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【 PICTURES IN CLINICAL MEDICINE 】

Erdheim-Chester Disease

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Key words: non-Langerhans histiocytosis, primary medicine

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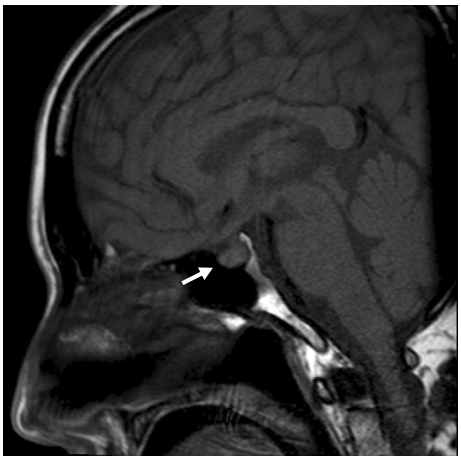
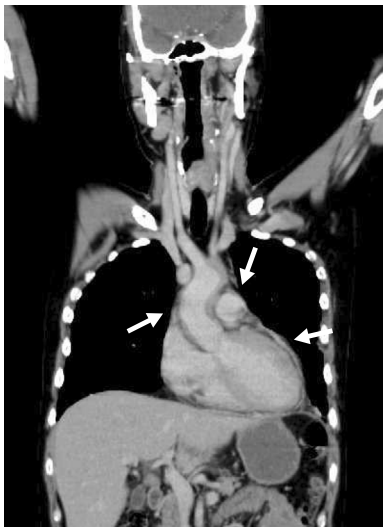
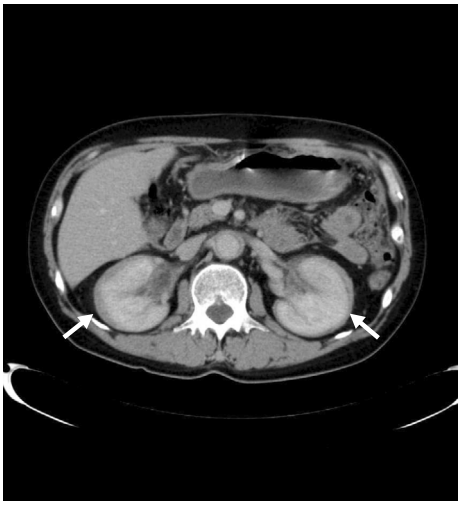


Picture 1.



Picture 2.



Picture 3.

A 51-year-old man presented with a 1-month history of fatigue and shortness of breath. His medical history included central diabetes insipidus with lymphocytic hypophysitis (Picture 1) diagnosed eight months earlier. A physical examination and laboratory tests revealed no significant abnormalities. Contrast-enhanced computed tomography revealed pleural thickening, pericardial fibrosis, a circumferential periaortic lesion called "coated aorta" (Picture 2), and rind-like infiltration surrounding the kidneys, also known as "hairy kidney" (Picture 3). Skin and pleural tissue biopsies showed foamy histiocytes with surrounding fibrosis [CD68(+), factor 13a (+), CD1a (-), and S-100 (-) ; Picture 4]. A diagnosis of Erdheim-Chester disease (ECD), a rare non-Langerhans histiocytosis, was made based on the histopathological findings and clinical and radiologic features. Bone and skin lesions were absent. The BRAF mutation was negative. We did not check for the NRAS mutation. He was treated with interferon-alpha, and his symptoms improved. ECD has multiorgan manifestations, including diabetes insipidus. Radiological imaging findings are unique and useful for the di-

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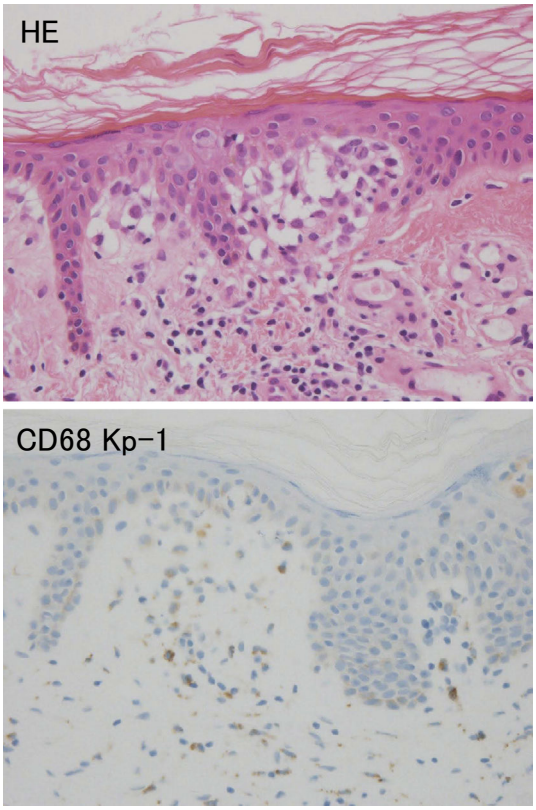

Picture 4.

agnosis of ECD (1, 2).

The authors state that they have no Conflict of Interest (COI).

References

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