

CIC-DUX4 fusion-positive round cell sarcoma

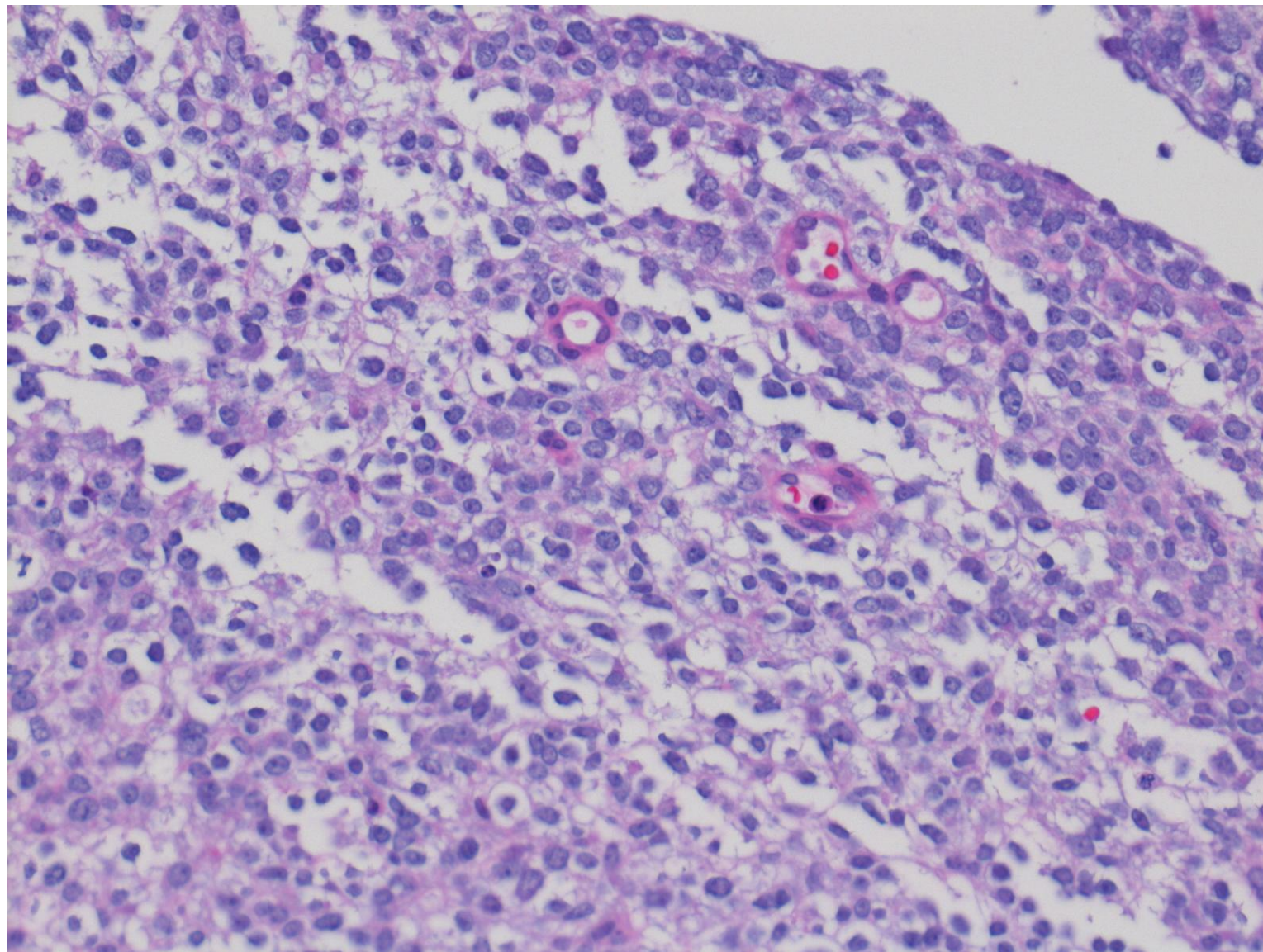
CIC-DUX4 fusion-positive round cell tumor (CIC-DUX4 sarcoma) represents undifferentiated Ewing-like (CD99-positive) small round cell sarcoma. The fusion oncoprotein comprise the transcriptional repressor Capicua (CIC) fused to the C-terminal transcriptional activation domain of DUX4. CIC-DUX4 sarcoma is a high-grade Ewing-like sarcoma affecting young adults and presents as a soft tissue mass. DUX4 immunostaining specifically detects this rare malignancy. The prognosis is poor. The present tumor presented as a huge omental mass in a young lady.

Ref.: Siegele B, et al. DUX4 immunohistochemistry is a highly sensitive and specific marker for CIC-DUX4 fusion-positive round cell tumor. Am J Surg Pathol 2017; 41(3): 423-429. doi: 10.1097/PAS.0000000000000772.

