original article

Cutaneous T-cell lymphoma in Saudi Arabia: retrospective single-center review

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BACKGROUND: Cutaneous T-cell lymphoma (CTCL) is an uncommon disease with various clinical presentations. The hypopigmented type is more common in individuals with a dark skin complexion. Moreover, childhood CTCL is more common in Mediterranean populations in comparison to the West.
OBJECTIVE: To describe CTCL in the Saudi population.
DESIGN: A retrospective collection of data on all cases of CTCL from 2010-2016.
SETTING: Dermatology clinic at a tertiary center in Riyadh, Saudi Arabia.
PATIENTS AND METHODS: We collected data on all cases of CTCL diagnosed clinically and confirmed pathologically.
MAIN OUTCOME MEASURE(S): The number of cases of CTCL, gender, age at diagnosis and clinical subtypes.
RESULTS: The most common presentation among 125 patients was the classic type (patches and plaques) followed by the hypopigmented and poikilodermatous variants. Males were 58% of the population (n=72). The median age at diagnosis was 41 years and the range was 5 to 86 years. Thirteen percent were younger than 20 years of age.
CONCLUSION: Hypopigmented and poikilodermatous types of CTCL are more common in our population than in the West.
LIMITATION: Retrospective, single-center data may not be generalizable since difficult cases are more likely to be referred to a tertiary center.

Cutaneous T-cell lymphoma (CTCL) is an uncommon non-Hodgkin lymphoma. It is the second most common extranodal type after the gastrointestinal tract with an overall incidence 1:100 000.[1] The male-to-female ratio is 2:1. The most common type of CTCL is mycosis fungoides (MF), which represents about 70% of all cases.[1] CTCL is a great mimicker, presenting with various clinical presentations, among them plaques and patches (classic type) being the most common. However, tumors and erythroderma can be initial presentations as well. Other less common clinical variants are hypo- and hyperpigmented, follicular, granulomatous and verrucous. Although 80% to 90% of those affected with early MF have a favorable prognosis, 10% to 20% can progress to more advanced disease with a marked increase in mortality (26% survival at 5 years).[2]

Different therapeutic modalities have been used to treat CTCL. Skin-directed therapy is the most common, which includes topical steroids, topical nitrogen mustard and phototherapy. Other options include retinoids, interferon, methotrexate, histone deacetylase inhibitors and extracorporeal photopheresis. Unfortunately, the response to chemotherapeutic agents is generally poor.We present epidemiological data and common clinical presentations of CTCL in a tertiary care center.

PATIENTS AND METHODS

We collected data from the records of all patients who had been diagnosed with CTCL based on clinical descriptions that were confirmed by histopathology at the Dermatology Department, King Faisal Specialist Hospital and Research Centre, Riyadh, Saudi Arabia, from 2010-2016.

Table 1. Clinical presentation of the 125 cases of cutaneous T-cell lymphoma.

Age group (years)	Number of cases	Gender		Comorbidities	Clinical subtypes (number of cases)				
		Male	Female		Patch	Plaques	Hypopigmented	Poikilodermatous	Others
0-10	4	4	-	-	0	1	3	0	0
10-20	12	6	6	-	1	0	8	1	Lichen planus –like (n=1), Hyperpigmented (n=1)
20-30	19	11	8	Thyroid carcinoma (n=1)	5	2	4	2	Folliculotropic (n=2), Erythrodermic (n=1), Hyperpigmented (n=1), Verrucous (n=1), 1 LYP(n=1)
30-40	26	14	12	-	2	11	2	1	Tumor (n=1), Hyperpigmented (n=1), Folliculotropic (n=4), Granulomatous (n=1), LYP (n=1), Sézary (n=1), Ichthyosis (n=1)
40-50	23	11	12	RA (n=1), Hodgkin lymphoma (n=1), breast cancer (n=1)	10	1	1	7	Linear folliculotropic (n=1), LYP (n=1)
50-60	21	11	10	RA (n=1), rectal cancer (n=1)	5	8	1	2	Erythrodermic (n=3), Tumor (n=1), Sézary (n=1)
60-70	5	5		RA (n=1)	1	3	-	-	Erythrodermic (n=1)
70-80	12	7	5	RA (n=1)	2	3	-	-	Erythrodermic (n=2), Tumor (n=1),CD30+ LPD (n=2), Transformed (n=1) PCTCL (n=1)
80-90	3	3	-	-	2	-	-	-	Sézary (n=1)
Total	**125**	**72**	**53**	**RA (n=4), malignancy (n=4)**	**28**	**29**	**19**	**13**	**36**

LYP: lymphomatoid papulosis, LPD: lymphoproliferative disorders, PCTCL: Peripheral cutaneous T-cell lymphoma, RA: rheumatoid arthritis.

