side face followed by a headache and diplopia. Physical examination revealed right abducens palsy and right oculomotor palsy. Visual acuity and visual field testing were intact, with absence of papilledema. MRI and CT scan showed a homogeneously enhancing mass in the clivus and sphenoid sinus, which extended into the right cavernous sinus and appeared to compress the optic chiasma. Nasal endoscopy revealed a multilobulated hypervascular mass on the right, emanating from the spheno ethmoidal recess down into the posterior nasopharynx. Biopsy results were compatible with a metastasis of malignant pheochromocytoma.

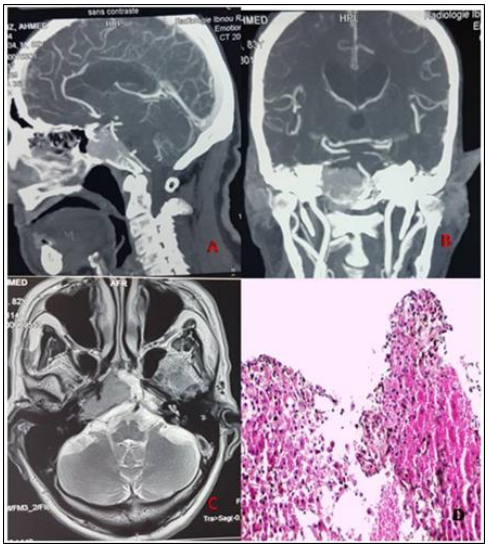

Fig-1: CT scan (A, B) and MRI (C) showing a homogeneously enhancing mass in the clivus and sphenoid sinus. Histopathological slides (H & E stain) (D) showing characteristic polygonal nests of pheochromocytoma cells.

DISCUSSION

Pheochromocytoma is an uncommon catecholamine-secreting tumor of the adrenal medulla [1, 2, 3]. Malignant pheochromocytomas constitute 10% of these tumors, as demonstrated by their potential to metastasize to distant sites that lack chromaffin tissue. Metastatic pheochromocytoma is most commonly reported in liver, kidney, lung, bone, and

lymph nodes [2, 3]. Currently, there are no reliable clinical, histological, or molecular predictors of metastatic potential or prognosis of pheochromocytoma, although criteria that may support malignant disposition have been suggested. The rate of metastasis has traditionally been accepted as 10%, but because of differences in histopathological diagnostic criteria and definitions of malignancy, reported rates vary between 5% and 26% (3). Even with treatment, the prognosis of malignant pheochromocytoma remains dismal with 50% reported mortality in 5 years (2).

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