MS165-ED-X

The Molecular Subtypes of Pheochromocytoma and Paragangliomas (PPGLs): The Cancer Genome Atlas Transcriptome Data Derived Taxonomic Scheme and 2017 WHO Classification System

All Day Room: MS Community, Learning Center Digital Education Exhibit

Participants
Sireesha Yedururi, MBBS, Sugar Land, TX (*Presenter*) Nothing to Disclose
Ajaykumar C. Morani, MD, Houston, TX (*Abstract Co-Author*) Nothing to Disclose
Nahyun Jo, MD, Houston, TX (*Abstract Co-Author*) Nothing to Disclose
Leonardo P. Marcal, MD, Houston, TX (*Abstract Co-Author*) Nothing to Disclose
Devaki Shilpa S. Surasi, MD, MBBS, Bellaire, TX (*Abstract Co-Author*) Nothing to Disclose
Nisha S. Ramani, Houston, TX (*Abstract Co-Author*) Nothing to Disclose
Venkata S. Katabathina, MD, San Antonio, TX (*Abstract Co-Author*) Nothing to Disclose
Srinivasa R. Prasad, MD, Houston, TX (*Abstract Co-Author*) Nothing to Disclose

For information about this presentation, contact:

syedururi@mdanderson.org

TEACHING POINTS

Pheochromocytomas and paragangliomas (PPGLs) are tumors arising from adrenal and extra-adrenal chromaffin cells. Currently associated with germline and/or somatic mutations in more than 20 genes. Up to 40% are associated with inherited germline mutations, most common genes include SDHB, SDHD, VHL, RET and NF1. Genotype/phenotype correlations exist between germline mutation, tumor location and biochemical secretion. Four main molecular subtypes are identified-1. Pseudohypoxia, 2. Kinase-signaling , 3.Wnt-signaling and 4. Cortical admixture. SDHB mutations are associated with increased risk of metastatic disease. Understanding tumor biology has expanded the scope of personlized care and treatment in PPGLs.

TABLE OF CONTENTS/OUTLINE

Introduction. The 2017 WHO clasiification system of PPGLs. Epidemiology and heritability of tumors. Oncogenic pathwyas of the four subtypes of tumors. Genetic syndromes predisposing to PPGLs. Common locations of tumor involvement and risk of metastatic disease. Imaging approach to diagnosis and staging. Emerging role of new functional agents in diagnosis and treatment. Emerging screening, treatment and follow up guidelines. Future precision oncology tretament options. Conclusion.