

2020



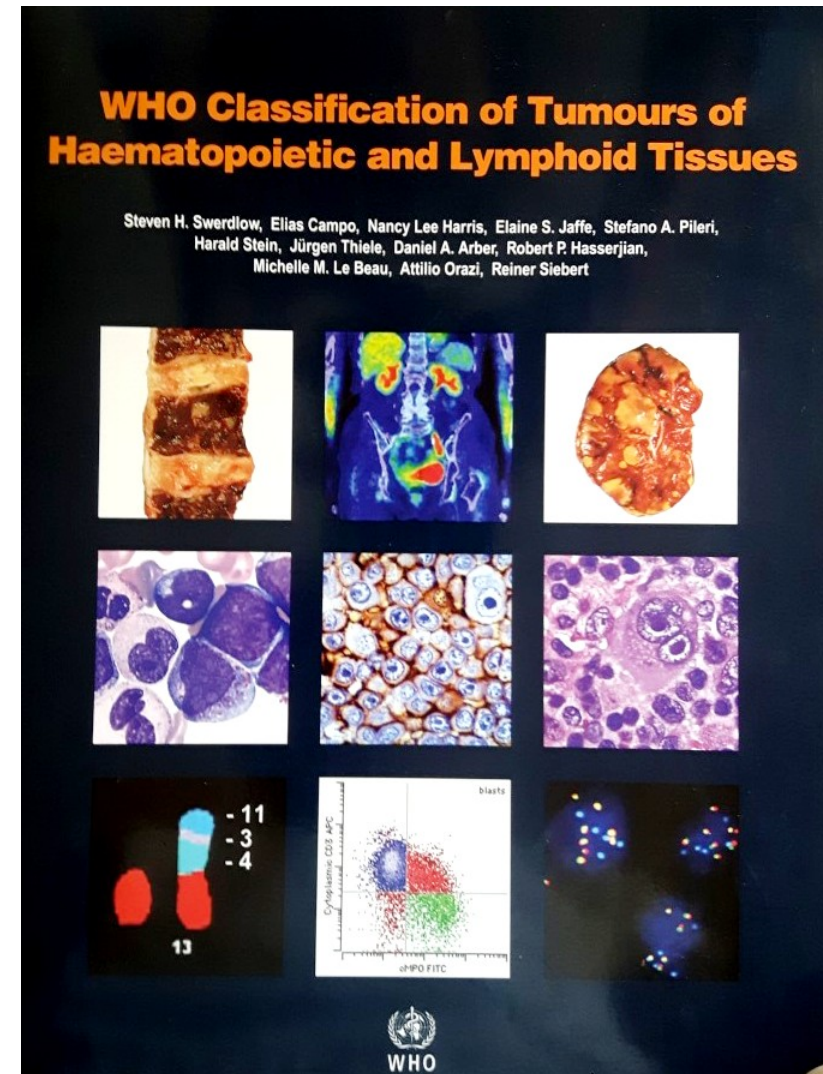
Progetto
Ematologia
Romagna

Sarcoma mieloide

Francesco Lanza – Ravenna

Myeloid sarcoma

A myeloid sarcoma **is a tumour mass consisting of myeloid blasts**, with or without maturation, **occurring at an anatomical site other than the bone marrow**. Infiltration of any site of the body by myeloid blasts in a patient with leukaemia **is not** classified as myeloid sarcoma unless it present with tumour masses in which the tissue is effaced.



