Paraneoplastic Mucous Membrane Pemphigoid, Not Your Typical Cause of Odynophagia

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Introduction

- Mucous Membrane Pemphigoid (MMP) is a rare inflammatory autoimmune disorder that primarily affects the oral, ocular and aerodigestive mucous membranes.
- IgG, IgA, and C3 autoantibodies target basal membrane zone causing subepithelial blisters with erosive lesions in the mucosa.
- It is rarely seen in the clinical setting, with an incidence of 2-10 cases in 100,000 individuals.
- High clinical suspicion with early diagnostic testing is warranted due to its strong association with underlying paraneoplastic disease.

Case Presentation

- An 88 year old female presented with a 5 month history of worsening odynophagia, dysphagia to solids and liquids, poor oral intake and a fifty pound weight loss.
- Physical examination was remarkable for diffuse erosive lesions of the oral mucosa, tense bullae on the soft palate, and bilateral temporal symblepharon along with inflammation of upper and lower eyelids.
- Laboratory workup demonstrated elevated Carbohydrate Antigen (CA)-125 antigen (574.20 U/mL).

Case Presentation

- Chest-abdominal-pelvic computed tomography scan demonstrated an irregular enhancing right adnexal mass, intra-abdominal lymphadenopathy, left hemiabdomen omental metastatic deposits and bilateral sub-centimeter pulmonary nodules.
- Computed tomography guided core biopsy of a left omentum deposit was remarkable for primary ovarian metastatic high grade serous carcinoma.
- Immunofluorescence study of biopsies from the left inferior conjunctiva and right lateral tongue lesions were remarkable for IgG, IgM, IgA inter-keratinocytic deposits at lower level of the epidermis and C3 linear deposits at the dermo-epidermal junction consistent with Paraneoplastic Mucous Membrane Pemphigoid.
- Due to refractory odynophagia and dysphagia, intravenous Solumedrol was initiated with subsequent improvement of symptoms.
- Patient was discharged with multidisciplinary team follow up for chemotherapy initiation and Rituxan induction.

Imaging

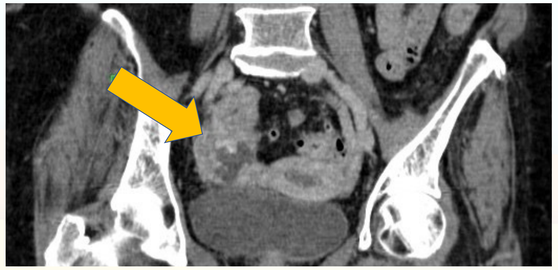

Figure 1: Irregular enhancing right adnexal mass

Conclusion

- Paraneoplastic MMP is an uncommon autoimmune mucocutaneous disease diagnosed when there is presence of stomatitis, histologic features of acantholysis or interface dermatitis, demonstration of anti-plakin autoantibodies on biopsy, with an underlying neoplasm.
- Less than 500 cases have been reported where clinical features, presentation, and autoantibodies profiles have varied widely between patients highlighting the challenge of diagnosis.

Take Home Message

- Recognition of Paraneoplastic MMP presenting as odynophagia is difficult due to the overlap of symptoms in more prevalent and benign conditions.

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