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Retroperitoneal pararenal dedifferentiated liposarcoma with epithelioid/neuroendocrine features, somatostatin-positive.

Research Article

Gabriele Gaggero[1]*, Paola Bet[1], Davide Taietti[2], Marta Ingaliso[3], Bruno Spina[1]

[1]*IRCCS Ospedale Policlinico San Martino, UO Anatomia patologica ospedaliera, Genova, Italy*
[2]*Pathology Unit, ASST del Garda, Desenzano del Garda (Brescia), Brescia, Italy*
[3]*Università di Genova, Scuola di Scienze Mediche e Farmaceutiche, Department of Integrated Surgical and Diagnostic Sciences (DISC), Division of Anatomic Pathology, Genova, Italy*

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Abstract: Liposarcomas are malignant mesenchymal neoplasms that, although derived from adipocytes, may dedifferentiate to other cell types. We present an exceedingly rare case of epithelioid/neuroendocrine dedifferentiation with somatostatin expression of a pararenal retroperitoneal liposarcoma.
Based on the immunomorphological features, the diagnosis was possible with the support of the molecular FISH investigation: it showed amplification of the MDM2 gene, which is present in most liposarcomas, even in dedifferentiated ones.

Keywords: *liposarcoma • dedifferentiated • epithelioid • neuroendocrine • somatostatin • MDM2*

Dear Editor-in-Chief

We have read with interest the article entitled 'Gastrointestinal Neuroendocrine Tumours: A Single-Centre Experience', published in Forum of Clinical Oncology[1].

We would like to point out, however, that not all intra-abdominal lesions with neuroendocrine phenotypic expression are gastro-entero-hepato-pancreatic neuroendocrine epithelial neoplasms.

We present the case of a woman in her sixties with a pararenal mass 12 cm in diameter, radiologically doubtful as to being of renal origin or from the pancreatic tail: surgery revealed that this mass was extra-renal and extra-pancreatic, thus sparing both organs.

Histology showed an epithelioid nested neoplasia, with necrosis and several mitoses (Fig. 1A); notably, spindle cell and adipocytic areas with cytological atypia are observed on the surrounding border (Fig. 1B). Immunohistochemistry results were the following: cytokeratins-; chromogranin-; synaptophysin-; NSE+ (Fig. 1C); CD56+; somatostatin+ (Fig. 1D); insulin-; glucagon-; gastrin-; serotonin-; pancreatic polypeptide-; pS100-; SOX10-; melan A-; HMB45-; vimentin-/+; CD31-; CD34-; FLI1-; desmin-; smooth muscle actin -/+; muscle specific actin-; CD117-; DOG1-; inhibin-; MUC1-; tyrosinase-; GATA3-; CD99-; BCL2+; WT1-.

The findings led us to a malignant neoplasm with neuroendocrine expression (NSE, somatostatin, CD56), whose true nature was, however, difficult to define: the epithelioid morphology found does not allow us to define an epithelial origin; furthermore, neuroendocrine tumours are cytokeratin-negative only rarely.

The following were considered in the differential diagnosis: pancreatic or gastro-enteric neuroendocrine tumour, paraganglioma, adrenal carcinoma, GIST, PEComa, schwannoma[2], and melanoma, but they are excluded by immunohistochemistry results.

* *E-mail: gabriele.gaggero@hsanmartino.it*

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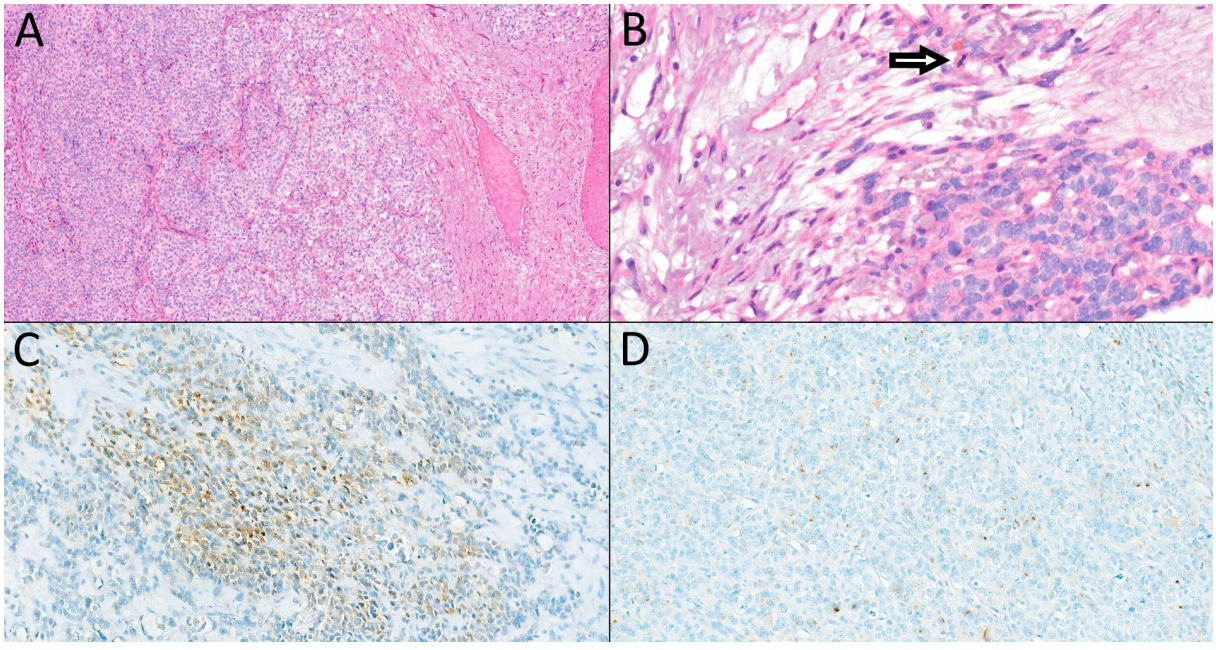