Incidence: up to 2017: 2000 cases; >100 cases (2020 yr)

- AML 3200 cases per year in Italy
- 0.05% of AML
- **Primary forms.**
- It might present the same genetic lesions as classical AML (trisomy 8; KMT2a-MLLT3)
- Aggressive chemotherapy is highly recommended; radioterapy to control symptoms
- **Secondary to allo transplanted pts (5-8%)**

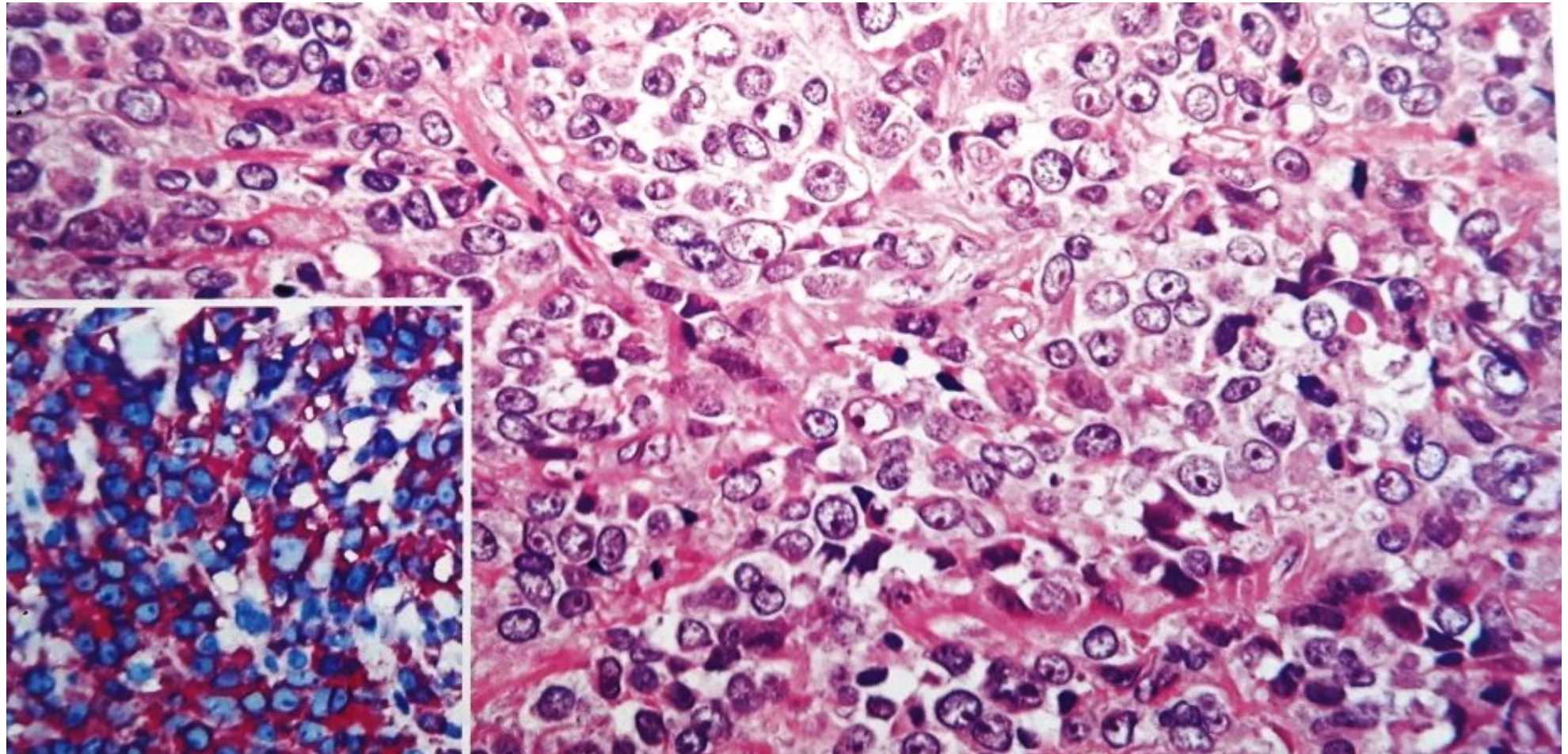


Fig. 1 Myeloid sarcoma. The tumour consists of blast with scant cytoplasm and round-oval nuclei with finely dispersed chromatin and minute but distinct nucleoli. Mitotic figures are numerous. Neoplastic cells strongly express MPO (insert)

