Original Research Paper

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Pathology

CELL BLOCK TO THE AID IN THE DIAGNOSIS OF A RARE CASE OF METASTATIC PARAGANGLIOMA- A DEFINITIVE WAY OUT.

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ABSTRACT Paragangliomas are neural crest derived neoplasms,commonly arising in head, neck and upper thorax. Head and neck paragangliomas are generally considered to be benign but rarely , about 5% produce metastasis. We report a case of a 41 year old male presenting with pulsatile left neck mass along with lytic rib lesion as detected radiologically. Fine needle aspirations from both the lesions showed similar cytomorphology of loose clusters of round to ovoid cells having granular cytoplasm, indistinct borders and occasional anisokaryosis. The differentials were metastatic paraganglioma and metastatic follicular carcinoma of thyroid. Surgery followed by histology revealed neck lesion to be paraganglioma. Immunohistochemistry on cell block prepared from rib lesion aspirate showed positivity for Chromogranin A, implying it to be metastatic paraganglioma. We present this rare entity (metastatic paraganglioma) where cell block study with immunohistochemistry came to the aid in diagnosing a lesion that was difficult to sample surgically.

KEYWORDS : Cell Block Preparation, Chromogranin A, Immunohistochemistry, Metastatic paraganglioma.

INTRODUCTION

Paragangliomas are neoplasms originating from embryonic neural crest throughout the body. These are rare slow growing, painless tumors with the ability to secrete catecholamines, thus presenting with hypertensive episodes and syncopal attacks. Paragangliomas of head and neck region account for 3% of total cases and are found in carotid body, vagus nerve and jugulotympanic area. Von Luschka in 1862 first described carotid body paraganglioma.[1] It has various synonyms such as chemodectomas, Glomus tumors, non-chromaffin tumors etc. Paragangliomas are often malignant and very rarely metastasise.[2]

CASE REPORT

A 41 year old male presented with a slow growing mass for 5 years in left side of neck without any systemic complaint. On local examination, the mass was pulsatile in nature. Routine physical examination of the patient revealed mild hypertension and tachycardia.

CECT of neck showed a soft tissue mass approximately 3.9cm×3.1cm in size with intense contrast enhancement and a small hypodensity within, located at left submandibular region near angle of mandible and lateral neck space[Fig 1], abutting the margin of internal carotid artery. A provisional diagnosis of Paraganglioma was given. A subsequent CT scan of thorax revealed an expansile lytic lesion at 2nd rib with soft tissue component [Fig 2].

Direct FNAC from neck lesion and CT guided FNAC from the rib lesion showed similar cytomorphology of mild anisonucleosis, stippled chromatin, scanty to moderate pale blue or finely granular cytoplasm with indistinct cytoplasmic border[Fig.3]. Scattered bare nuclei were also seen in background and some loose cell clusters showed admixture of spindle cells. Occasional follicular formation of cells also noted. Later surgical excision of the neck lesion followed by histology was done.

Grossly it was firm, brownish, well circumscribed and partially capsulated. On cut section there were areas of hemorrhage and cystic degeneration. Microscopically well-defined nests of cuboidal cells with abundant granular basophilic cytoplasm arranged in distinctive 'Zellballen' pattern, separated by prominent fibrovascular stroma present in a focal chronic inflammatory background, confirmed it to be Paraganglioma.

As the rib was a difficult site for excisional biopsy, cell block preparation was done[Fig.4] from the aspirate of the rib lesion followed by immunohistochemistry. Microscopy revealed clusters as well as follicular formation by the lesional cells. These cells showed strong cytoplasmic positivity for Chromogranin-A[Fig.5], hence excluded the differentials other than metastatic paraganglioma.

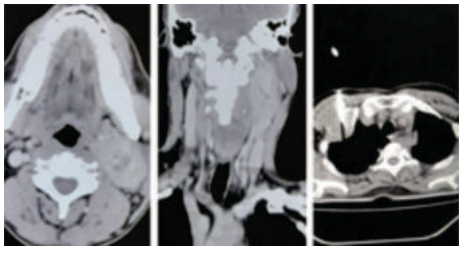

Fig 1:-CECT Of Neck(Horizontal & Coronal View) & CT Scan Of Rib Lesion

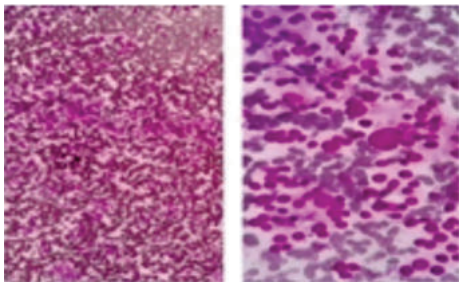

Fig 2:-10X & 40X View Of FNA Cytology Of Rib Lesion

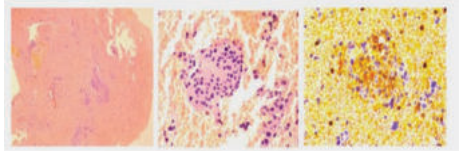

Fig 3:- CB Preparation(10X & 40X view) & Its Positivity For Chromogranin A

DISCUSSION

Parganglioma is tumor of chromaffin cells that rarely metastasize.[3] Most of the malignant paragangliomas are SDHB or SDHD gene mutation associated. Lymphovascular or perineural invasion does not indicate its aggressiveness.

Although paragangliomas can occur throughout the body, carotid body paraganglioma is one of the most common. It has more or less equal incidence in males and females. It commonly gets diagnosed between third and fifth decade of life.

Presentation of paraganglioma is variable. Classical symptoms , headache, palpitation, sweating, episodic hypertension are due to increased secretion of catecholamines. Often hyperglycemia, fever, panic attacks, weight loss, myocardial infarction, osteolytic lesions can be manifested . Early diagnosis and appropriate treatment of it can reduce morbidity associated with symptomatic and malignant disease. Microscopically as in our case, classical paragangliomas are highly vascular tumor consisting of two types of cells, Type I: Chief cells which are numerous and containing catecholamine bound neurosecretory granules and Type II: Sustentacular cells present in the periphery of the 'Zellballen' pattern.[1] This pattern can be also found in carcinoid, malignant melanoma and medullary thyroid carcinoma. The cell block of rib aspirate in this case consisted of round cells in follicular pattern as well as in clusters. This microscopic feature along with the concurrent presence of a neck lesion raised the differentials of a metastatic follicular carcinoma of thyroid or a metastatic paraganglioma. However Immunohistochemistry of Chromogranin-A strong positivity helped to reach a conclusive diagnosis of metastatic paraganglioma.

Metastatic paraganglioma is a very rare entity. Cell block preparation along with IHC can be very helpful in challenging cases that are difficult to sample surgically.

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