Occurrence of giant focal forms of congenital hyperinsulinism with incorrect visualization by (18) F DOPA-PET/CT scanning.

Abstract:

Congenital hyperinsulinism (CHI) is a rare disease characterized by severe hypoglycaemic episodes due to pathologically increased insulin secretion from the pancreatic beta cells. When untreated, CHI might result in irreversible brain damage and death. Currently, two major subtypes of CHI are known: a focal form, associated with local distribution of affected beta cells and a nonfocal form, affecting every single beta cell. The identification of focal forms is important, as the patients can be cured by limited surgery. (18) F DOPA-PET/CT is an established non-invasive approach to differentiate focal from nonfocal CHI. The purpose of this study was to identify possible limitations of (18) F DOPA-PET/CT scan in patients with focal forms nonfocal CHI. A retrospective chart review of 32 patients (from 2008 through 2013) who underwent (18) F DOPA-PET/CT and partial pancreatectomy for focal CHI at the reference centres in Berlin, Germany and London, UK. In most cases (n = 29, 90.7%), (18) F DOPA-PET/CT was sufficient to localize the complete focal lesion. However, in some patients (n = 3, 9.3%), (18) F DOPA-PET/CT wrongly visualized only a small portion of the focal lesion. In this group of patients, a
so-called 'giant focus' was detected in histopathological analysis during the surgery. Our data show that in most patients with focal CHI (18) F DOPA-PET/CT correctly predicts the size and anatomical localisation of the lesion. However, in those patients with a 'giant focal' lesion (18) F DOPA-PET/CT is unreliable for correct identification of 'giant focus' cases.