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Clinical outcome and prognosis of young patients with mycosis fungoides

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Abstract

Background—Mycosis fungoides (MF) in young patients is rare and may have atypical presentations. There are limited data in these patients.

Objective—Determine the clinical outcome and prognosis of young patients with MF.

Methods—A search of our institutional cancer registry database was conducted for patients diagnosed with MF at ≤ 30 years of age.

Results—Our study includes 74 patients (median age at diagnosis= 25.5 years). Most patients (n=65, 88%) presented with early stage disease and variants of MF (n=44, 59%) leading to a median delay in diagnosis of 2.5 years. Hypopigmented MF (n=27, 36.5%) was the most common variant affecting predominantly African-American (44.4% vs.19%, p= 0.02) and younger age group (20 vs. 26 years, p<0.001). All patients with hypopigmented MF presented with early stage disease and were less likely to develop progressive disease (PD) compared to other variants (11% vs. 34%, p= 0.03). Overall, nineteen patients (26%) developed PD during median follow-up of 3.5 years which was associated with advanced stage disease (89% vs. 17%, p<0.0001), older age (>20

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years) [31% vs. 13%, $p=0.08$ (NS)], African-American race (52.6% vs. 20%, $p=0.0090$) and poikilodermatous presentation ($p<0.01$). The overall survival was favorable (97.2% and 95.9% at 5 and 10 years) despite delay in diagnosis and atypical presentation.

Conclusions—PD is associated with older age, African-American race, the poikilodermatous variant, and advanced-stage disease. The hypopigmented variant is a common presentation in young patients and has an indolent disease course.

Our study confirms an overall favorable prognosis in young patients with MF.

Keywords

Cutaneous T-cell lymphoma; mycosis fungoides; Sezary syndrome; hypopigmented mycosis fungoides; young patients; overall survival; prognosis

INTRODUCTION

Mycosis fungoides (MF) is the most common type of primary cutaneous T-cell lymphoma (CTCL).¹ It typically affects older adults with the majority of patients diagnosed after 50 years of age.² Although the survival of patients diagnosed with early stage disease is similar to that of age matched controls, patients diagnosed with advanced disease (stage IIB) have a median survival ranging from 17 months to 4.7 years.^{3, 4}

Primary CTCL is rare in children. In the United States, only 14 cases of CTCL in children <9 years of age and 33 cases in the age group 10–19 years were reported to cancer registries between 1973–2002, resulting in age-adjusted incidence levels of 0.1 and 0.3 per 1,000,000 persons-years, respectively.⁵ In a Surveillance Epidemiology and End Results (SEER) population based study, the incidence of MF/Sézary syndrome (SS) was reported as 0.3 per 1,000,000 person-years for patients <20 years of age and 1.0 per 1,000,000 person-years between 20–29 years of age.⁶ In a recent study by Kempf et al., MF was reported as one of the most prevalent cutaneous lymphomas in children.⁷ The diagnosis of MF may be delayed in young patients due to its low incidence and varied clinical presentation, as well as physician reluctance to perform skin biopsies in younger individuals.^{8, 9}

Retrospective studies suggest that the prognosis of MF in young patients is favorable compared to adults, but there are no large studies that define the disease course and long-term outcomes in young patients.^{10, 11} Based on the large SEER population study,⁶ we decided to use 30 years as the cutoff age for our study. In this study, our objective was to analyze clinical features, disease course, risk of disease progression, and overall survival of a cohort of patients diagnosed with MF at a young age.

METHODS

Following approval by the Institutional Review Board at Memorial Sloan Kettering Cancer Center (MSKCC), a search of the cancer registry database at our institute was conducted. ICD-O histology codes for MF (M9700), pathology key words (“mycosis fungoides”/ “cutaneous T-cell lymphoma”/ “parapsoriasis”) and age at tumor diagnosis date, “years is

less than or equal to 30", were the search terms specified. All patients retrieved from the time of the database creation in 1990 to the date of search in March 2015, were reviewed.

