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IMAGE OF THE MONTH

Distinct [^{18}F]FDG-PET imaging features of a newly recognized and yet uncharacterized RDD-ECD overlap disease entity

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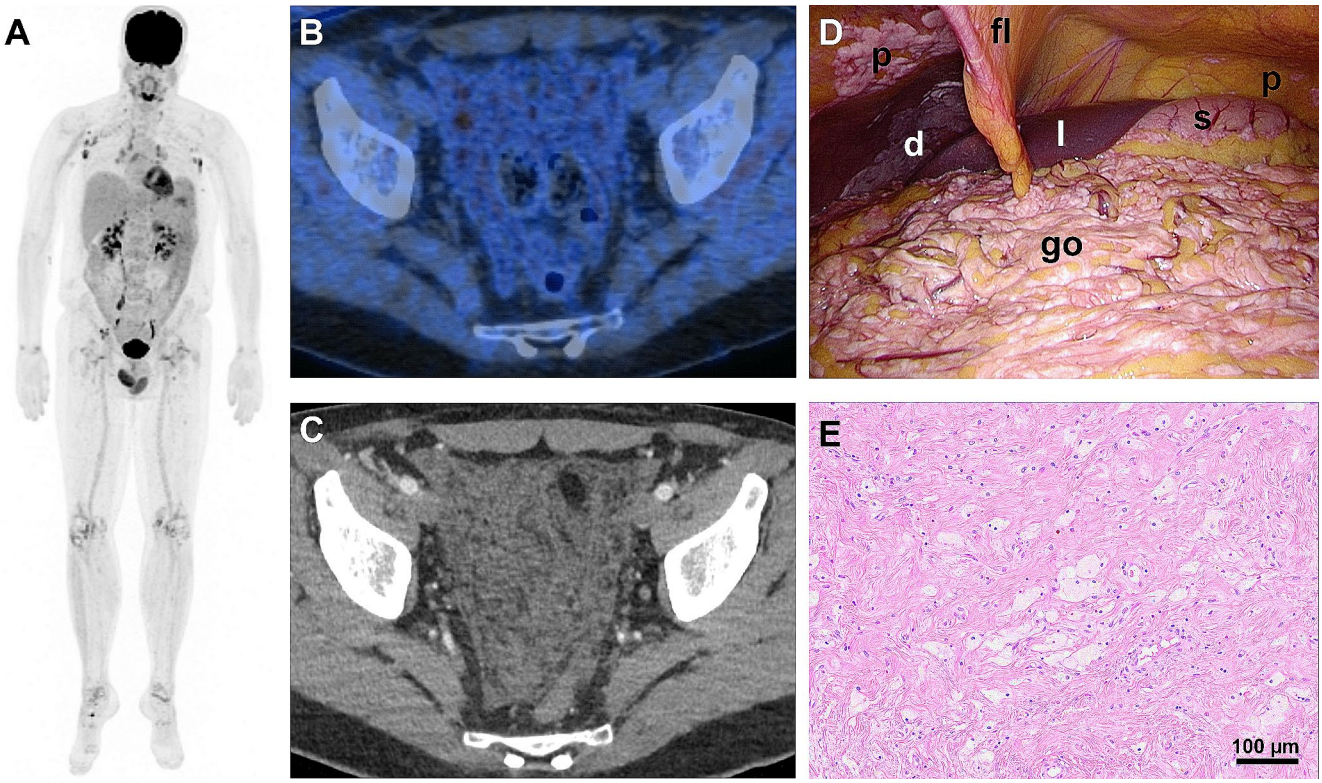

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A newly recognized histiocytosis entity, encompassing clinical and histopathologic features of Rosai-Dorfman disease (RDD) and Erdheim-Chester disease (ECD), is driven by MAP2K1 mutations [1, 2]. [^{18}F]fluorodeoxyglucose ([^{18}F]FDG) positron emission tomography (PET) features have not yet been reported.

This 46 year-old man presented with a two-year history of clinical hallmarks resembling RDD rather than ECD, including lymphadenopathy and painless testicle enlargement [3], being also visible on [^{18}F]FDG-PET (**A**). Testicular RDD-ECD involvement was also reported in 6/13 patients by Razanamahery et al. [2]. Diffuse omental proliferations,

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manifesting as faintly [^{18}F]FDG-avid omental thickening resembling a fishing net (SUV_{max} 5.5; **A, B, C**), and symmetric large-joint synovitis were reported as specific features of RDD-ECD [1, 2], Notably, none of these features are characteristic of RDD or hitherto known ECD subtypes. Other RDD and/or ECD features were absent [4–7].

Open biopsy targeted peritoneal lesions (**D**) localized on the diaphragm (d), peritoneum (p) and greater omentum (go). Histopathology revealed nodular fibrosis, foamy cell infiltrates, pigment deposits and chronic perivascular inflammatory infiltrates (**E**). Molecular genetic analyses confirmed presence of a characteristic MAP2K1 mutation (p.Q56P).

Diamond et al. effectively treated a patient harboring the identical mutation with MEK inhibitors [8]. FAPI-PET focusing on fibrosis aspects of histiocytosis might help determining disease extent and assessing treatment response [9, 10].

In summary, the newly recognized RDD-ECD overlap histiocytosis demonstrates distinct [^{18}F]FDG-PET features setting it apart from RDD and ECD. The concurrent presence of omental proliferations, symmetric large-joint synovitis, and high testicular uptake should raise suspicion for this yet uncharacterized disease.

Author contributions All authors contributed to the conception and design. Image analysis / image compilation and design of the final image were performed by M.H. The first draft of the manuscript was written by M.H: and all authors commented on previous versions of the manuscript. All authors read and approved the final manuscript.

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Declarations

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