Ecthyma gangrenosum of the sinonasal tract mimicking acute invasive rhinosinusitis in an immunocompromised patient

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ABSTRACT

OBJECTIVE: Ecthyma gangrenosum [EG] is an uncommon cutaneous necrotizing infection classically associated with *Pseudomonas aeruginosa* bacteremia. Only one case of sinonasal ecthyma gangrenosum has been reported to date. Here, we present two additional cases of sinonasal ecthyma gangrenosum with unique, previously unreported features.
STUDY DESIGN: Case series.
METHODS: Retrospective review of clinical cases.
RESULTS: Patient 1. A 35-year-old female on tacrolimus and prednisone 3 months following peripheral blood stem cell transplant for acute lymphoblastic leukemia was evaluated for acute-onset fever, left-sided facial pain and swelling, without visual changes or facial dysesthesia. Nasal endoscopy revealed an eschar within the middle meatus. Biopsy showed no fungal elements. Due to concern for acute invasive fungal rhinosinusitis, the patient underwent endoscopic and open sinonasal debridement of the necrotic tissue. Histopathologic analysis showed no fungal elements. *Pseudomonas aeruginosa* was isolated. The patient improved following debridement and therapy with intravenous tobramycin and cefepime.
Patient 2. A 20-year-old male on tacrolimus after bone marrow transplant for chronic myelocytic leukemia presented with headache, facial swelling, and altered mental status. The patient had asymmetric facial swelling, with dark mucoid nasal discharge. The patient was taken to the operating room for concern for invasive fungal rhinosinusitis. Operative findings included extensive pallor and eschar in the uncinate process, maxillary sinus, and nasal septum. Repeat debridement was necessary. Tissue culture demonstrated *Pseudomonas aeruginosa*. The patient showed significant improvement following ceftazadime therapy initiation.
CONCLUSION: A diagnosis of sinonasal EG should be considered in an immunocompromised patient with an acute necrotizing rhinosinusitis in whom biopsies fail to show fungal elements.

BACKGROUND

Ecythyma gangrenosum [EG] is a clinical syndrome characterized necrotizing cutaneous lesions. Though it is most commonly reported in the gluteal and perineal regions, only one case has previously reported in the sinonasal mucosa. Here we present two cases of sinonasal echythma gangrenosum in immunocompromised patients. Though the clinical presentation of EG may overlap with those of acute invasive fungal rhinosinusitis (AIFRS), early consideration and appropriate treatment may limit the morbidity of aggressive debridements.

CASE 1 REPORT

A 35-year-old female with history of peripheral blood stem cell transplant for acute lymphoblastic leukemia, recent acute graft-versus-host disease, and psuedomonal septicemia was evaluated for the acute onset of fever, severe left-sided hemifacial pain with swelling. Nasal endoscopy demonstrated a large eschar within the left middle meatus, and the maxillary sinus antrum had an abnormally widened appearance (**Figure 1**). CT demonstrated scattered opacification of the paranasal sinuses bilaterally with demineralization left ethmoid labyrinth and dehiscence of the posterolateral wall of the left maxillary sinus (**Figure 2**).

Intraoperative endoscopy of the left nasal cavity revealed frank necrosis of the inferior turbinate and lateral nasal wall mucosa, enlarged maxillary sinus antrum, and pale, poorly vascularized maxillary sinus mucosa. A decision was made to proceed with aggressive resection. Final histopathologic assessment showed a mixture of non-inflamed respiratory mucosa and mucosa showing necrotizing inflammation. Methenamine silver stains were negative for fungi. Gram stain did not identify bacteria; however, multi-drug resistant *Pseudomonas aeruginosa* was isolated from tissue cultures. The patient showed improvement with continued antipseudomonal antibiotics.

CASE 2 REPORT

This is a 20 year-old male with a history of chronic myelocytic leukemia, who underwent allogeneic stem cell transplantation complicated by graft versus host disease, who presented with worsening headache and facial swelling. Approximately one month prior to this encounter, he was admitted for fever and pseudomonas aeruginosa bacteremia. On initial otolaryngology consultation, he reported symptoms of dark, mucoid rhinorrhea, nasal congestion and rhinorrhea.

Physical exam was notable for dark mucoid rhinorrhea and edematous nasal mucosa. Nasal endoscopy showed for bleeding mucosa and copious mucoid secretions, but was limited by the cooperation of the patient. CT showed opacification of bilateral ethmoid, sphenoid, and maxillary sinuses, as well as infiltration of retroantral fat posterior to the left maxillary antrum (Figure 4). Final histopathologic assessment showed a mixture of non-inflamed respiratory mucosa and mucosa showing necrotizing inflammation. Methenamine silver stains were negative for fungi. Multi-drug resistant *Pseudomonas aeruginosa* was isolated from tissue cultures. The patient showed significant clinical improvement with appropriate antibiotic therapy.