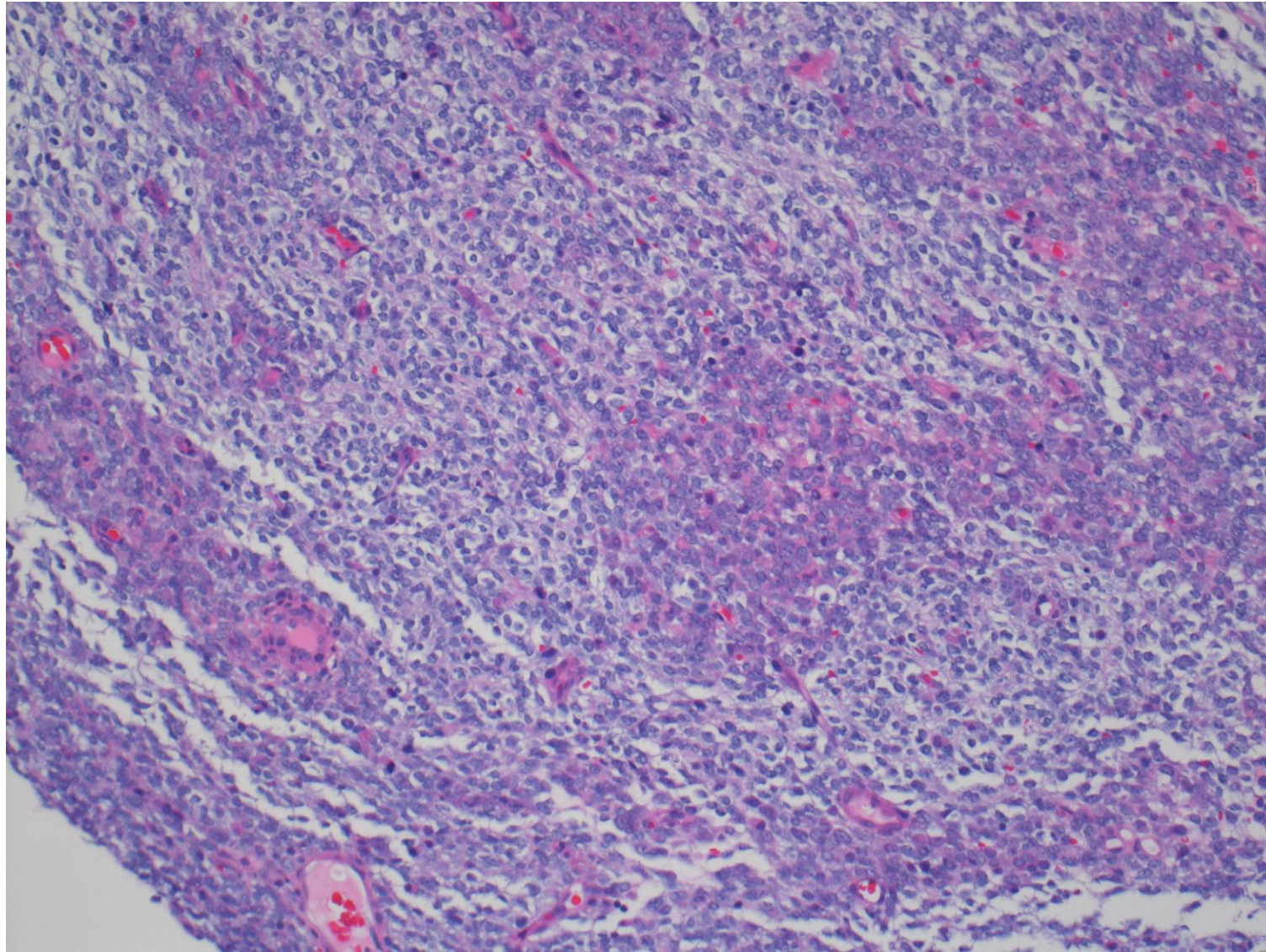
CIC-DUX4 fusion-positive round cell sarcoma of the omentum (19F)

A huge (adult head-sized) omental tumor weighing 1.7 kg was surgically removed from a 19-year-old female patient. Massive hemorrhagic ascites (4,000 mL) was associated. No direct contact with the bowel, oviduct and ovary was detected.

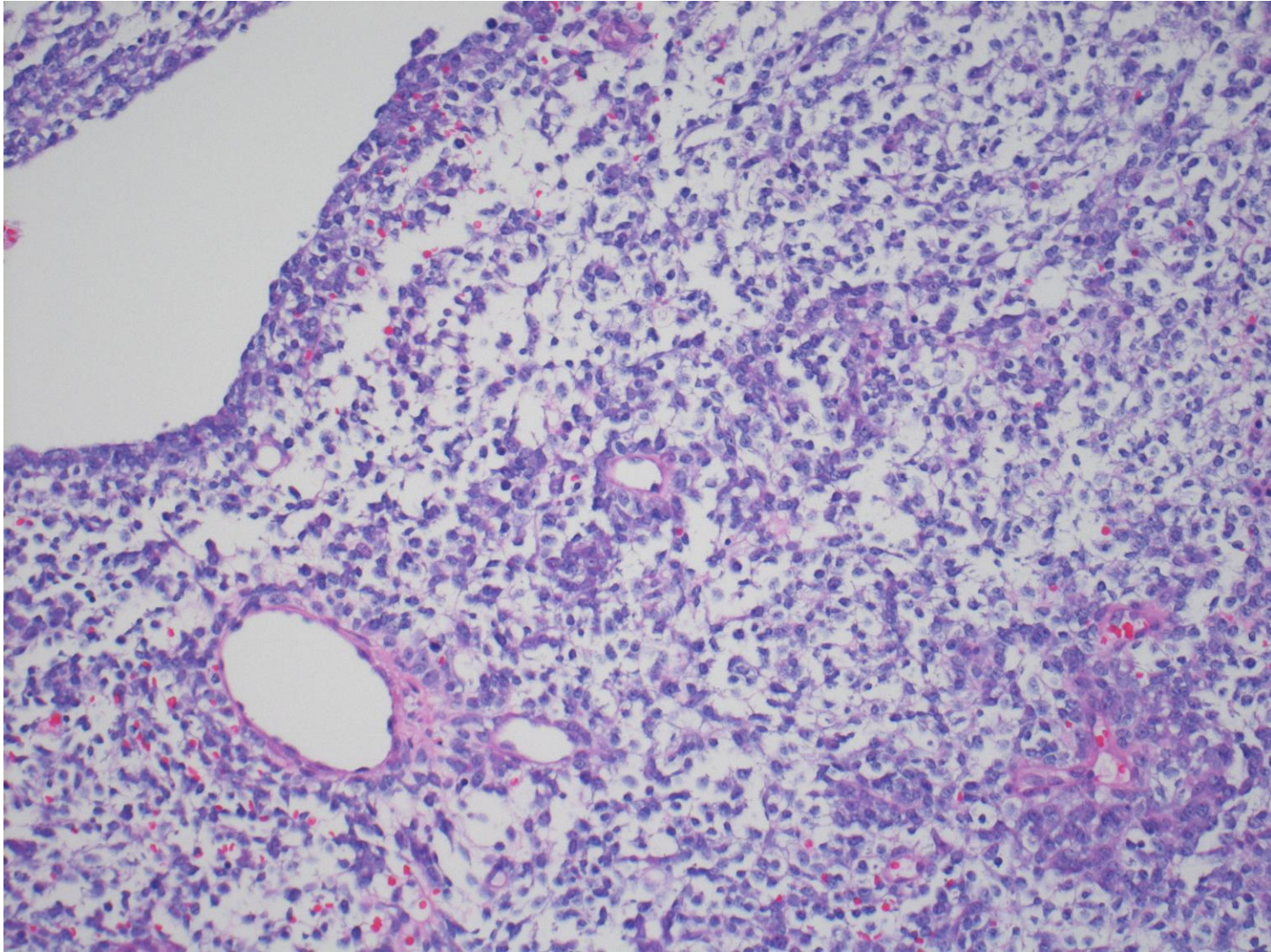
Microscopically, undifferentiated (Ewing-like) small round cells massively grow with multinodular fashion and sheet-like arrangement. Microcystic change is focally observed. Immunohistochemically, the tumor cells are positive for CD99 (MIC2), but negative for cytokeratins, EMA, alpha-SMA, desmin, myogenin, TLE1, S-100 protein, NSE, synaptophysin and LCA. By RT-PCR analysis, CIC-DUX4 fusion gene was detected, while EWS-FLI1/ERG fusion genes were negative.



