Assessing the Utility of the Newly-Published IMACS Guidelines for Detecting Malignancy in Paraneoplastic DM Patients: A Retrospective Cohort Study

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Background

- The International Myositis Assessment and Clinical Studies Group (IMACS) recently published the first consensus-based cancer screening guidelines for patients with idiopathic inflammatory myopathies (IIM) including dermatomyositis (DM).[1]
- Given the known association between IIM and cancer,[2,3] these recommendations aim to provide practical guidance for malignancy screening by stratifying patients as “high,” “intermediate,” or “low” risk for malignancy based on a constellation of factors including IIM diagnosis, age of disease onset, autoantibody profiles, and clinical features.
- We sought to investigate the utility of these guidelines in patients with DM.

Methods

- We performed a single-center, retrospective cohort study of 525 DM patients at the University of Pennsylvania.
- 44 patients with DM and a diagnosis of malignancy within 5 years of DM symptom onset were identified.
- Patients with a diagnosis of non-melanoma skin cancer were excluded.
- Following IMACS guidelines (figure 1), we stratified patients based on cancer risk.

	‘High risk’ factors	‘Intermediate risk’ factors	‘Low risk’ factors
IIM subtype	☐ Dermatomyositis	☐ CADM ☐ Polymyositis ☐ IMNM	☐ ASSD ☐ CTD-associated IIM
MSA and MAA	☐ Anti-TIF1-γ antibodies ☐ Anti-NXP2 antibodies	☐ Anti-SAE1 antibodies ☐ Anti-HMGCR antibodies ☐ Anti-Mi2 antibodies ☐ Anti-MDA5 antibodies	☐ Anti-SRP antibodies ☐ Anti-Jo1 antibodies ☐ Non-Jo1 ASSD antibodies ☐ MAA[a]
Clinical features	☐ Age >40 years at IIM onset ☐ Persistent high disease activity despite therapy ☐ Dysphagia (moderate to severe) ☐ Cutaneous necrosis	☐ Male sex	☐ Raynaud phenomenon ☐ Inflammatory arthropathy ☐ Interstitial lung disease
Total			

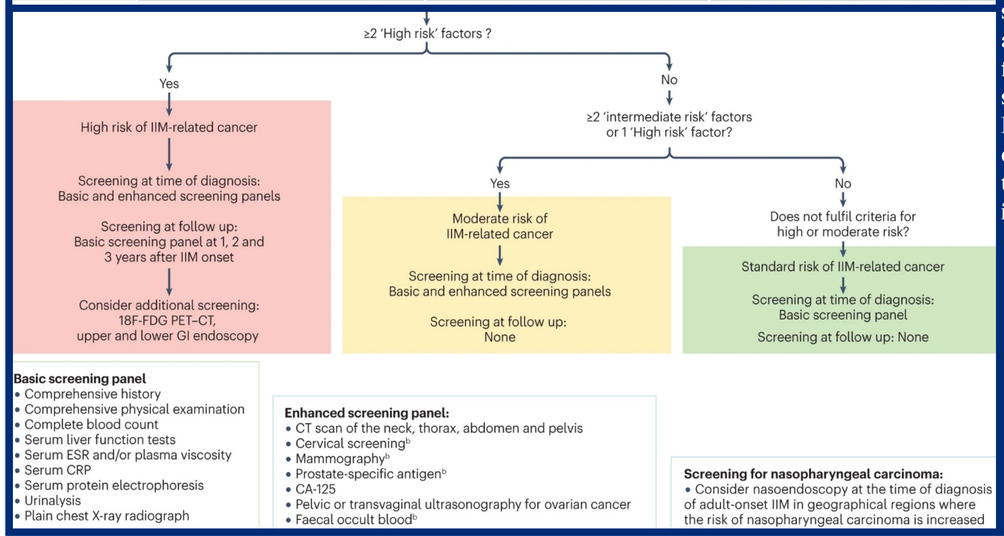

Figure 1: Risk stratification and frequency of screening for IIM-related cancer from the IMACS initiative.[1]

Results

Paraneoplastic Dermatomyositis Patients

	High-risk patients n=19 No	%	Intermediate-risk patients n=9 No	%
Sex				
Female	16	84.20%	9	100%
Male	3	15.80%	0	--
Race				
White	17	89.50%	9	100%
Black	2	10.50%	0	--
Type of DM				
Classic DM	16	84.20%	1	11.10%
Amyopathic DM	3	15.80%	7	77.80%
Hypomyopathic DM	0	--	1	11.10%

Years elapsed from DM diagnosis to cancer diagnosis	n=11 patients diagnosed with cancer after DM		n=3 patients diagnosed with cancer after DM	
0 years	6	54.50%	2	66.60%
1 year	4	36.40%	1	33.30%
2 years	1	9.10%	0	--

Cancer Screening				
Cancer detected by basic screening technique	5	26.30%	1	11.10%
Cancer detected by enhanced screening technique	14	73.70%	8	88.90%

Type of cancer				
Breast	9	47.40%	5	55.60%
Lymphoma	3	15.80%	0	--
Lung	2	10.50%	0	--
Bladder	1	5.30%	0	--
Thyroid	1	5.30%	0	--
Testicular	1	5.30%	0	--
Ovarian	1	5.30%	1	11.10%
Pancreatic	0	--	1	11.10%
Melanoma	0	--	1	11.10%
Prostate	1	5.30%	0	--
Vulvar	0	--	1	11.10%

Autoantibody profile				
Anti-TIF1-gamma	4	21.10%	0	--
Anti-NXP2	1	5.30%	0	--
Anti-Mi2	1	5.30%	1	11.10%
anti-MDA5	0	--	1	11.10%
Anti-Jo1	2	10.50%	0	--
Other*	4	21.10%	2	22.20%

*Other: anti-PM-Scl, anti-Ku, anti-RNP, anti-SSA/Ro, anti-SSB/La antibodies

Clinical features				
Age of disease onset >40 yrs old	17	89.50%	7	77.80%
Dysphagia (moderate to severe)	11	57.90%	0	--
Cutaneous necrosis/ ulceration	4	21.10%	0	--
Raynaud Phenomenon	4	21.10%	1	11.10%
Inflammatory arthropathy	4	21.10%	2	22.20%
Interstitial lung disease	6	31.60%	1	11.10%

- Of the 44 patients assessed, 28 had paraneoplastic DM, with the majority falling into the 'high' risk category according to IMACS guidelines.
- 10 patients were categorized as high risk exclusively due to age of onset > 40 years old and a diagnosis of classic DM.
- The data also reveals a temporal proximity between DM and cancer diagnoses, underscoring the critical window for cancer screening within the first two years after DM onset.
- No adverse events were reported in the context of cancer screening.

Conclusion

Adherence to IMACS guidelines would have detected all cancers, supporting their use for high-risk patient identification and malignancy screening.

References

1. Oldroyd, Alexander G S et al. “International Guideline for Idiopathic Inflammatory Myopathy-Associated Cancer Screening: an International Myositis Assessment and Clinical Studies Group (IMACS) initiative.” Nature reviews. Rheumatology vol. 19,12 (2023): 805-817. doi:10.1038/s41584-023-01045-w
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