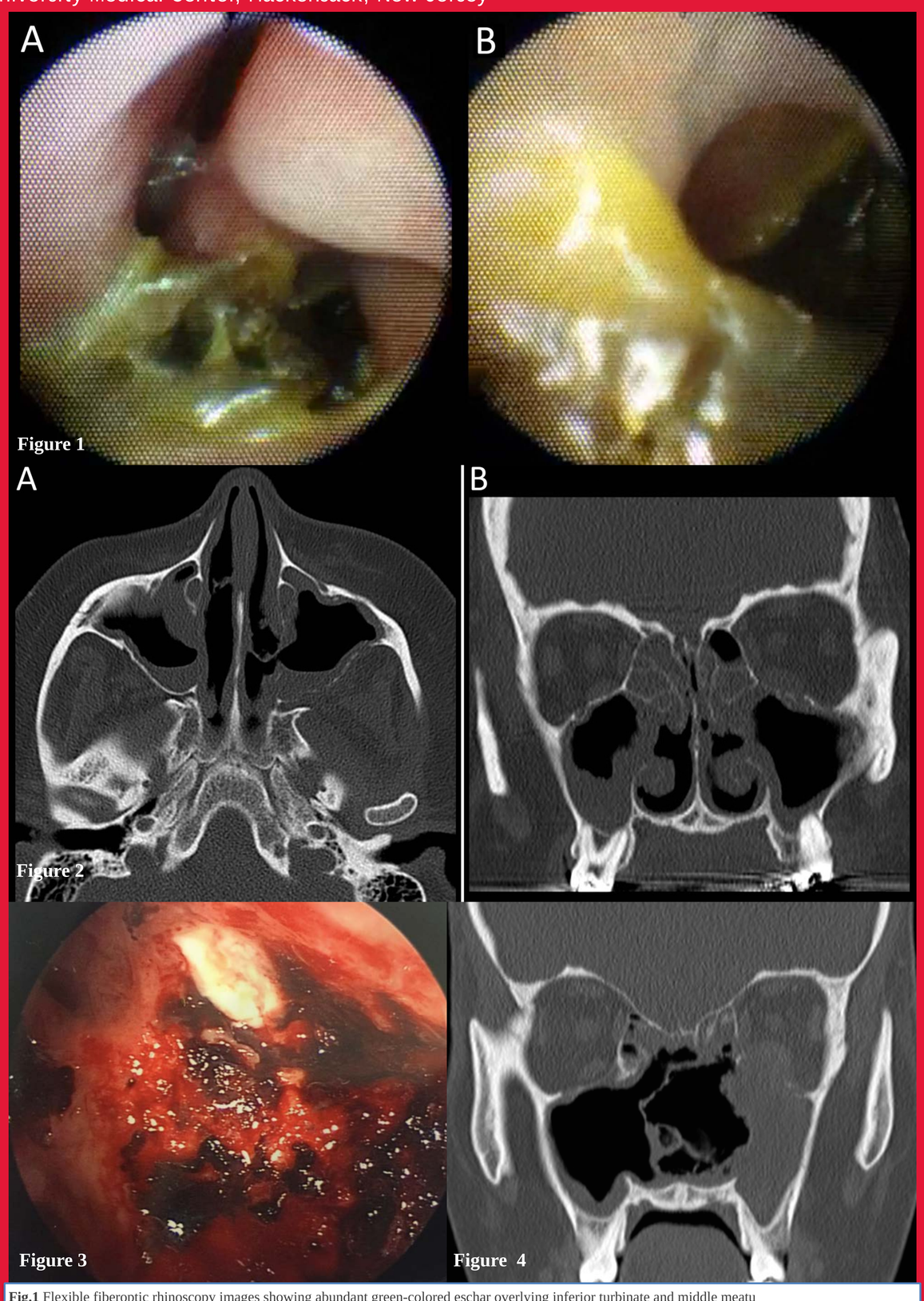

Fig.1 Flexible fiberoptic rhinoscopy images showing abundant green-colored eschar overlying inferior turbinate and middle meatu
Fig. 2(A) Axial computed tomography (CT) in showing demineralization and dehiscence of the left posterior maxillary sinus wall. (B) Coronal reconstruction showing opacification of the bilateral maxillary and ethmoid sinuses.
Fig 3. Intraoperative endoscopy showing extensive necrosis and bony erosion of the left maxillary sinus
Fig 4. Coronal CT imaging showing soft tissue opacification of the left maxillary sinus with destruction through the left orbital floor.

DISCUSSION

EG may arise in as many as 1- 6% of immunocompromised patients with recent history of pseudomonal sepsis.[2,3] The disease system manifests as cutaneous lesions that evolve from maculopapular eruption to hemorrhagic bullae to eschar, corresponding to invasion of the agent through the adventitia and media of involved vasculature.[4]
Although EG classically associated with pseudomonal sepsis, recent reports have linked it a number of other bacteria and fungi.[2] A non- septicemic variant has been described for EG which carries a substantially lower mortality rate.[5]
Sinonasal EG has been reported once in the English language literature by Hekiert, et al.[1] Each reported case thus far has occurred in the setting of iatrogenic immunosuppression. Distinctive in these reports are evidence of bony destruction, a feature which may further which may further obfuscate the diagnosis of sinonasal EG. In addition, one of our patients presented eight months later with recurrent disease, suggesting an indolent yet destructive element to this disease, requiring diligent postoperative surveillance.

CONCLUSION

EG an uncommon necrotizing vasculitis that can affect the sinonasal tract. Here we present two cases affecting the sinonasal tract, and document for the first time its ability to cause bony erosion. The disease appears to respond to targeted antiinfective therapy, and may be amenable to more conservative debridement than acute invasive fungal rhinosinusitis.

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