After identification of 171 unique potential patients, we reviewed the charts and pathology of all patients for inclusion/exclusion criteria. Patients with clinically and histopathologically confirmed MF were selected. Our inclusion criteria included diagnosis at 30 years of age, the clinical diagnosis of "mycosis fungoides" and histology "consistent with", "compatible with", "most in keeping with" MF, as well as the presence of an "atypical epidermotropic lymphocytic infiltrate" with positive clonality. Exclusion criteria included patients who were never seen by a dermatologist at MSKCC or did not receive histopathological confirmation of MF by a dermatopathologist at MSKCC. Pathology reports described as "possible MF", "probable MF", "suspicious for MF", "worrisome for MF", and "not unequivocal for MF" were excluded. In total, 97 patients were excluded, resulting in 74 patients who were included in the study.

A retrospective chart review was performed for each patient using the electronic medical record. Data gathered included: (a) demographic variables - age, gender, race, date of birth, date of reported onset of rash and date of initial diagnosis; (b) past medical history - presence of comorbidities, specifically, asthma, eczema and other lymphoma/lymphoproliferative disorders; (c) family history of lymphoma/leukemia; (d) physical examination findings at presentation and at follow-up visits including lymph node/s and organomegaly status; (e) disease stage at presentation; and (f) treatment(s), types of response, and dates of response. Complete remission (CR) was defined as complete resolution of cutaneous/extracutaneous disease. Partial remission (PR) was defined as equal or more than 50% resolution of cutaneous/extracutaneous disease. Stable disease (SD) was defined as less than 50% response with no evidence of disease progression. Progressive disease (PD) was defined as >25% increase in skin disease from baseline or the development of new tumors or increase in stage of the disease. Relapse was defined as recurrence of disease after complete remission. Disease course and treatment responses were recorded from clinical encounters with the cutaneous lymphoma experts at our institute and from the images that were taken at each follow-up visit.

Statistical Analysis

All descriptive and comparative statistical analyses were performed using Stata software (version 12.0; StataCorp LLC). Descriptive analysis of the patients is summarized in Table I. When exact dates (for initial diagnosis of MF, last follow-up visit, and/or death), were not available, we used the first day of the month of occurrence or June 1 of the year of occurrence of the event, depending on how much information was present. Delay in diagnosis was calculated from date of reported onset of rash to the date of initial diagnosis whether at MSKCC or an outside institution. Overall survival was defined as the percentage of patients alive at the end of 5 and 10 years of follow-up, respectively. Total years of follow-up were calculated from the date of initial diagnosis of MF to the most recent clinic visit at the time of data collection or date of death.

RESULTS

Patient characteristics

Median age at diagnosis was 25.5 years (range 6–30 years). Fifty-one (69%) patients were in the age group 21–30 years (median age 26.7 years), while the remaining 23 (31%) were 20 years of age (median age 17 years) at the time of diagnosis. Median age of reported onset of rash was 19 years (range 6–29 years) and median delay in diagnosis was 2.5 years (range 0–28 years). Three patients presented at the age of 12 years (two were below 10 years) with no reported delay in diagnosis. Median duration of follow-up was 3.5 years (range 1 month–34 years) (Table 1).

Clinical characteristics

Sixty-five (88%) patients presented with early stage MF (I–IIA), while the remaining 9 (12%) patients presented with advanced stage (IIB–IV) disease. Both patients diagnosed at <10 years of age presented with early stage (IB) hypopigmented MF. Of patients with advanced stage disease (n=9), 5 (55.5%) were African-American (AA) (p=0.054) (NS), 5 (55.5%) had classic MF characterized by patch, plaque and/or tumor lesions, while 2 (22.2%) patients each had poikilodermatous and folliculotropic disease, respectively.

Clinically, patches were present in 37 (50%) patients, plaques in 22 (30%), papules in 8 (11%), ulcers in 2(3%), and skin tumors in 4 (5%) patients. Lower extremities (81%) were most commonly involved, followed by trunk (76%) and upper extremities (61%). Head and neck were involved in twelve (16%) patients.

Thirty patients (40.5%) presented with classic mildly erythematous, scaly patches and/or plaques. However, the remaining 59.5% patients presented with non-classic variants; 27 (36.5%) with hypopigmented, 6(8.1%) with hyperpigmented, 4(5.4%) with poikilodermatous, and 7(9.5%) with folliculotropic MF. Hypopigmented MF was most commonly reported in AA patients (44.4%) and Asians (18.5%).

