Abstract 324 Table 1 Lynch syndrome and risk-reducing surgery

	Analysis at age 45 for MLH1/MSH2 mutations				Analysis at age 50 for MLH1/MSH2 mutations			
	Women who did not undergo risk-reducing surgery by age 45 (n, %)	Women who did undergo surgery by age 45 (n, %)	Compliance rate (%)	p-value	Women who did not undergo risk-reducing surgery by age 50 (n, %)	Women who did undergo surgery by age 50 (n, %)	Compliance rate (%)	p-value
All patients	43	39	47.6%		24	52	68.4%	p=0.001
Mutation				p=0.33				p=0.08
MLH1	20 (46.5%)	14 (35.6%)	41.2%		14 (58.3%)	19 (36.5%)	57.6%	
MSH2	23 (53.5%)	25 (64.1%)	52.1%		10 (41.7%)	33 (63.5%)	76.7%	
Surgical and Radiation History				p=0.11				p=0.001
Prior bowel surgery or pelvic radiation	17 (39.5%)	9 (23.1%)	34.6%		14 (58.3%)	10 (19.2%)	41.7%	
No prior bowel surgery or pelvic radiation	26 (60.5%)	30 (66.7%)	53.6%		10 (41.7%)	42 (80.8%)	80.8%	
Type of Surgery								p=0.88
Hysterectomy with BSO		33 (91.7%)				46 (88.5%)		
Hysterectomy only (at least 1 ovary remains)		5 (12.8%)				5 (9.6%)		
BSO only (uterus remains)		1 (2.6%)				1 (1.9%)		

Guidelines recommend discussing prophylactic hysterectomy and bilateral salpingo-oophorectomy (BSO) by age 40–45 for women with MLH1 or MSH2 mutations but lack consensus for timing of surgery. This study aims to define factors that impact decision-making for RRS.

Methods This IRB-approved retrospective study assessed 282 women with Lynch Syndrome with records from 2002–2020. Those preoperatively diagnosed with endometrial hyperplasia or cancer were excluded (n=75). The cohort was divided by mutation and age. Medical history was collected. Comparisons were made with Chi-Squared, McNemar, and Fisher exact tests. Compliance was calculated as the proportion of patients who underwent RRS by the specified age compared to all who met that age.

Results For MLH1 and MSH2 mutation carriers, compliance increased from 47.6% by age 45 to 68.4% by age 50 (p=0.001). Ten patients with prior bowel surgery or pelvic radiation underwent RRS by age 50 compared to 42 patients without this history (p=0.001). Compliance was 41.7% and 80.8% respectively. Surgery by age 50 included: 46 (88.5%) hysterectomy with BSO, 5 (9.6%) hysterectomy alone, and 1 (1.9%) BSO alone. The patient who underwent BSO alone had prior bowel surgery and radiation.

Conclusion The decision to undergo RRS in women with Lynch Syndrome is complex and often individualized. Factors that impact the compliance and timing of surgery include age, mutation status, and prior bowel surgery or pelvic radiation.

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DERMATOMYOSITIS AS A PARANEOPLASTIC SYNDROME IN HIGH GRADE SEROUS OVARIAN CARCINOMA: A CASE REPORT AND REVIEW OF LITERATURE

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Dermatomyositis occurring as a paraneoplastic syndrome in a high grade serous ovarian carcinoma is rare and treating the disease condition is a challenge.

A 46-year-old, nulligravid presented with an eight-month history of rash, joint pain, and progressive muscular weakness. Dermatomyositis was diagnosed in the background of cutaneous manifestations, progressive muscle weakness, elevated muscle enzymes and electromyographic findings. She was treated with prednisone, however during the course of

Abstract 325 Figure 1

Abstract 325 Table 1 Review of case reports of dermatomyositis with high grade serous ovarian carcinoma describing temporal relationship, treatment given and outcomes

