Malignant PEComa of Stomach: A Rare Malignancy

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Background: Perivascular epithelioid cell tumor (PEComa) is characterized by perivascular location and the tumor cells have mostly epithelioid and spindle appearance with clear to lightly granular eosinophilic cytoplasm, a round-to-oval centrally located nucleus. Immunohistochemically, nearly all PEComas show reactivity for melanocytic (HMB-45 and/or Melan-A) and smooth muscle (actin and/or desmin) markers. Malignant Gastric PEComa is extremely rare and only 3 cases have been reported to best of our knowledge. We report a 4th case of malignant gastric PEComa.

Case: We are presenting a case of 48 years old Caucasian female who presents to emergency department with complains of progressively worsening of intermittent abdominal pain for few weeks associated nausea, vomiting and decreased appetite. CT Abdomen/Pelvis with contrast showed a mass at greater curvature of stomach. Patient underwent resection of mass and found to have malignant PEC-oma.

Conclusion: Perivascular epithelioid cell tumors (PEComas) are family of rare mesenchymal tumors composed of histologically and immunohistochemically distinctive perivascular epithelioid cells. These tumors co-express muscle and melanotic markers. Surgical resection is the best treatment option. They arise most commonly in the retroperitoneum, visceral and abdominopelvic sites. Colon is most the common site followed by small intestine in GI tract