32 yrs old pt seen at Ravenna Institute



Myeloid sarcoma

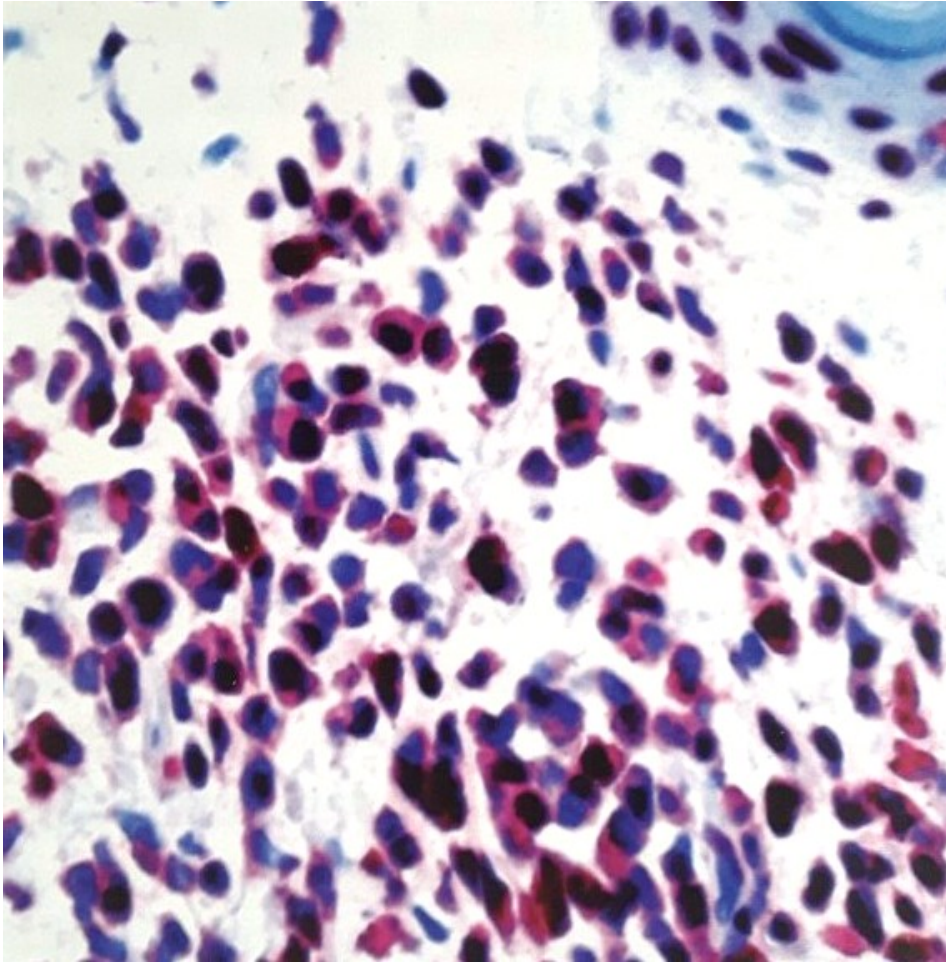


Fig. 2 Myeloid sarcoma. On skin biopsy with staining for NPM1, leukaemic cells infiltrating the derma show, in addition to the expected nuclear positivity, aberrant cytoplasmic expression of NPM1, which indicates the presence of *NPM1* mutation, cells of the overlying epidermis show a nuclear-restricted positivity for NPM1,

