Chronic Malignant Acanthosis Nigricans

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We have no financial disclosures
Clinical presentation

• 40-year-old Somali male
• Presented to dermatology clinic one month after immigrating to Minneapolis from an Ethiopian refugee camp
• Described 8 year history of progressive bumps on head, neck, extremities and mucosae
• Stiff hands and tongue, decreased appetite
• Feels otherwise well, socially embarrassed
Clinical presentation

• Family History
  – 2 children in Ethiopia, healthy
  – No history of renal or GI cancer

• Social History
  – Estranged from wife and children due to social stigma
  – Non-smoker

• Medications
  – None at time of initial presentation
Clinical impression

• Cutaneous/mucosal papillomas
• Acanthosis nigricans-like intertriginous plaques
• Velvety palms
Clinical impression: differential diagnosis

- Cutaneous/mucosal papillomas
  - Verruca vulgaris
  - Epidermodysplasia verruciformis
  - Malignancy associated
- Acanthosis nigricans-like intertriginous plaques
- Velvety palms
Clinical impression: differential diagnosis

• Cutaneous/mucosal papillomas
  – Verruca vulgaris
  – Epidermodysplasia verruciformis
  – Malignancy associated

• Acanthosis nigricans-like intertriginous plaques
  – Acanthosis nigricans (AN)
    • Metabolic disease
    • Syndromic AN
    • Autoimmune AN
    • Familial
    • Malignancy associated
  – Confluent and reticulated papillomatosis
  – Intertriginous granular parakeratosis

• Velvety palms
Clinical impression: differential diagnosis

- Cutaneous/mucosal papillomas
  - Verruca vulgaris
  - Epidermodysplasia verruciformis
  - Malignancy associated
- Acanthosis nigricans-like intertriginous plaques
  - Acanthosis nigricans (AN)
    - Metabolic disease
    - Syndromic AN
    - Autoimmune AN
    - Familial
    - Malignancy associated
  - Confluent and reticulated papillomatosis
  - Intertriginous granular parakeratosis
- Velvety palms
  - Malignancy associated
  - Severe form of acrokeratosis verruciformis of Hopf
Impression & Plan

• Strongly suspecting malignancy associated process
• Biopsy to rule out HPV
• Initiate laboratory and systemic malignancy work up
Plan

• Ruled out HPV
• Initiate search for underlying malignancy
Plan

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  - CT of chest, abdomen, pelvis unrevealing
  - CBC with absolute eosinophils 1.09
Plan

• Ruled out HPV
• Initiate search for underlying malignancy
  – CT of chest, abdomen, pelvis unrevealing
  – CBC with absolute eosinophils 1.09
    • Positive Schistosoma IgG 0.38
    • Tx: Praziquantel 12/2015

• Referred to oncology and gastroenterology
Further workup and management

- Ruled out HPV
- Initiate search for underlying malignancy
  - CT of chest, abdomen, pelvis unrevealing
  - Upper endoscopy and colonoscopy with normal biopsies
  - CBC with absolute eosinophils 1.09
    - Positive Schistosoma IgG 0.38
    - Tx: Praziquantel 12/2015
- Symptomatic referrals:
  - Ophthalmology, otolaryngology
- Multidisciplinary approach to care:
  - Oncology following q 6 months
  - GI with negative scopes
  - Urology consult secondary to positive Schistosomiasis
• Ongoing malignancy surveillance for 9 months was negative
Acanthosis Nigricans

Florid cutaneous (mucosal) papillomatosis

Tripe palms
Initial presentation

4 month followup
Acitretin 50 mg/day
Initial presentation

4 month followup
Acitretin 50 mg/day
Urologic evaluation

• 5/20/16 cystoscopy
  – Occult papillary tumor at the left trigone
  – **Not identified on prior CT C/A/P**
  – Bx = papillary urothelial carcinoma, low grade
  – Mitomycin D intraoperative treatment

• 11/30/16: persistent tumor → OR fulguration

• 12/22/16: BCG instillation q6weeks

• 3/2017: Followup cystoscopy planned
6 weeks status-post intraoperative mitomycin D injection of low-grade papillary urothelial carcinoma
Initial presentation

6 weeks status-post mitomycin C
Discussion

• Florid cutaneous papillomatosis
  – Also known as Schwartz-Burgess Syndrome
  – Most commonly associated with gastric adenocarcinoma

• Malignant acanthosis nigricans
  – Most commonly associated with gastric adenocarcinoma

• Tripe palms
  – GI (30%), lung (20%)
Discussion

• Bladder urothelial carcinoma is also known as transitional cell carcinoma (TCC)
• Most common bladder cancer in Europe and USA
• Risk factors: cigarette smoking, chemical and environmental exposures
• Association between *S. haematobium* and bladder cancer
  – Most commonly high-grade SCC > noninvasive TCC
Discussion

• Review of the literature:
  – Malignant AN + tripe palms + papillomatosis = rare
  – Reported in renal urothelial carcinoma however never in bladder carcinoma

• The longest duration to diagnosis previously reported was 2 years

• The stimulating factor is hypothesized to be a substance secreted by the tumor
  – Transforming growth factor (TGF)-alpha is structurally similar to epidermal growth factor

• FGFR3 has been shown to be mutated in more than 30% of bladder TCC which is also mutated in AN
Teaching points

• Florid mucocutaneous papillomatosis, acanthosis nigricans, and tripe palms occurring in association with bladder urothelial carcinoma
• Rare to not have rapid onset
• Highlights possibility of oral retinoids and palliative surgery for treatment of papillomatosis
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References