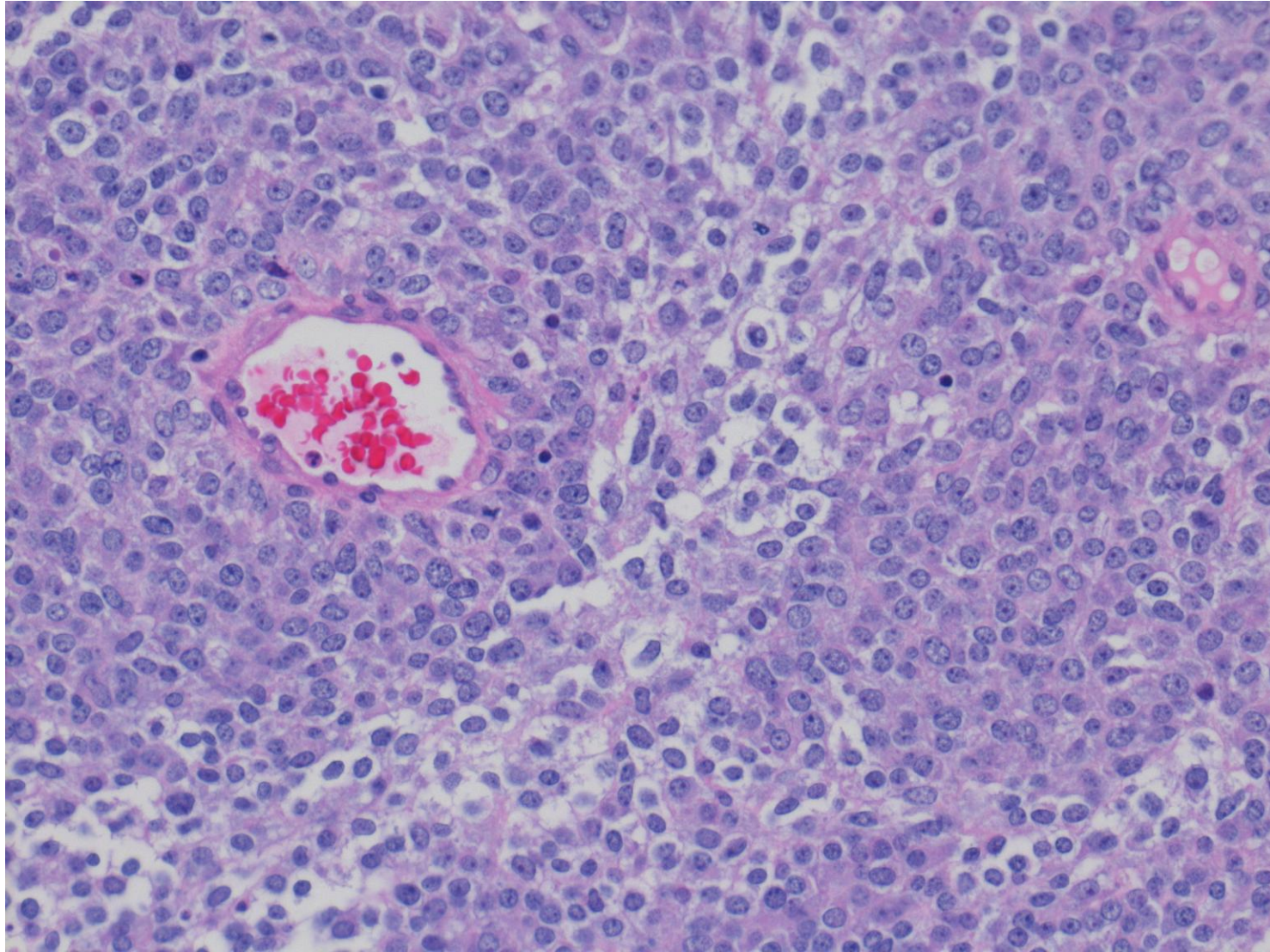
CIC-DUX4 sarcoma seen in the omentum of a 19-year-old female patient. Undifferentiated (Ewing-like) small round cells massively grow with a sheet-like arrangement. H&E-1