Figure 1: Photomicrographs of liposarcoma with epithelioid/neuroendocrine dedifferentiation: A) histological view of a neoplasm organised in nests separated by thin fibrovascular branches, cytologically of epithelioid appearance (Hematoxilin and Eosin, 20x); B) at higher magnification, spindle/elongated neoplastic elements, also with mitosis (arrow), are observed at the edges of the epithelioid neoplasm, suggesting a possible mesenchymal origin (Hematoxilin and Eosin, 40x); C) immunohistochemistry for NSE, showing positivity in scattered neoplastic cells (10x); D) immunohistochemistry for somatostatin, showing ‹dot like› positivity in epithelioid neoplastic cells (10x).

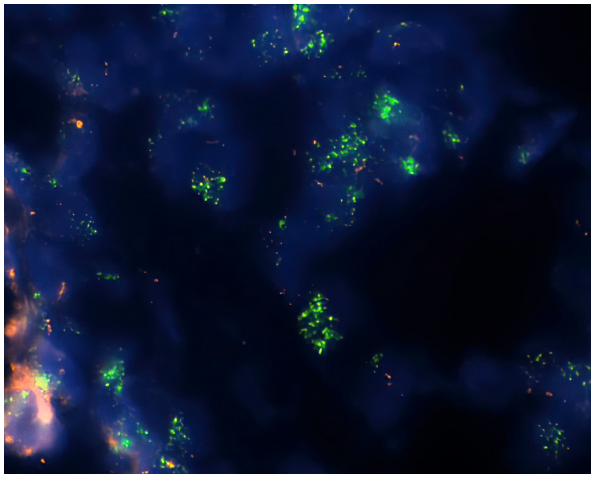


Figure 2: FISH photomicrograph showing a liposarcoma tissue section with amplification of the MDM2 gene (green signal), compared to the control CEN 12 (orange signal).

The positivities for vimentin and actin, although in rare neoplastic elements, led to the suspicion of a mesenchymal nature, and the atypical features in the surrounding fibroadipose tissue reinforced this hypothesis: we therefore proceeded with the assessment of *MDM2* gene amplification by *FISH*. The result was: amplified (Fig. 2).

Pararenal malignancies may originate from the renal capsule or the adrenal gland[3,4]; however, more frequently, they arise from the retroperitoneal soft tissues: the most frequent is liposarcoma. Such a neoplasm derives from adipocytes but can dedifferentiate to many histological lines: the epithelioid/neuroendocrine is one of the rarest[5]. In any case, the vast majority of liposarcomas, even dedifferentiated ones, show amplification of the MDM2 gene.

It is therefore thanks to the above FISH result that it was possible to label this lesion as a liposarcoma, although exceptionally rare in this form of epithelioid/neuroendocrine dedifferentiation with somatostatin expression[6].

Abbreviations

BCL2- B-cell lymphoma 2
CD- cluster of differentiation
CEN 12- centromeric region in chromosome 12
DOG1- discovered on GIST type 1
FISH- fluorescence in situ hybridization
FLI1- friend leukemia virus integration 1
GATA3- GATA Binding Protein 3
GIST- gastrointestinal stromal tumors

HMB45- human melanoma black 45
MDM2- mouse double minute 2 homolog
Melan A- melanoma antigen recognized by T cells 1
MUC1- transmembrane glycoprotein mucin 1
NSE- neuron specific enolase
PEComa- perivascular epithelioid cell tumor
SOX10- sry-related hmg-box gene 10
WT1- Wilms' tumor suppressor gene1

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