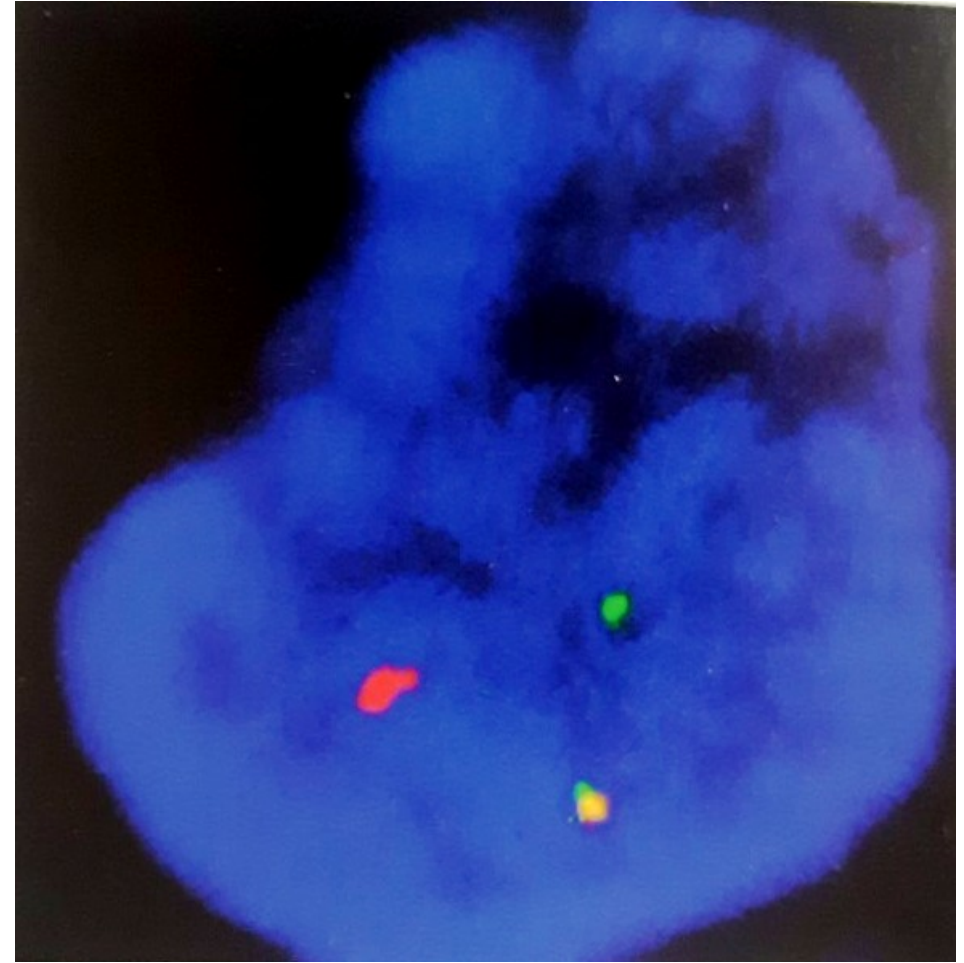


Fig. 3 Interphase FISH with *CBFβ* dual-colour break-apart rearrangement probe shows splitting of the gene: one red and one green signal appear separately in the nucleus, and the normal *CBFβ* allele appears as a fused red/green signal.