RESULTS

Of 125 patients diagnosed with CTCL, 72/125 (58%) were males and 53/125 (42%) females. The median age at diagnosis was 41 years (range, 5-86 years; IQR, 27-54 years). Sixteen (12.8%) patients were younger than 20 years of age and 35 (28%) were younger than 30 years of age (**Table 1**).

The most common clinical presentations were plaques (23%) and patches in (22%) (**Table 1, Figure 1**). **Figure 2** shows presentations of typical cases. Other types encountered once each were ichthyosis-like CTCL, linear, lichen-planus pigmentosus-like, verrucous and granulomatous. Four patients had rheumatoid arthritis as a comorbidity and all were females. Another four patients had a second neoplasm which included one case each of Hodgkin lymphoma, thyroid, rectal and breast carcinomas.

Most patients had received narrowband UVB phototherapy along with topical steroids. Patients with thick plaques had oral PUVA therapy (psoralen and UVA). Two patients had total skin electron beam. Systemic therapy was limited to pegylated interferon (due to availability), methotrexate and retinoids (isotretinoin and acitretin).

DISCUSSION

CTCL can present as different clinical types so the diagnosis requires a high index of clinical suspicion and histopathologic confirmation. The median age at diagnosis was 41 in our study, which is older than the age reported by Alghamdi et al,[3] the single previous report from the same region. The median age in our study was younger than that reported by Kim et al[4] (57 years). Twenty-eight percent of our patients were younger than 30 years of age, which is fewer than in the previous report by Alghamdi et al (50%). This could be explained by referral bias as we follow more advanced cases.

The most common presentation is the classic type, which includes patches and thin plaques. However, other clinical variants are not uncommon. The hypopigmented type occurred in 15.2% of our patients, which is lower than reported by Alghamdi et al (42%). This could also be explained by referral bias. The clinical presentations in our population correspond to the general belief that the hypopigmented variant is more common in a population with a dark skin complexion.[5] However, the poikilodermatous variant is a common presentation in our population. This could be attributed to the dark skin complexion, which makes the hypopigmentation more obvious. Moreover, the cosmetic appearance of hypopigmentation and poikilodermatous changes and the fear of vitiligo stigma causes affected individuals to seek medical advice early on.

