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Summary

AN EXTREMELY RARE CASE OF PRIMARY EPITHELIOID RHABDOMYOSARCOMA OF THE STOMACH

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An 85-year-old man with primitive gastric epithelioid rhabdomyosarcoma presented with metastatic disease at diagnosis and died 20 days later, highlighting the aggressive nature and diagnostic challenges of this extremely rare tumor.

KEY POINTS

- Epithelioid rhabdomyosarcoma is a recently recognized highly aggressive rare variety not yet included in the WHO classification, characterized by poorly differentiated cells with desmin expression and specific skeletal muscle markers
- The patient presented with a 2.5 cm gastroesophageal junction lesion with rapid symptom onset over less than a month, and metastases to multiple sites including the brain were identified on CT scan
- Diagnosis required specific immunohistochemical staining showing negativity for cytokeratin (MN116) and positivity for CD10, Vimentin, Desmin, and MyoD1, with fluctuating p53
- The patient was eligible only for best supportive care due to advanced disease and clinical condition, dying approximately 20 days after diagnosis
- Proper immunohistochemical staining is crucial when evaluating malignant cells with epithelioid morphology, as gastric epithelioid rhabdomyosarcoma presents diagnostic challenges even for experienced pathologists

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