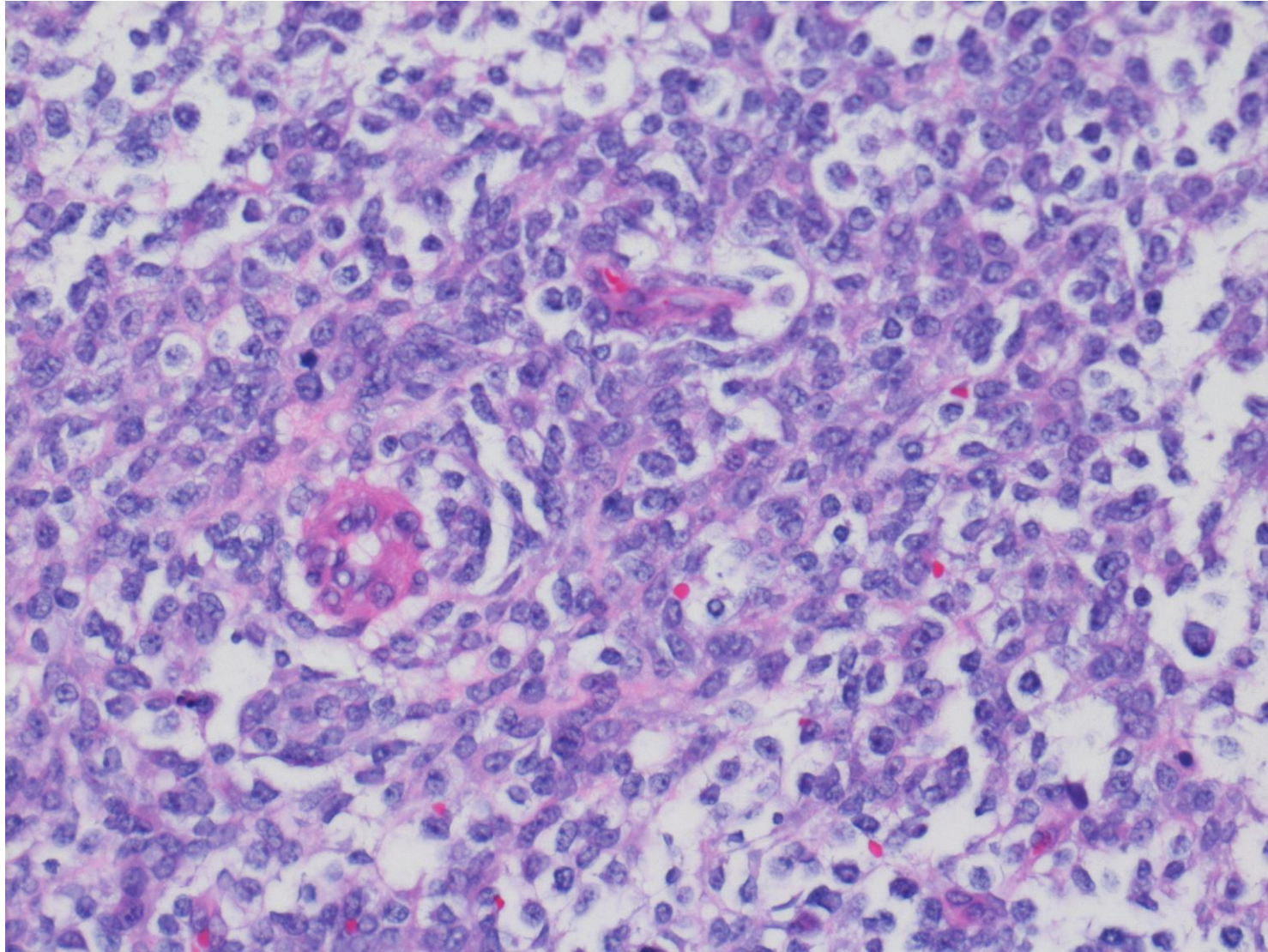
CIC-DUX4 sarcoma seen in the omentum of a 19-year-old female patient. Undifferentiated (Ewing-like) small round cells massively grow with a sheet-like arrangement and focal clustering. H&E-2