Pancreatic Myeloid Sarcoma

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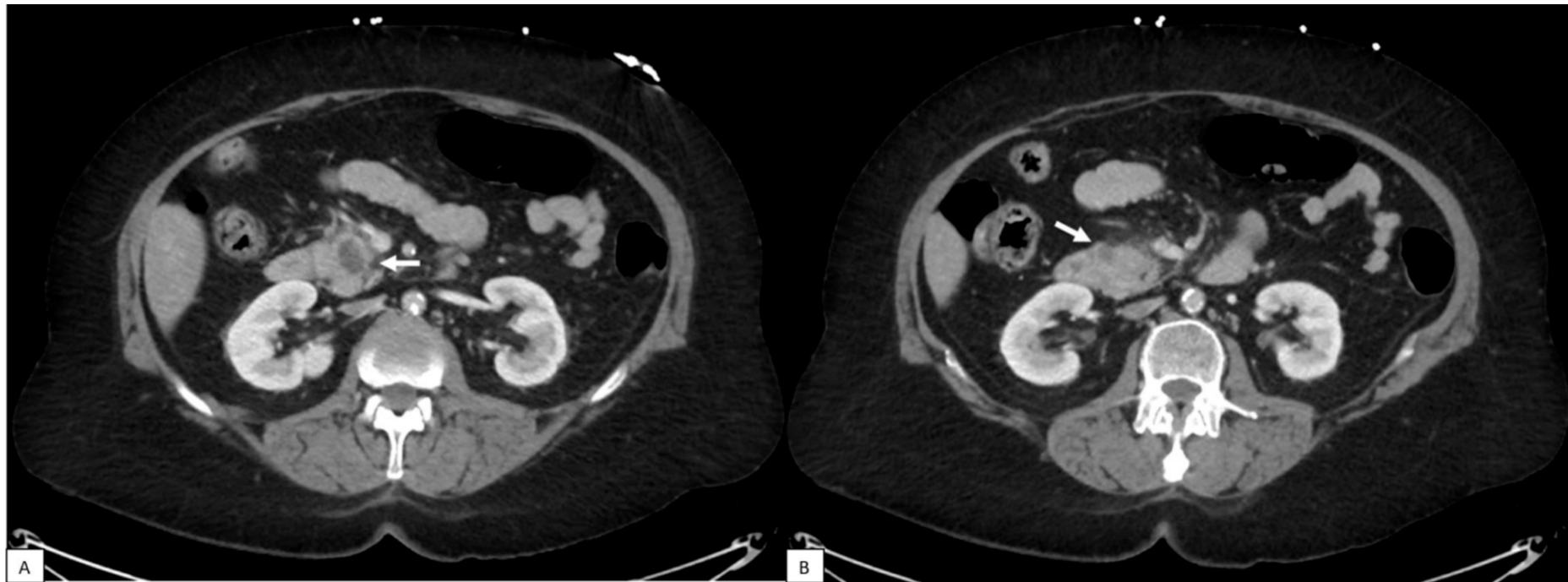


Fig. 4: Abdominopelvic imaging demonstrating a new pancreatic mass. (A) CT scans of the abdomen and pelvis with contrast shows a new ellipsoid area of hypoenhancement within the pancreatic head near the region of the ampulla that measures 2.2 x 1.5 cm. (B) A separate area of hypo-enhancement seen more laterally and inferiorly measures 1 cm. The pancreatic duct is not distended.