Studies	Age of patient	Temporal relationship ¹	Stage	Treatment	Outcomes
Nakanishi (1999)	75 yo	Antecedent	IIIA	Tumor debulking then chemotherapy with carboplatin and paclitaxel x 6 cycles	Improvement of symptoms after surgery and chemotherapy No recurrence noted after 3 months
Yehouizen (2005)	48 yo	Antecedent	IIA	Unilateral salpingo-oophorectomy then chemotherapy with carboplatin and paclitaxel x 6 cycles	Improvement of symptoms after surgery and chemotherapy No recurrence of both condition after 16 months
Ngo (2009)	73 yo	Antecedent	IIIB	Hysterectomy with surgical staging	Improvement of symptoms after surgery
Mozani (2012)	2 cases: 1 st case: 46 yo 2 nd case: 58 yo	2 cases: Antecedent	1 st case: IB 2 nd case: stage IV	1 st case: Hysterectomy with complete surgical staging 2 nd case: Neoadjuvant chemotherapy of carboplatin and paclitaxel x 4 cycles then tumor debulking, followed by 2 more cycles of chemotherapy	1 st case: No evidence of disease for both condition for 2 years and 4 months 2 nd case: ovarian cancer progression after completed chemotherapy Death from disease after 1 year and 4 months
Hong (2013)	48 yo	Antecedent	IIIA	Tumor debulking then chemotherapy with dose dense paclitaxel and carboplatin x 6 cycles	Improvement of symptoms after surgery Complete remission of ovarian cancer at 15 months after surgery
Christie (2013)	57 yo	Antecedent	unknown stage	Neoadjuvant chemotherapy with carboplatin and paclitaxel x 6 cycles then tumor debulking	Recurrence of ovarian carcinoma after 3 months paralleled with exacerbation of dermatomyositis symptoms Death from disease 18 months after diagnosis
Salmeron (2016)	50 yo	Antecedent	IC	Hysterectomy with complete surgical staging then chemotherapy of carboplatin and paclitaxel x 6 cycles	Improvement of symptoms after surgery and chemotherapy No evidence of disease for 2.5 years
Arshad (2016)	60 yo	Subsequent	IIIB	Tumor debulking	Improvement of symptoms after surgery
Baizman (2017)	63 yo	Antecedent	IIIA	Surgery (optimally debulked) then chemotherapy with carboplatin and paclitaxel 3 cycles	Improvement of symptoms for 5 months
Field (2017)	43 yo	Antecedent	IIIC	Tumor debulking (sub-optimally debulked) then chemotherapy with carboplatin and paclitaxel x 6 cycles and bevacizumab for 16 months	No evidence of disease for 1 year Ovarian cancer recurrence paralleled with recurring dermatomyositis symptoms

¹Antecedent: Dermatomyositis was diagnosed before diagnosing ovarian carcinoma.
Subsequent: Dermatomyositis developed after the diagnosis of ovarian carcinoma.

treatment, she had an emergency exploratory laparotomy for a pelvic mass in complication. The patient underwent total abdominal hysterectomy and bilateral salpingo-oophorectomy, right hemicolectomy, and ileocolic side to side anastomosis. Histopathology revealed a high grade serous ovarian carcinoma, with a presumptive stage IIB. Post-operatively, the myositis partially improved. Steroid therapy was continued for two months. Chemotherapy was delayed because of repeated infections due to her immunosuppressed state.

Dermatomyositis is highly associated with malignancy. Early detection as well as treatment of the underlying malignancy can improve survival of this simultaneous condition.

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326 PURE TUBULAR CARCINOMA OF THE BREAST: A RARE ENTITY

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Introduction Pure tubular breast carcinoma (PTC) is a rare and well-differentiated tumor with good outcomes and a low local recurrence rate. Our study aims to determine the clinical, radiological, and appropriate management and the prognosis of this rare entity.

Materials and Methods A review of Salah Azaiez institute registry from 2004–2019 was performed including seven cases of PTC of the breast.

Results Seven patients were included in this study. The average age was 54 years old (from 36 to 70 years) and all patients were female. Physical exam revealed a breast lump in 6 patients. Six cancers were clinically detected on physical examination. In one patient the diagnosis was made by mammography. The radiologic findings were variable (BIRADS 3 in 6 cases and BIRADS 5 in one case). In our study, we found that 5 patients presented with T1 and N0 according to the TNM classification. The radiological assessment did not show any metastasis. The conservative surgery was more frequently performed (5 cases). All patients had hormone-receptor positivity and HER-2 negativity. Six patients were luminal A, and one patient was luminal B. In one case we found axillary lymph node metastasis. The adjuvant radiotherapy was indicated in all cases of breast conservation and only in one case of radical surgery. Only one patient received chemotherapy.