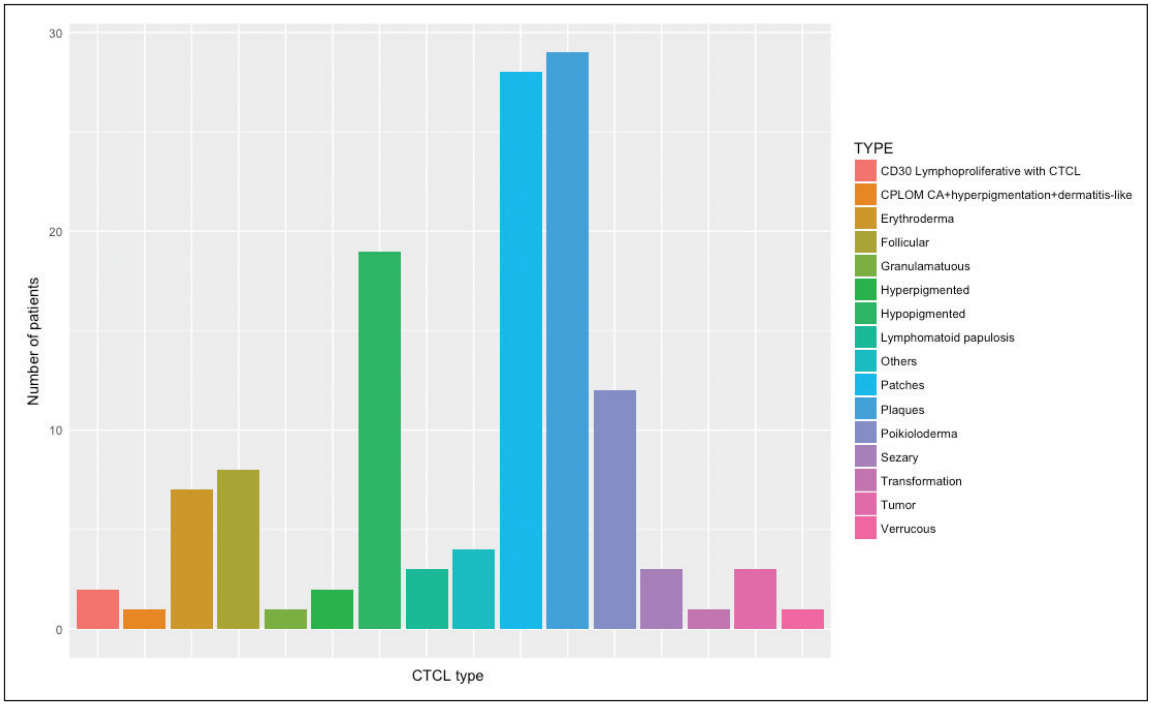

Figure 1. Type of cutaneous T-cell lymphoma at time of diagnosis.

The higher incidence in younger people (less than 20 years of age) (12.8%) is similar to reports from Kuwait and Israel, but much higher than a report in a Caucasian population.[6-8] In our study, the most common presentationin the age group younger than 20 years was the hypopigmented variant, which occurred in 11 of 16 patients (69%).

This study is the second report of CTCL in Saudi Arabia, but includes data on more patients. However, our study is limited by possible referral bias as we see more complex cases. A multicenter epidemiological study and national registry would be the best way to study such an uncommon and important disease.

Data presented at the 3rd World Congress of Cutaneous Lymphoma, October 2016, New York, NY.

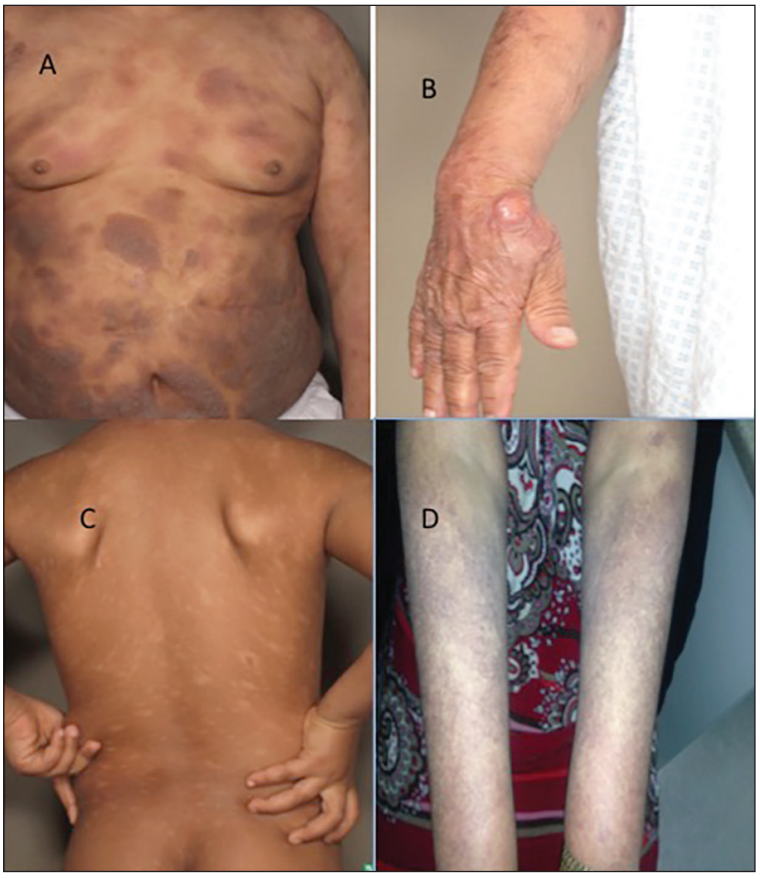

Figure 2. Different clinical presentations: A) plaques, B) tumor, C) hypopigmented, D) poikilodermatous.

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