

Long-Term Outcome of 490 Patients with Early-Stage Extra-nodal Marginal Zone Lymphoma

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OBJECTIVE

To report the long-term outcome of a large cohort of extra-nodal MZL and assess the impact of patient and disease characteristics, organ site, and treatment strategy on disease control and survival.

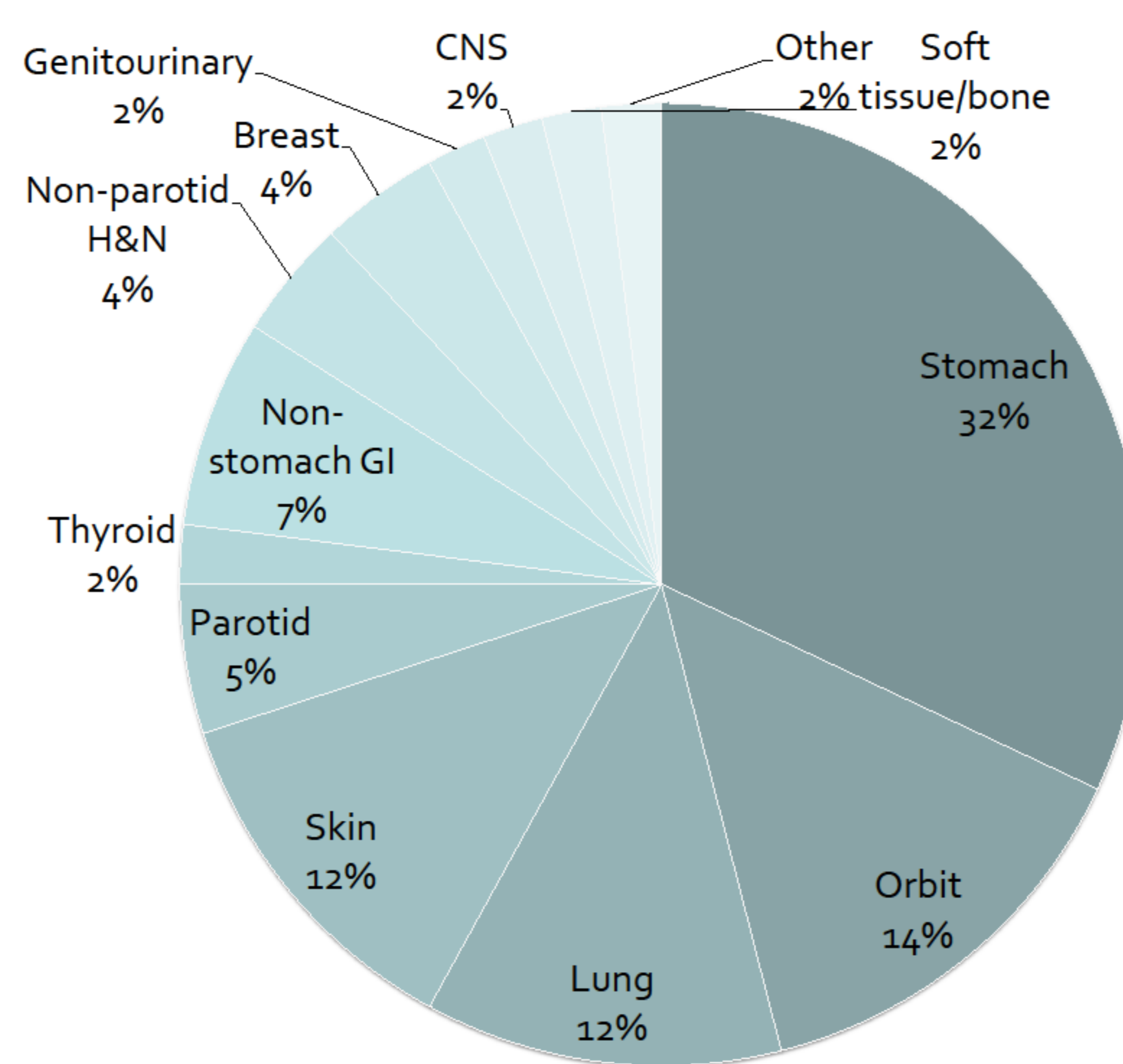
METHODS

- 490 consecutive patients with stage IE or IIE MZL referred from 1992 - 2012 to Memorial Sloan Kettering Cancer Center.
- Pathology was confirmed by hematopathologists.
- Patient characteristics, disease factors, and treatment types were analyzed for association with relapse-free survival (RFS), overall survival (OS), and cumulative incidence of relapse.

RESULTS

Patient Characteristic		N=490
Age	Median in years (Range)	60 (9-92)
Gender		
	Female	278 (57%)
	Male	212 (43%)
Stage		
	IE	436 (89%)
	IIE	54 (11%)
IPI risk group		
	Unknown	2 (0%)
	Low risk (0-1 points)	463 (94%)
	Low-intermediate risk (2 points)	25 (5%)
B-symptoms		
	Not present	466 (95%)
	Present	18 (4%)
	N/A	6 (1%)
Initial Treatment type		
	Surgery	147 (30%)
	Chemotherapy	10 (2%)
	RT	244 (50%)
	ChemoRT ^a	1 (0%)
	Immunotherapy	19 (4%)
	Observation	43 (9%)
	Other	26 (5%)

Figure 1: Primary Sites of Disease at Diagnosis.