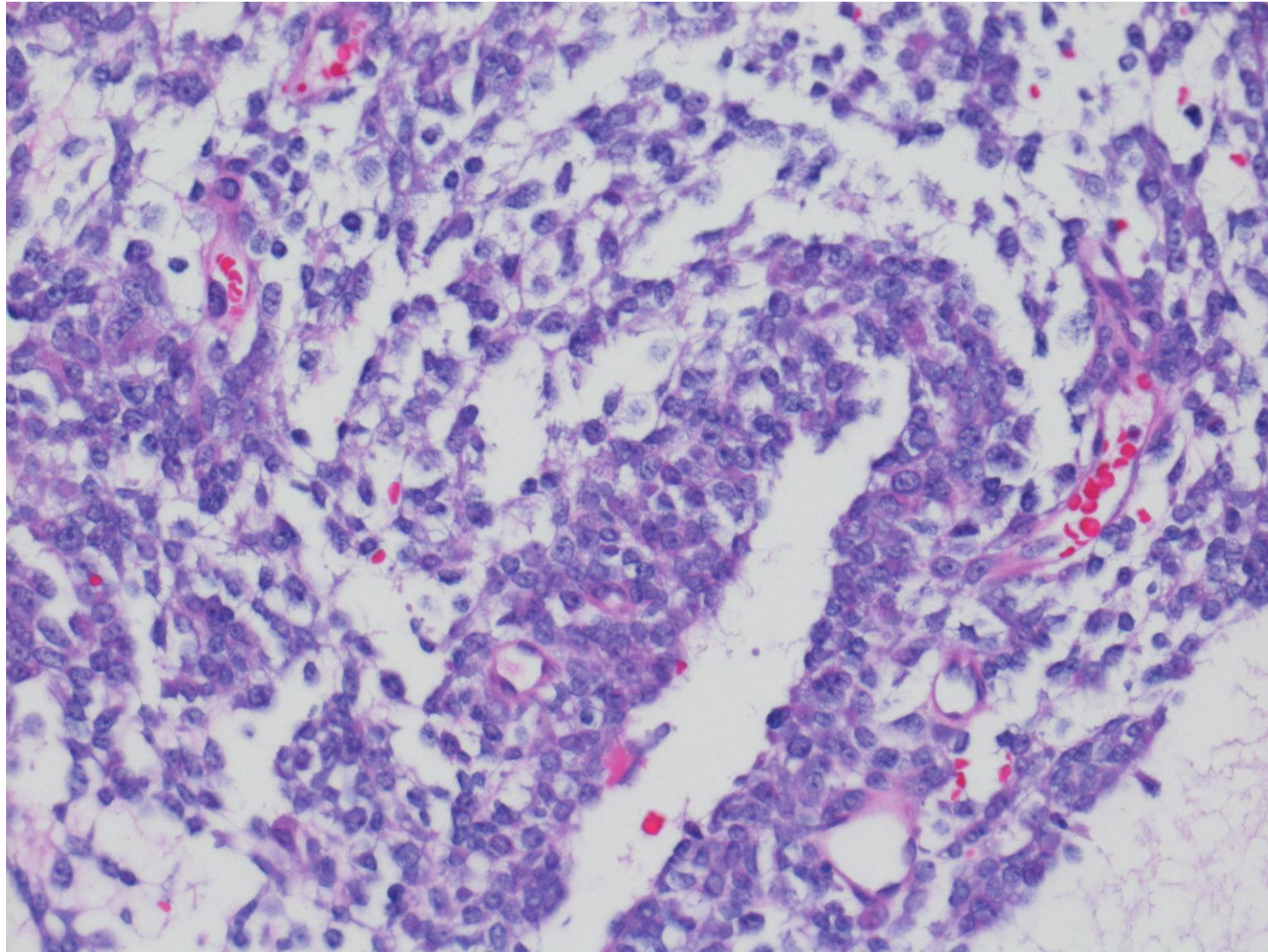
CIC-DUX4 sarcoma seen in the omentum of a 19-year-old female patient. Undifferentiated (Ewing-like) small round cells massively grow with a sheet-like arrangement and focal microcystic change. H&E-3



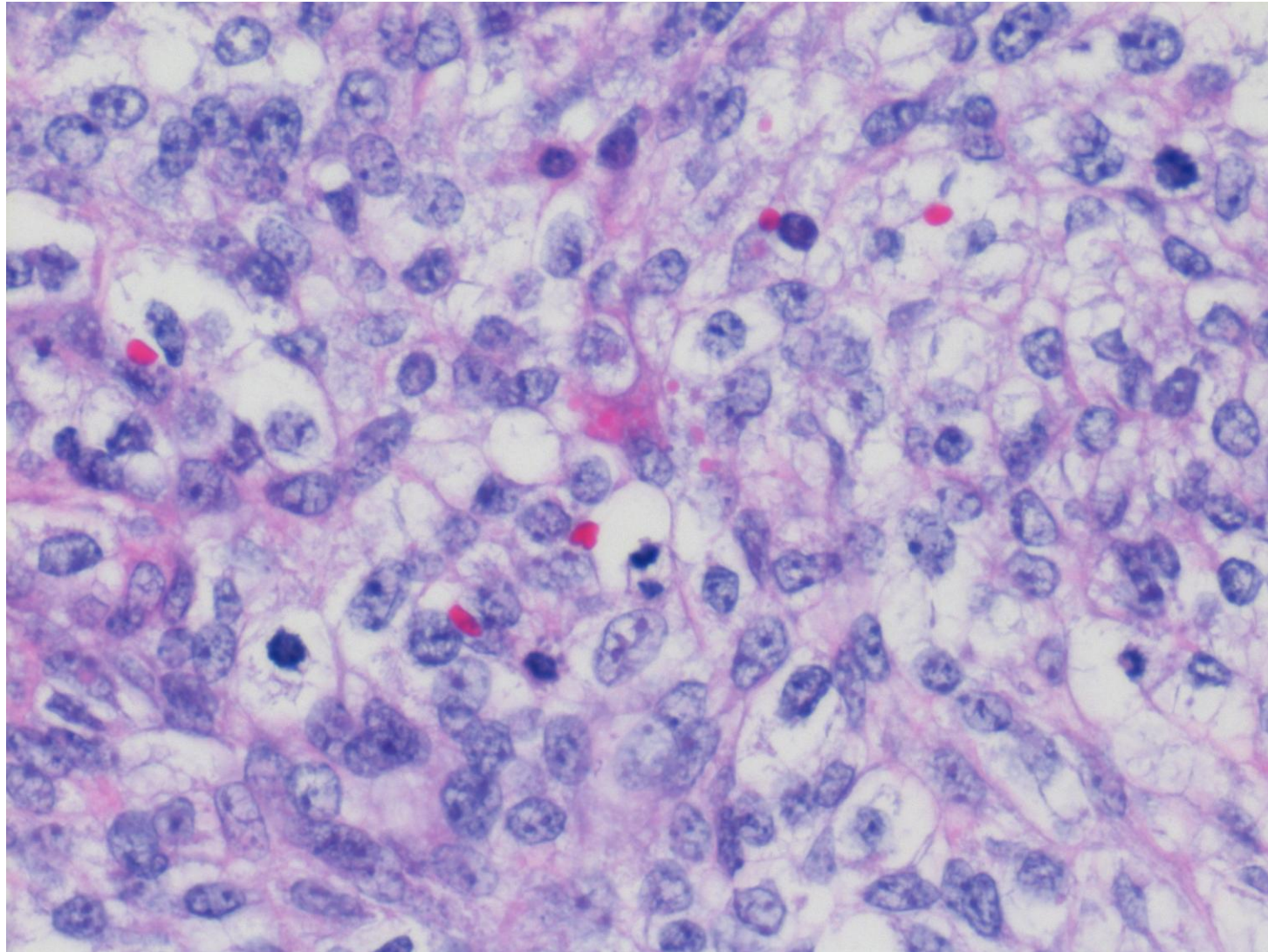
CIC-DUX4 sarcoma seen in the omentum of a 19-year-old female patient. Undifferentiated (Ewing-like) small round cells massively grow with a sheet-like arrangement. The cytoplasm of the tumor cells is clear or eosinophilic. H&E-4



