Symmetric giant xanthogranulomas in Erdheim-Chester disease

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A 33-year-old woman admitted to our hospital presenting with polyarthralgia, diabetes insipidus and xanthogranulomas in a subcutaneous precordial region and long bones was histologically diagnosed with Erdheim-Chester disease (ECD) [1]. Despite 5 years of treatment with prednisolone, cyclophosphamide and imatinib mesylate after the original diagnosis, subcutaneous xanthogranulomas again developed. ^{18}F -FDG PET/CT revealed symmetric xanthogranulomas in the retroauricular regions, axillae, in tissue surrounding the psoas major muscle and in the long bones (arrows). These were histologically and immunologically confirmed as being consistent with ECD. Symmetric lesions of the long bones have generally been observed in ECD patients [2, 3]; however, our PET/CT images clearly showed symmetric cutaneous xanthogranulomas. Such symmetric anatomical changes, especially at tendon attachments or in connective tissue, may be an important clue to clarifying the aetiology of ECD.

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