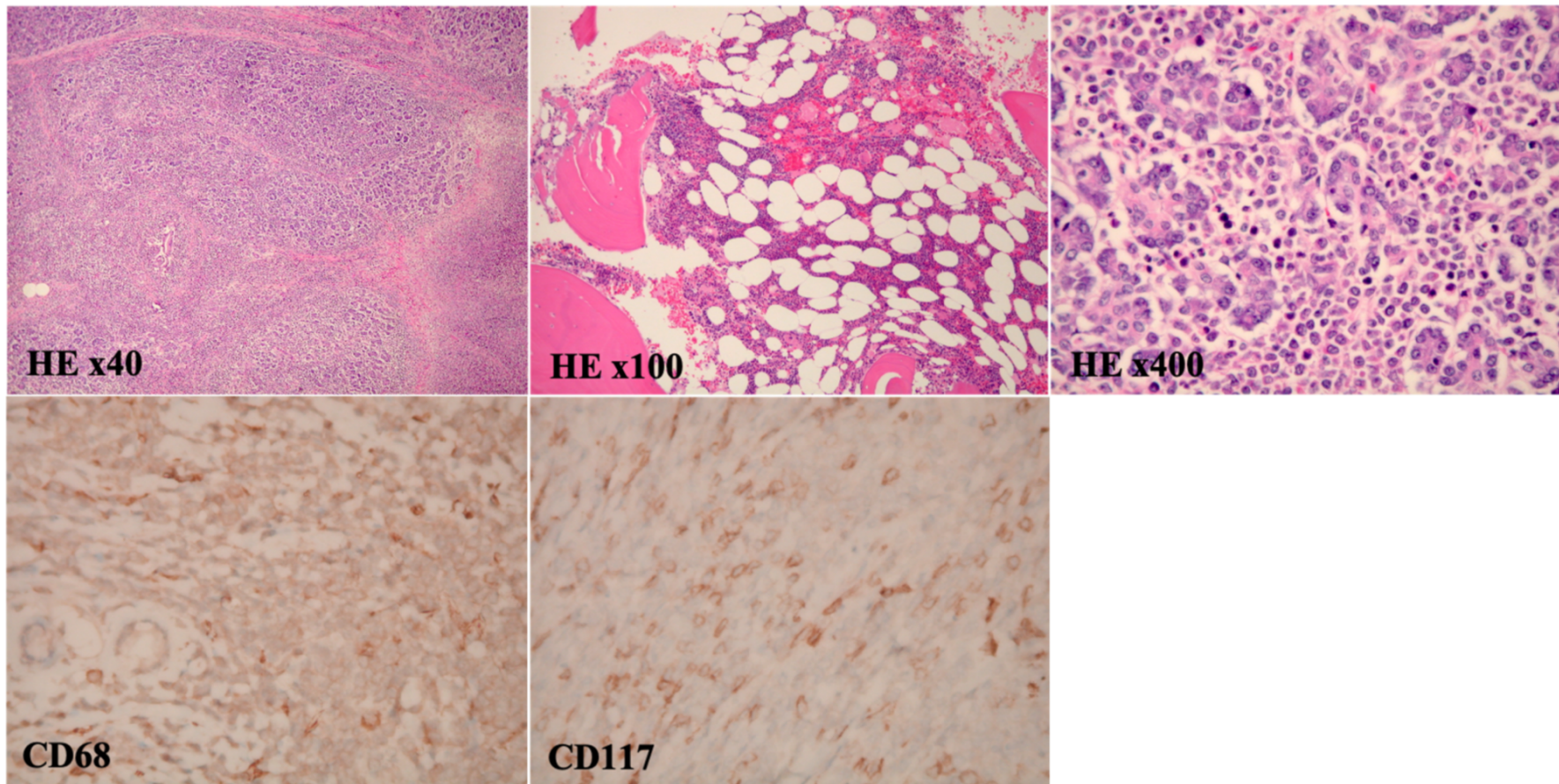


FIGURE 2: Pathology provided by Whipple resection shows a pancreatic myeloid sarcoma.

H&E staining of pancreatic tissue specimens shows an infiltrate comprised of diffuse sheets of atypical cells with scant cytoplasm, irregular nuclei, and prominent nucleoli. Myeloid blast cells infiltrate between and through benign pancreatic ducts and acini (H&E x40, x100, and x400). No lymph node involvement is present. The lesional cells show immunoreactivity with cytoplasmic CD68 and dimly with CD117 on immunohistochemical staining. Thus, aberrant B-cell and T-cell expressions are observed, which further supports the diagnosis of MS.

H&E, hematoxylin and eosin

Open Access Case
Report

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“Extramedullary Myeloid Sarcoma with Symptoms of Acute Pancreatitis Mimicking Pancreatic Carcinoma”

Sean Clausen, Mansi Oberoi, Douglas Lynch, George Maher, Timothy Ridgway, Muslim Atiq

Case Reports S D Med. 2020 Jul;73(7):305-307.

“Gastrointestinal Myeloid Sarcoma”

Trisha S. Pasricha, Diane Abraczinskas

Case Reports N Engl J Med. 2020 Aug

27;383(9):858. doi:

10.1056/NEJMicm2001235.

Therapy: standard AML induction therapy

“Venetoclax combined with azacitidine in the treatment of elderly patients with acute myeloid leukemia or myeloid sarcoma: Three cases reports and literature review”

F.Q. Tian, L.S. Zhang, J.H. Li, M.Q. Tang, J. Jiang, X.H. Cheng, X.C. Zhang, M. Jiang

PMID: 32942828 DOI: 10.3760/cma.j.issn.0253-2727.2020.08.016

Conclusioni

- **Patologia rara ma non rarissima**
- **escludere coinvolgimento periferico e midollare**
- **può nascere in qualunque parte del corpo**
- . **Terapia simile ad AML + SCT e RT per controllo sintomi: stretto follow-up**
- **Pochissimi studi clinici per testare ruolo nuovi farmaci**