- Median follow-up 4.8 years.
- 5 year **OS** 90% and **CSS** 98.7% (Figure 2)
- **Cumulative incidence of relapse** was 29% by 5 years and 39% by 10 years.
- Amongst the 384 patients with complete response (CR), 99 experienced relapse.
- 11 patients (2%) transformed to large-cell lymphoma.
- After RT, only 11 patients experienced in-field failure (5-year cumulative incidence of 2.4%).
- Distant relapses most common.
- Second tumors in irradiated sites developed in 3 patients: 2 of these were breast ductal carcinoma in situ cured with surgical resection.

Figure 2: Overall and Cause-Specific Survival.

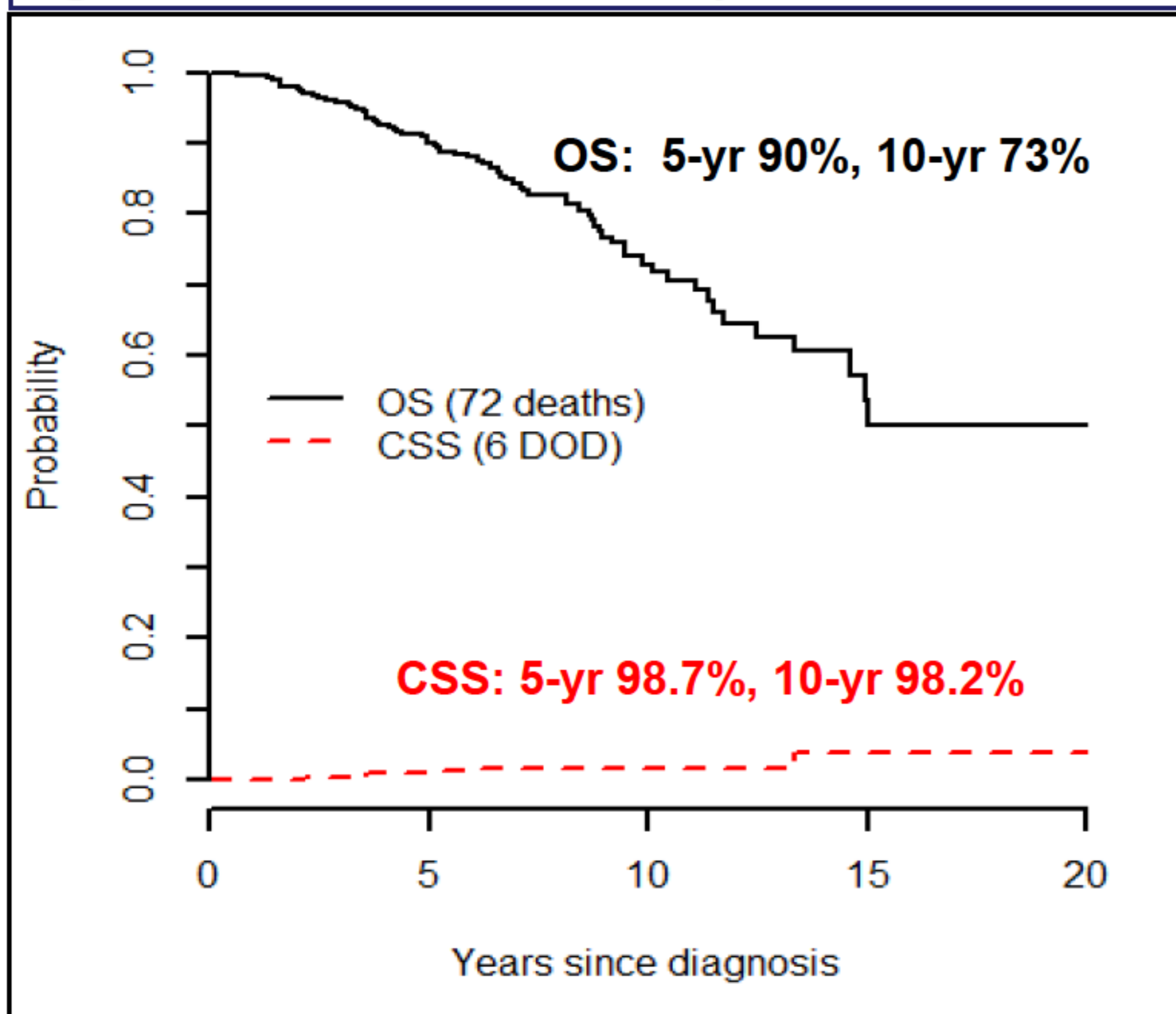
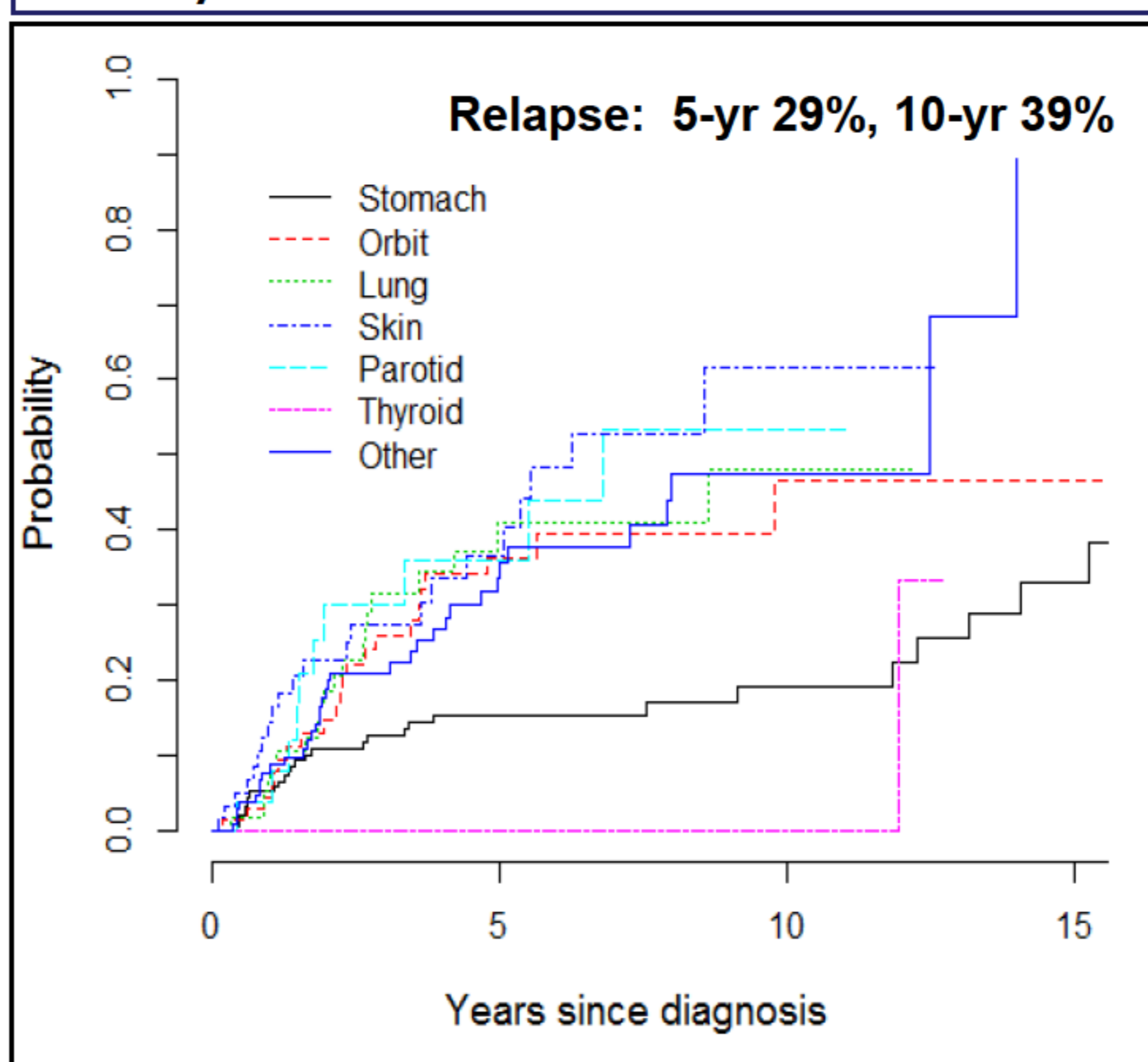


Figure 3: Cumulative Incidence of Relapse by Primary Disease Site.



Factors associated with Relapse:

- On multivariable analysis, initial treatment type and primary disease site were independently associated with RFS and relapse (all $p < 0.005$). All disease sites ($HR > 2.0$, $p \leq 0.01$) except for thyroid ($p = 0.8$) had worse RFS relative to stomach (Figure 3).
- Compared with RT, chemotherapy or immunotherapy had worse RFS ($HR 2.2$, $p = 0.004$) while surgery was no different ($p = 0.52$).

CONCLUSIONS

- Overall and cause-specific survival are excellent in early-stage extra-nodal MZL.
- Treatment with RT or surgery was associated with longer RFS and reduced the need for salvage.
- Relapses are common after initial remission, and most frequently occur in distant sites.
- Transformation to large cell lymphomas is rare.
- Stomach cases are less likely to relapse than other anatomic primary sites, perhaps in part because the entire organ is irradiated, versus other sites that are either bilateral or only part of the organ is treated (skin, lung).
- This study supports the use of local therapies to treat stage IE and IIE MZL.