The atoll sign

A man presented as an urgent referral to the interstitial lung disease outpatient clinic with a 5 week history of fever, night sweats, shortness of breath and malaise. His admission chest x-ray showed patchy consolidation within both lungs, and a subsequent CT demonstrated multiple parenchymal ring opacities (figure 1 and figure 2A). On the basis of the unusual CT appearances he subsequently proceeded to an open lung biopsy which demonstrated polypoid buds of intra-alveolar granulation tissue or Masson bodies, consistent with a histopathological diagnosis of organising pneumonia (figure 3). He was treated with intravenous methylprednisolone and hydroxychloroquine to which he initially responded. Over the next 5 years he had several relapses clinically and radiologically which required pulse doses of methylprednisolone. Once stable, his medication was tapered and eventually stopped. Repeat CT imaging at this time revealed no residual parenchymal abnormalities. No definite cause for his illness was found and, following a multidisciplinary evaluation of his case, a final diagnosis of cryptogenic organising pneumonia was made.

Crescentic or ring-shaped opacities surrounding an area of ground-glass opacification occasionally seen in the setting of organising pneumonia were first described by Voloudaki and later coined the ‘Atoll sign’ by Zompatori. Kim et al. documented it in 19% of cases of cryptogenic organising pneumonia. The word Atoll is derived from the Maldivian word ‘atholhu’ meaning an island consisting of a circular coral reef surrounding a lagoon (figure 2B). Although originally thought to be specific for organising pneumonia, this CT appearance has been described in a variety of other diseases including sarcoidosis, Wegener granulomatosis, lymphomatoid granulomatosis, pulmonary paracoccidiomycosis and non-specific interstitial pneumonia, and under different names including the ‘reversed-halo’ sign and the ‘fairy-ring’ sign. Histopathologically the ground-glass opacification centrally represents septal inflammation, and cellular debris within the airspaces and the peripheral crescentic or ring-shaped opacity represents organising pneumonia in the alveolar ducts. From a learning perspective, although this striking CT appearance is well documented in organising pneumonia, it should alert the radiologist and physician to other possible diagnoses, of which Wegener granulomatosis is particularly important.

Learning points

► The Atoll sign is a well recognised CT pattern associated with organising pneumonia.
► This sign has also been reported in a number of other important conditions which should be considered in the differential diagnosis when interpreting it. In particular, consideration should be given to Wegener granulomatosis.

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