Invasive Fungal Pneumatisi and Carotiditis (IFPAC) Syndrome in Immunocompromised Hosts: An Unrecognized, Often Catastrophic Invasive Fungal Disease (IFD)

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Infections involving the petrous portion of the temporal bone are rare and primarily caused by Pseudomonas aeruginosa. This presentation can occur in the setting of progressive malignant otitis externa (skull base osteomyelitis) or sinus disease.

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Background

Local invasion from the external auditory canal or sinus can result in inflammation and disseminated infection of vessels, with a particular pre-disposition to involvement of the internal carotid artery due to its course through the skull base.

Methods

We conducted a retrospective study of patients with fungal skull base infections and carotitis at our institution between 2003 and 2018. We collected data including demographics, presentation, imaging, diagnosis, treatment, microbiology, and outcomes on patients diagnosed with IFPAC.

We conducted a search of the PUBMED database. All cases in adults with imaging, pathology, or autopsy findings of skull base infection and carotiditis at our institution between 2003 and 2018.

Results

We identified 4 cases of IFPAC at our institution from 2003 to 2018. Median age at presentation was 73 years (range, 66-79). Three were male.

IFD risk factors included diabetes (n=3), glucocorticoid use (n=3), and lymphoid malignancy (n=2). Ten patients were on T-cell immunosuppressants.

All cases were caused by Aspergillus spp. Two cases were otogenic, two were sinusosal in origin. Two patients developed cranial nerve deficits (III, V, VI).

Median time from symptom onset to diagnosis was 17 days (range, 6-36). Two patients were treated with anti-Aspergillus antigens with initial improvement in symptoms, imaging, or decrease in serum galactomannan levels.

All patients subsequently presented with occlusion of the internal carotid artery (ICA) and multiple cerebral infaracts. Two patients developed mycotic aneurysms.

References


