Histological tumour type (Core)

Histological diagnosis is based on the 2020 World Health Organization Classification of Soft Tissue and Bone Tumours, 5th edition.[1] Gastrointestinal stromal tumours (GISTs) are most often of spindle cell type, followed by epithelioid type and mixed epithelioid and spindle cell type;[1] the latter two histological types are most common in the stomach. The histological tumour type may be associated with mutational status (e.g., most *PDGFRA*-mutant GISTs are of epithelioid type)[2] or particular syndromes (e.g., Carney triad and Carney-Stratakis syndrome-associated GISTs are usually of epithelioid or mixed type),[3] although this is not always the case.

Pleomorphic morphology in GIST is rare (<2%). Dedifferentiated GIST, defined as the abrupt transition from conventional spindle cell or epithelioid GIST to an anaplastic sarcomatous appearance, usually accompanied by loss of the expression of lineage markers (e.g., KIT and ANO1/DOG1), is exceptionally rare.[4]

References

1 WHO Classification of Tumours Editorial Board (2020). *Soft Tissue and Bone Tumours. WHO Classification of Tumours, 5th Edition, Volume 3*. IARC Publications, Lyon.

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3 Zhang L, Smyrk TC, Young WF, Jr., Stratakis CA and Carney JA (2010). Gastric stromal tumors in Carney triad are different clinically, pathologically, and behaviorally from sporadic gastric gastrointestinal stromal tumors: findings in 104 cases. *Am J Surg Pathol* 34(1):53-64.

4 Antonescu CR, Romeo S, Zhang L, Nafa K, Hornick JL, Nielsen GP, Mino-Kenudson M, Huang HY, Mosquera JM, Dei Tos PA and Fletcher CD (2013). Dedifferentiation in gastrointestinal stromal tumor to an anaplastic KIT-negative phenotype: a diagnostic pitfall: morphologic and molecular characterization of 8 cases occurring either de novo or after imatinib therapy. *Am J Surg Pathol* 37(3):385-392.