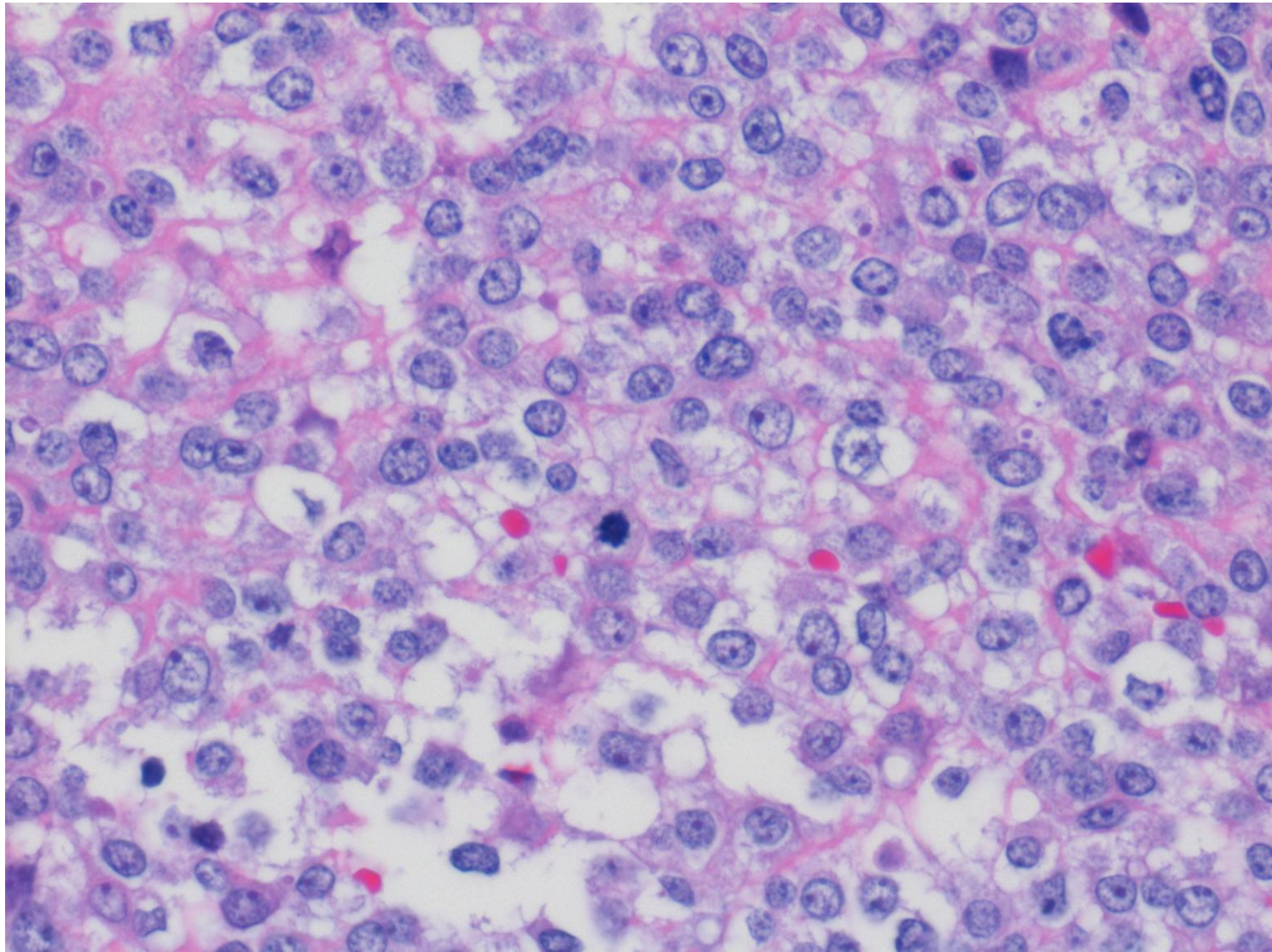
CIC-DUX4 sarcoma seen in the omentum of a 19-year-old female patient. Undifferentiated (Ewing-like) small round cells massively grow with a sheet-like arrangement. The cytoplasm of the tumor cells is clear or eosinophilic. H&E-5