We compared the characteristics of hypopigmented MF (n=27, 36.5%) to non-hypopigmented MF patients (n=47, 63.5%) (Table 2). Patients with hypopigmented MF were more likely to be AA (44.4% vs.19%, p= 0.02) and of younger age at diagnosis (20 vs. 26 years, p<0.001). All patients with hypopigmented MF presented with early stage (I–IIA) while 19% with non- hypopigmented MF had advanced stage disease at presentation (p=0.01). At last follow-up, 16(34%) patients with non-hypopigmented MF and 3(11%) with hypopigmented MF had evidence of PD, which was significantly different (p= 0.03).

Histopathology

Patch and plaque lesions of MF were typically described with the characteristic epidermotropic atypical lymphocytes, allowing definitive pathologic confirmation. For a diagnosis of folliculotropic MF, an atypical folliculotropic lymphoid infiltrate with or without mucinosis was required. Reported immunohistochemical findings confirmed the presence of some CD4+ lymphocytes in all evaluated cases. However, the epidermotropic infiltrates were found to be CD8-predominant in most cases of hypopigmented MF. T-cell

receptor gene rearrangements were monoclonal in thirty-eight (51%) cases. Thirteen (18%) cases were not evaluated by molecular techniques.

Disease course and outcome

Nineteen patients (26%) developed progressive disease (PD) during follow up. A higher, although not statistically significant, percentage of those age >20 years at diagnosis developed PD than those in the younger age group (31% vs. 13%, $p=0.08$) [Table 3]. AA patients had a higher risk of PD ($p<0.01$) compared to others. Sex of the patient, family history of lymphoma, past medical history of other lymphoma/lymphoproliferative disorders and other comorbidities were not significantly associated with disease progression.

All evaluated patients ($n=8$, 89%) diagnosed with advanced MF (stage IIB-IV) had evidence of PD during the follow up period. In contrast, most (73%) patients with stage I/II disease had stable disease with good response to treatment. Of note, one of the patients with stage II was lost to follow up after initial consult.

All four (100%) patients with poikilodermatous MF had PD ($p<0.01$). Of these, three patients progressed to develop skin tumors (T3N0M0B0) or nodal/systemic disease (T4NXM0B0, T2N3M1B2), the fourth patient had progression limited to the skin (T2N0M0B0).

Three of seven (42.8%) patients with folliculotropic MF ($p=0.2510$) progressed to advanced stage (T3N0M0B0, T2N3M0B0, T4N2M1B1) disease, while the remaining four had stable cutaneous disease (T1-2N0M0B0).

Overall survival of our cohort of 74 patients at 5 and 10 years was 97.2% and 95.9%. Four patients in our cohort died. Three patients died at the age of 28, 32 and 30 years due to rapidly progressive MF/Sezary syndrome with systemic/blood involvement within 1, 2, and 8 years of diagnosis, respectively. All three had classic MF and were >20 years of age at diagnosis. A fourth patient died 11 years after MF diagnosis due to complications of coexisting metastatic thymoma at the age of 38 years.

Treatment

Patients with early-stage disease received skin-directed treatments. Phototherapy (narrow band ultraviolet B [nbUVB], psoralen and ultraviolet A [PUVA], UVA-1) was the most commonly prescribed primary treatment for the group ($n=36$, 48.6%) followed by topical agents (mid- and high-potency corticosteroids, topical nitrogen mustard or bexarotene) ($n=22$, 29.7%). Most patients received nbUVB ($n=34$, 46%); PUVA was the primary treatment for two (2.7%) patients. Of those receiving nbUVB, 15 of 34 patients (44.1%) achieved complete remission, 3 (8.8%) progressed while on nbUVB; while all others (47.1%) remained stable or had partial remission. Three patients had mild cutaneous relapses with limited lesions after initial complete remission, which were subsequently controlled with topical steroid use.

All patients with stage IV ($n=5$, 6.75%) disease at presentation received chemotherapy (CHOP, EPOCH or doxorubicin and gemcitabine) as first line treatment. Local radiation was

used to treat skin tumors in 2 patients (2.7%). Five patients (6.8%) required treatment with systemic agents (methotrexate, bexarotene, mogamulizumab/anti-CCR-4) for rapidly progressive disease.

