

Material for Online Repository
 JACI Submission
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Table 1. Cases and literature review of HIES patients with endemic fungal infections.

Case [Reference]	Age*/gender/race	STAT3 mutation (domain)	Endemic mycosis	Pulmonary disease	Extra pulmonary disease	Therapy **	Complications
H1	10/M/W	V432M (DNA binding)	<i>Histoplasma</i>	Y	Assumed dissemination to liver and spleen	Liposomal amphotericin, itraconazole, posaconazole	Renal insufficiency from amphotericin
H2	15/F/W	F621V (SH2)	<i>Histoplasma</i>	N	Gastrointestinal	Liposomal amphotericin, itraconazole	Duodenal stricture, renal insufficiency from amphotericin
H3	22/F/W	W479C (DNA binding)	<i>Histoplasma</i>	N	Laryngeal	Ketoconazole	Reconstructive laryngoplasty
H4[E1]	4/F/ND	ND	<i>Histoplasma</i>	N	Rectal	Unknown	ND
H5[E2]	14/F/ND	ND	<i>Histoplasma</i>	N	Ileocecal	Itraconazole (12 mo)	ND
H6[E3]	27/F/B	ND	<i>Histoplasma</i>	N	Cecum, colon, bone marrow	Amphotericin B (ND), ketoconazole (2 mo)	ND
H7 [E4]	16/M/ND	ND	<i>Histoplasma</i>	N	Ileum, cecum, regional lymph nodes	Ketoconazole (6 mo)	Misdiagnosis as Crohn's disease
H8[E5]	2/M/ND	K591M (SH2)	<i>Histoplasma</i>	Y	Disseminated	ND	Respiratory distress; hypoxia
H9[E6]	33 and 37/M/W	ND	<i>Histoplasma and Cryptococcus</i>	Age 33: <i>Histoplasma</i>	Age 33: Tongue (<i>Histoplasma</i>)	Ketoconazole (ND); amphotericin B (ND); <i>Histoplasma</i>);	Lobectomy for <i>Histoplasma</i> -related

					Age 37: Meningitis (<i>Cryptococcus</i>)	fluconazole (ND; <i>Cryptococcus</i>)	bronchiectasis
Cr1	18/M/A	R423Q (DNA binding)	<i>Cryptococcus gattii</i>	<i>Cryptococ- cus gattii</i>	<i>Cryptococcus gattii</i> meningitis	Liposomal amphotericin B, flucytosine, voriconazole	Papilledema and increased opening pressures without vision changes
Cr2[E7]	29/F/B	ND	<i>Cryptococcus neoformans</i>	N	Colon, perirectal abscess	Amphotericin B (10 wk) and 5- flucytosine (ND)	ND
Cr3[E8]	31/M/W	ND	<i>Cryptococcus neoformans</i>	N	Esophageal	Amphotericin B (6 wk) and 5- flucytosine (6 wk)	ND
Cr4[E9]	13/M/ND	ND	<i>Cryptococcus neoformans</i>	N	Meningitis	Amphotericin B (10 wk), 5-flucytosine (6 wk), dexamethasone (12 wk), fluconazole (6 mo)	Myeloperitoneal shunt
Co1	4/F/W	V713M (Trans- activation)	<i>Coccidioides immitis</i>	Y	Meningitis	Liposomal amphotericin, fluconazole	Stroke, hydrocephalus, bilateral VP-shunt hypercalcemia
Co2[E10]	17/F/ND	T412S (DNA binding)	<i>Coccidioides immitis</i>	Y	Meningitis, cerebral abscess	Amphotericin B (3 wk), fluconazole (lifelong)	Altered mental status requiring temporary intubation
Co3 [E11]	4/F/ND	ND	<i>Coccidioides immitis</i>	N	Meningitis	Amphotericin B (ND), fluconazole (ND)	Strokes

* Age is reported in years

** Remains on secondary prophylaxis, unless stated otherwise

A: Asian, B: Black, ND: Not Documented, W: White

[E] references can be found in the Online Repository.

References of Literature Review Patients

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