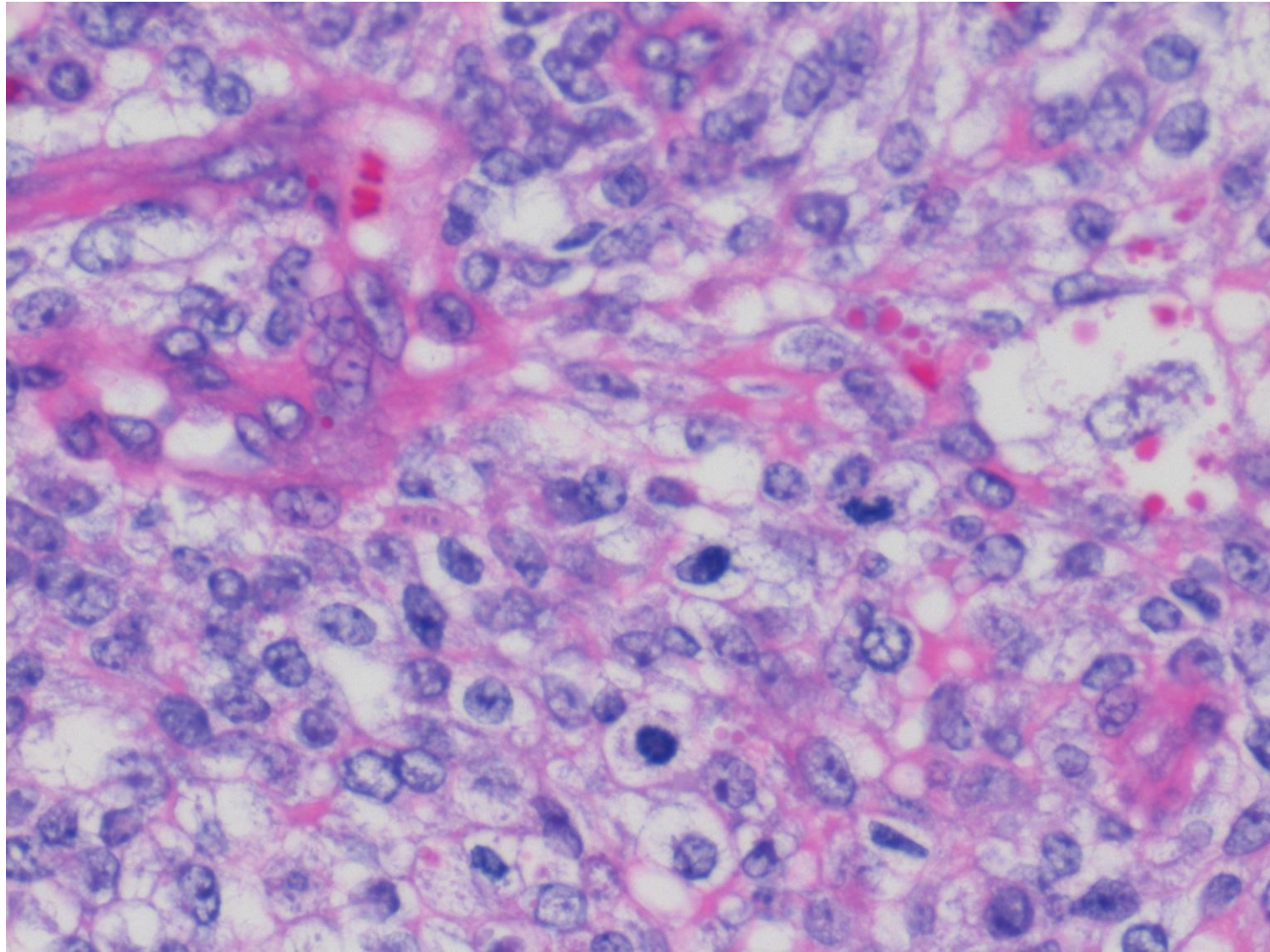
CIC-DUX4 sarcoma seen in the omentum of a 19-year-old female patient. Undifferentiated (Ewing-like) small round cells massively grow with a sheet-like arrangement. The tumor cells accumulate around microcystic spaces. H&E-6



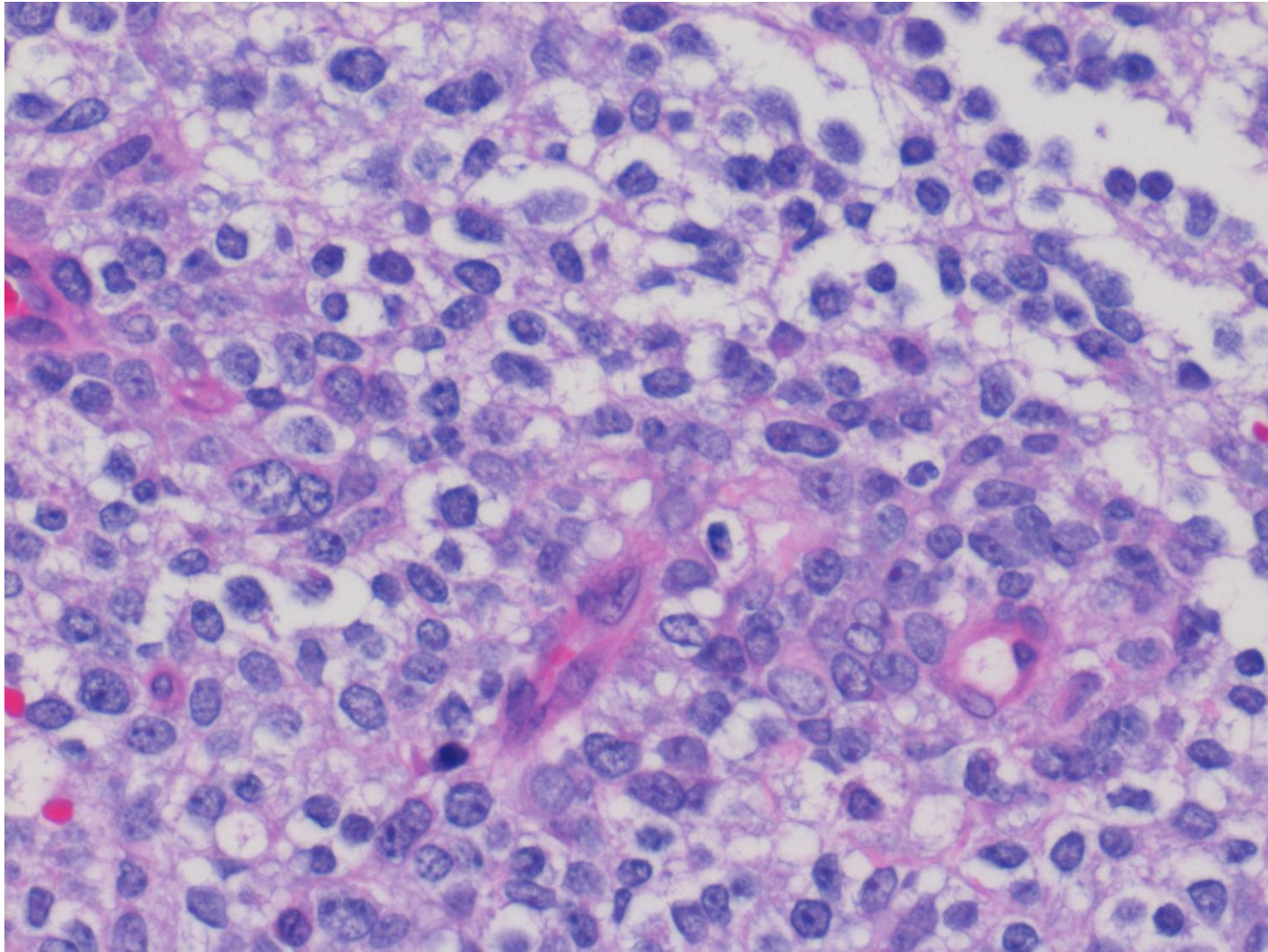
CIC-DUX4 sarcoma seen in the omentum of a 19-year-old female patient. Undifferentiated (Ewing-like) small round cells massively grow with a sheet-like arrangement. The cytoplasm of the tumor cells is clear or eosinophilic. Mitoses are scattered. H&E-7