DISCUSSION

Mycosis fungoides (MF) in young patients is rare and often misdiagnosed, leading to delay in diagnosis. This is partly because MF may clinically mimic benign rashes, particularly atopic dermatitis, eczema, pityriasis alba or vitiligo. The clinical resemblance to other pediatric eruptions, low index of suspicion and reluctance to biopsy have led to a median delay in diagnosis of 2.5 years in our patients.¹² MF typically presents in the fifth and sixth decades of life, but can present as early as the first decade; the youngest patient in our cohort was 6 years old at the time of diagnosis. An increasing incidence was seen with age, but without any sex predominance, similar to a large population-based study.¹

We noticed a higher occurrence of MF variants in our young patient population compared to reported data on adult patients¹³: 36.5% with hypopigmented, 8% with hyperpigmented, 5% with poikilodermatous, and 9.5% with folliculotropic MF. We compared the characteristics of patients with hypopigmented MF to the remaining group. All patients with hypopigmented MF presented with early stage disease (IA-IB), followed by an indolent disease course with a favorable response to phototherapy. It most commonly affected patients age < 20 years and AA patients. Our results are similar to the findings of a large retrospective study of pediatric hypopigmented MF by Castano et al., which demonstrated hypopigmented MF as the most common variant (72.5%) in a group of 69 patients diagnosed before 21 years of age.¹⁴

Our data showed an overall survival of 97% and 96% at 5 years and 10 years, respectively. Disease progression was seen in a quarter of our patients (26%); and nearly half (42%) of these patients, who progressed, had advanced stage disease at initial presentation. Age >20 years, advanced stage, and AA race were found to be independent risk factors for disease progression. In support of our findings, a retrospective analysis from 1987–2007 at M.D. Anderson Cancer Center, showed that AA patients were more than twice as likely to be diagnosed before the age of 40 years, as compared to Caucasians.¹⁵ A more recent analysis by Balagula et al. confirmed an increase in the incidence of early-onset MF in AA females, with a higher risk of disease progression and poorer prognosis.¹⁶

In addition, all patients with poikilodermatous MF in our cohort showed disease progression. These data are in contrast to previously published data where only 2 (4%) of 49 patients progressed.¹⁷ This discrepancy may be due to significantly older median age presented as compared to our study (44 vs. 25.5 years). This suggests that younger patients with poikilodermatous variant may have a more aggressive course than those who are older, although it is difficult to extrapolate definitive conclusions given the limited number of patients with poikilodermatous MF in our group.

Treatment options and outcome in young patients are not well-defined. Narrowband UVB therapy for early MF in children is one of the first line regimens recommended due to its low

side-effect profile.^{18, 19} Skin-directed regimens, including topical steroids, phototherapy or combinations thereof, were the most commonly applied treatment regimens in our group, which reflects previously published data from other centers.^{14, 20} Most patients responded favorably to these regimens. Cutaneous relapses were generally easily controlled with topical steroids or further phototherapy courses. Treatment with nbUVB was generally well tolerated, with occasional mild side-effects such as pruritus and/or erythema.

Data from retrospective studies suggest that, despite relapses, the outcome of CTCL in children seems more favorable than in adults, when monitored over a median follow-up period of 43 months-12 years.^{11, 14, 18} Our data showed similar favorable outcomes, despite a delay in diagnosis. Most patients presented with early stage disease that rarely developed into advanced stage MF. Progression of disease was most commonly seen in patients with limited skin disease (IA) as development of more widespread cutaneous lesions (IB). Disease progression into advanced (IIB-IV) stages was associated with advanced disease at initial presentation, poikilodermatous and folliculotropic subtypes of MF.

One of the limitations of our study was being a single center retrospective study. Due to the rarity of the disease in the pediatric population, we had a low number of patients diagnosed with MF under 10 years of age. Our study could benefit from collaboration among large centers across the U.S. to study this rare disease, specifically in the younger population. We studied the clinical course and outcome of young patients with MF, and did not review their histopathology, although we excluded cases in which the diagnosis was not confirmed by pathologists at MSKCC. It would also be useful to study the histopathological aspect of the disease in correlation with disease course and outcome in young patients.

In summary, our study is a large single-institution-based study of clinical outcomes and prognosis of young patients with MF. It confirms the overall favorable prognosis and outcome in young patients. Narrowband UVB and topical steroids were the most commonly prescribed treatment options with complete or partial responses achieved in most patients. Our patients with advanced stage disease had worse prognosis and outcomes similar to reported outcomes in adult MF.⁴ However, large prospective case control studies are needed to compare the clinical course and outcomes in young vs. older patients with mycosis fungoides.

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ABBREVIATIONS/ ACRONYMS

AA	African American
BSA	Body surface area
CHOP	Cyclophosphamide, hydroxydoxorubicin, Oncovin Vincristine), prednisone

CR	Complete remission
CTCL	Cutaneous T-cell lymphoma
EPOCH	Etoposide, prednisone, Oncovin (vincristine), cyclophosphamide, hydroxydoxorubicin
ICD-O	International Classification of Diseases for Oncology
LDH	Lactate dehydrogenase
MF	Mycosis Fungoides
MSKCC	Memorial Sloan Kettering Cancer Center
nbUVB	Narrowband ultraviolet B
PD	Progressive disease
PR	Partial remission
PUVA	Psoralen and ultraviolet A
SD	Stable disease
SEER	Surveillance, Epidemiology and End Results

Table 1

Clinical characteristics of the patients with mycosis fungoides

Characteristic	Category	All ages (n=74)	Ages 0–20 (n=23)	Ages 21–30 (n=51)
Age at diagnosis, median (range), years		25.5 (6–30)	17 (6–20)	27 (21–30)
Race	Caucasian	34 (46.0%)	8 (34.8%)	26 (51.0%)
	AA	21 (28.4%)	7 (30.4%)	14 (27.5%)
	Asian	7 (9.5%)	4 (17.4%)	3 (5.9%)
	Hispanic	9 (12.2%)	2 (8.7%)	7 (13.7%)
	White/Hispanic	1 (1.4%)	0 (0%)	1 (2.0%)
	AA/Hispanic	1 (1.4%)	1 (4.4%)	0 (0%)
	Did not self-report	1 (1.4%)	1 (4.4%)	0 (0%)
Gender	Male	38 (51.4%)	12 (52.2%)	26 (51.0%)
	Female	36 (48.7%)	11 (47.8%)	25 (49.0%)
Duration of follow up, median (range), years		3.5 (0–34)	2 (0–18)	4 (0–34)
MF stage	I NOS	4 (5.4%)	2 (8.7%)	2 (3.9%)
	IA (T1N0M0B0)	29 (39.2%)	7 (30.4%)	22 (43.1%)
	IB (T2N0M0B0)	32 (43.2%)	14 (61%)	18 (35.3%)
	IIA	0 (0%)	0 (0%)	0 (0%)
	IIB (T3N0M0B0, skin lesions only)	4 (5.4%)	0 (0%)	4 (7.8%)
	III	0 (0%)	0 (0%)	0 (0%)
	IIIA	0 (0%)	0 (0%)	0 (0%)
	IIIB	0 (0%)	0 (0%)	0 (0%)
	IVA1 (T4NXM0B2)	1 (1.4%)	0 (0%)	1 (2.0%)
	IVA2 (T4N3M0B0, T4N3M0B2)	2 (2.7%)	0 (0%)	2 (3.9%)
	IVB (T2N3M1B2, T4N2M1B1)	2 (2.7%)	0 (0%)	2 (3.9%)
Clinical variant	Hypopigmented	27 (36.5%)	15 (65.2%)	12 (23.5%)
	Classic	30 (40.5%)	6 (26%)	24 (47.1%)
	Hyperpigmented	6 (8.1%)	0 (0%)	6 (11.8%)
	Poikilodermatous	4 (5.4%)	0 (0%)	4 (7.8%)
	Folliculotropic	7 (9.5%)	2 (8.7%)	5 (9.8%)
Response	No progression of disease	55 (74.3%)	21 (91.3%)	34 (66.7%)
	Progression of disease	19 (25.7%)	3 (13.0%)	16 (31.4%)

IA-IVB: ISCL/EORTC revised staging of mycosis fungoides and Sezary syndrome; NOS: not otherwise specified, skin involvement with incomplete information on extent of disease.