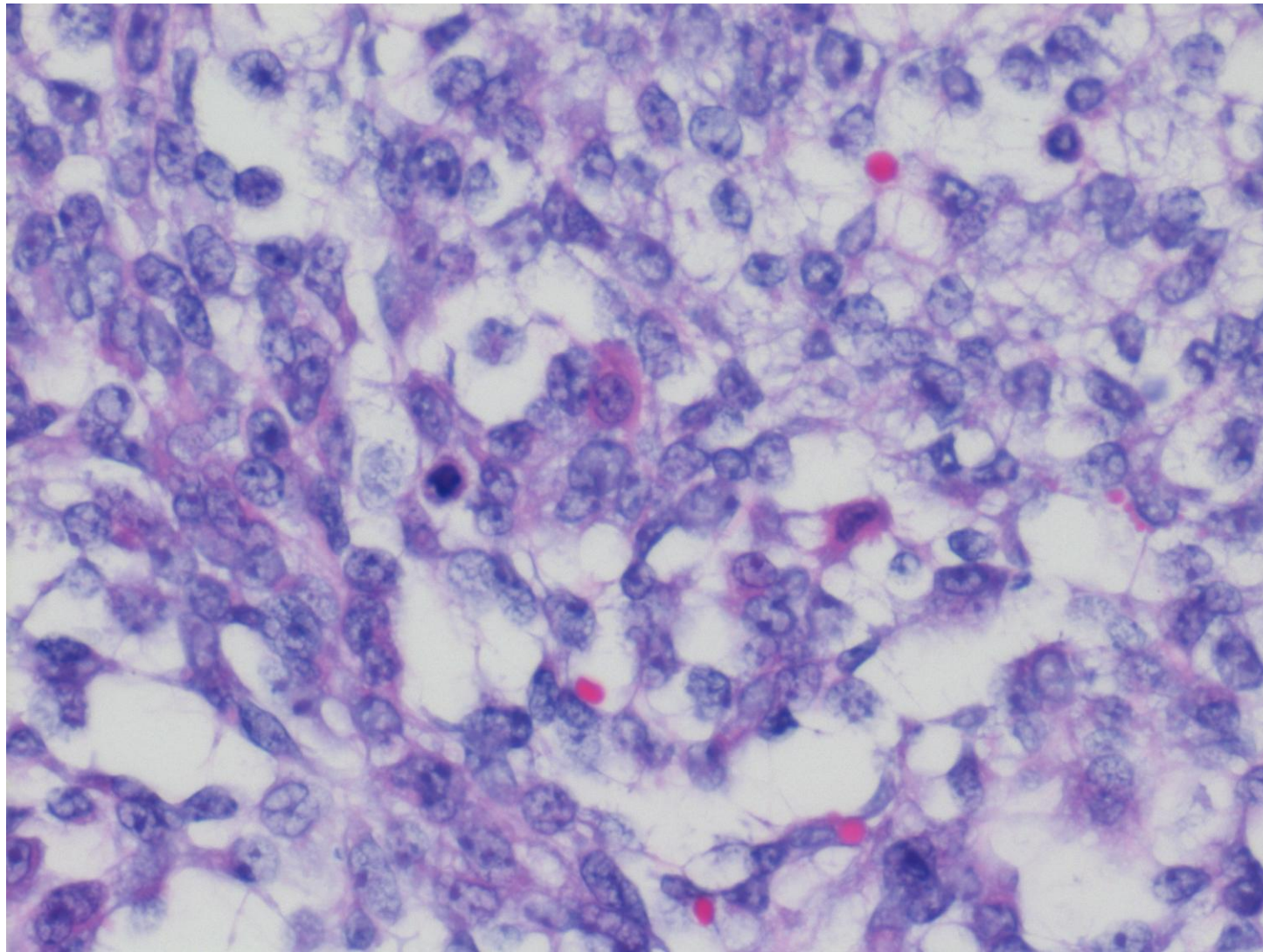
CIC-DUX4 sarcoma seen in the omentum of a 19-year-old female patient. Undifferentiated (Ewing-like) small round cells massively grow with a sheet-like arrangement. The cytoplasm of the tumor cells is clear or eosinophilic. Mitoses are scattered. H&E-8



CIC-DUX4 sarcoma seen in the omentum of a 19-year-old female patient. Undifferentiated (Ewing-like) small round cells massively grow with a sheet-like arrangement. The cytoplasm of the tumor cells is clear in this area. H&E-9



CIC-DUX4 sarcoma seen in the omentum of a 19-year-old female patient. Undifferentiated (Ewing-like) small round cells massively grow with a sheet-like arrangement. The cytoplasm of the tumor cells is clear in this area. H&E-10



CIC-DUX4 sarcoma seen in the omentum of a 19-year-old female patient. Undifferentiated (Ewing-like) small round cells massively grow with a sheet-like arrangement. The cytoplasm of the tumor cells is clear or eosinophilic. H&E-11