Table 2

Clinical characteristics of patients with non-hypopigmented vs. hypopigmented MF

Characteristic	Category	Non-hypopigmented MF (n=47)	Hypopigmented MF (n=27)	p-value *
Age at diagnosis (n)	Age (0–20)	8 (17.0%)	15 (55.6%)	0.0010
	Age (21–30)	39 (83.0%)	12 (44.4%)	
Age at diagnosis, median (range) years		26 (12–30)	20 (6–30)	0.0006
Race	Caucasian	30 (63.8%)	4 (14.8%)	0.0000
	AA	9 (19.1%)	12 (44.4%)	0.0210
	Asian	2 (4.3%)	5 (18.5%)	0.0570
	Hispanic	5 (10.6%)	4 (14.8%)	0.4270
	White/Hispanic	0 (0%)	1 (3.7%)	0.3650
	AA/Hispanic	1 (2.1%)	0 (0%)	0.6350
	Did not self-report	0 (0%)	1 (3.7%)	0.3650
Gender	Male	28 (60.0%)	10 (37.0%)	0.0520
	Female	19 (40.4%)	17 (63.0%)	
MF stage	Early stage (I-IIA)	38 (80.9%)	27 (100.0%)	0.0120
	Advanced stage (IIB-IVB)	9 (19.1%)	0 (0%)	
Duration of follow up, median (range) years		4 (0–34)	3 (0–11)	0.4527
Response	Non-progression of disease	31 (66.0%)	24 (88.9%)	0.0260
	Progression of disease	16 (34.0%)	3 (11.1%)	
Family Hx of lymphoma/leukemia	Absent	44 (93.6%)	24 (88.9%)	0.3810
	Present	3 (6.4%)	3 (11.1%)	
Comorbidity: other lymphoma/ lymphoproliferative disorders	Absent	46 (97.9%)	26 (96.3%)	0.6000
	Present	1 (2.1%)	1 (3.7%)	
Comorbidity: asthma	Absent	44 (2.1%)	22 (81.5%)	0.1110
	Present	3 (6.4%)	5 (18.5%)	
Comorbidity: eczema	Absent	45 (95.7%)	25 (92.6%)	0.4640
	Present	2 (4.3%)	2 (7.4%)	

* Fisher's exact test

Table 3

Clinical characteristics of patients with non- progressive disease vs. progressive disease (PD)

		Disease Progression		<i>p</i> -value*
		No (n=55)	Yes (n=19)	
Age at diagnosis (n)	Age (0–20)	20 (36.4%)	3 (15.8%)	
	Age (21–30)	35 (63.6%)	16 (84.2%)	0.0800
Age at diagnosis, median (range) years		25 (6–30)	26 (14–30)	0.0971
Gender	Male	28 (50.9%)	10 (52.6%)	
	Female	27 (49.1%)	9 (47.4%)	0.5550
Race				0.0880
	Caucasian	28 (50.9%)	6 (31.6%)	0.1160
	AA	11 (20.0%)	10 (52.6%)	0.0090
	Asian	7 (12.7%)	0 (0%)	0.1130
	Hispanic	6 (10.9%)	3 (15.8%)	0.4190
	White/Hispanic	1 (1.8%)	0 (0%)	0.7430
	AA/Hispanic	1 (1.8%)	0 (0%)	0.7430
	Did not self-report	1 (1.8%)	0 (0%)	0.7430
MF stage	I-IIA	54 (98.2%)	11 (57.9%)	
	IIB- IV	1 (1.8%)	8 (42.1%)	0.0000
Family Hx of lymphoma/leukemia	Absent	52	16	
	Present	3	3	0.1548
Comorbidity: Other lymphoma/ lymphoproliferative disorders	Absent	53	19	
	Present	2	0	0.3994
Comorbidity: Asthma	Absent	48	18	
	Present	7	1	0.3664
Comorbidity: Eczema	Absent	52	18	
	Present	3	1	0.9746
Clinical variant	Hypopigmented	24 (43.6%)	3 (15.8%)	0.0260
	Classic	22 (40%)	8 (42.1%)	0.5400
	Hyperpigmented	5 (9%)	1 (5.3%)	0.5130
	Poikilodermatous	0 (0%)	4 (21%)	0.0030
	Folliculotropic	4 (7.3%)	3 (15.8%)	0.2510

* Fisher's exact test.

Abbreviations; AA, African American