Endonasal Dacryocystorhinostomy in Wegener Granulomatosis

Wegener granulomatosis (WG) is a life-threatening autoimmune disease of unknown etiology first described in 1936. The classic clinical triad consists of necrotizing granulomatous inflammation of the respiratory tract, necrotizing glomerulonephritis, and systemic vasculitis. Prior to the introduction of modern immunosuppressants, WG was usually fatal due to renal complications. Involvement of the eye and adnexal structures is common, occurring in 29% of patients. Nasolacrimal duct obstruction (NLDO) following necrosis of the nasal tissue is seen in 7% of patients, who may seek relief through lacrimal surgery.

Dacryocystorhinostomy (DCR) has been shown to be a safe, effective intervention but failures and complications are more common in this patient group. While conventional teaching recommends an external approach, increasing use of the endonasal approach (EN-DCR), with or without an endoscope, raises the question of whether it can be used safely and successfully in WG. We present 4 cases treated endonasally (cases 1 and 2 using endoscopy by D.S., cases 3 and 4 using direct visualization by P.J.D., customary for each surgeon) and offer further advice on management of this patient group.

Report of Cases. Case 1. A 54-year-old woman, diagnosed 4 months previously with WG and treated with prednisolone and cyclophosphamide, presented with a 2-year history of right epiphora and recurrent episodes of acute dacryocystitis. Syringing and dacryocystogram showed complete NLDO. Computed tomographic scan demonstrated destruction of the ethmoids, medial wall of the left maxilla, and anterior cribiform plate with a narrow nasal cavity. Surgery was deferred until her disease was controlled and 20 months later endoscopic EN-DCR was performed, with Ear, Nose, and Throat assistance because of grossly disturbed anatomy. The lacrimal bone was eroded and there was dense scar tissue adjacent to the nasolacrimal sac; perioperative prednisone (1 mg/kg tapered over 4 weeks) was administered. The patient was asymptomatic following surgery and the ostium was patent at 14 months using the endoscopic dye test.

Case 2. A 49-year-old woman presented with a 12-month history of left epiphora and a preceding episode of acute dacryocystitis and orbital cellulitis. She had known WG for 2 years associated with severe sinusitis and a stricture in the left main bronchus and was treated with prednisolone and azathioprine. She had a saddle nose deformity due to destruction of the nasal bridge. Syringing showed complete NLDO and computed tomographic scan showed a deviated septum but good resolution of sinus disease. Left endoscopic EN-DCR and septoplasty were performed leading to relief of epiphora and a patent ostium on the endoscopic dye test at 12 months.

Case 3. A 47-year-old woman presented with 6 months of left epiphora complicated by 2 episodes of dacryocystitis, once requiring abscess drainage. She had a 10-year history of WG controlled with cyclophosphamide treatment; her last clinical recurrence had been 2 years previously and results of a recent antineutrophil cytoplasmic antibody test were negative. Left nonendoscopic EN-DCR was performed and she was symptom free and irrigated freely 6 months later. At that time, she had developed right NLDO and an EN-DCR was per-
formed on that side. A year later, she developed recurrent right epiphora during a flare-up of her WG, with granulation tissue obscuring the original ostium. Once this was controlled medically, a repeated EN-DCR was performed and she has been epiphora free for 5 years.

Case 4. A 49-year-old man presented with left optic neuropathy and apical compression. His computed tomographic scan showed a left orbital apical mass lesion (Figure 4) and WG was diagnosed from the biopsy specimen. Although visual acuity returned with systemic prednisolone and methotrexate treatment, he developed right NLDO 3 months later. Once the disease was deemed to be quiescent 4 months later, a right nonendoscopic EN-DCR was performed with insertion of Crawford tubes. These were removed 3 months later and the patient remained asymptomatic 9 months following surgery.

Comment. These cases demonstrate the complexities of managing NLDO in patients with WG, similar to previous case series, and that EN-DCR can be used successfully in this group. However, various arguments have been raised against EN-DCR in WG, including disease reactivation, ostium size, bleeding, and endonasal navigation.

In terms of disease reactivation, there is less tissue disruption endonasally, so the risk of postoperative inflammation is potentially lower than with an external approach. Apart from case 1, no perioperative steroids were administered be-
cause surgery was performed when the disease was in remission.

While ostium size may be smaller using an endonasal approach, this should not increase failure risk, since WG reactivation would threaten ostia of any size. Indeed, external DCR requires more tissue dissection in the diseased area, which may increase the risk of postoperative inflammation and external scar complications, such as fistula formation and wound necrosis. The smaller endonasal ostium may help prevent complications, such as granulomas extending into the orbit through the rhinostomy.

Another possible disadvantage of the endonasal approach is bleeding from inflamed nasal mucosa obscuring the view, making the surgery more technically challenging and causing epistaxis and scarring. Inflamed nasal mucosa (with hyperplasia, erythema, and possible petechiae) suggests active disease and the surgery should be avoided using either approach unless the systemic disease is under control, as advised by the internist based on systemic and laboratory findings. In our experience, the scarred, necrotic mucosal tissue bleeds less.

While recent radiologic navigating systems might be considered in cases where anatomic landmarks are absent, the standard vitrectomy light pipe inserted via the superior canaliculus into the nasolacrimal sac used in all 4 cases eliminated any possible source of confusion and acted as a target for the surgeon allowing safe access to the lacrimal sac.

Patients with WG should be warned about the potential reactivation of their disease and that the risk of failure is higher because of the unpredictable nature of their postoperative inflammation. We recommend preoperative imaging, that an experienced surgeon perform the operation, and that systemic disease is quiescent at the time of surgery; therefore, a multidisciplinary approach is also advocated. We would encourage readers to consider the use of EN-DCR in patients with WG as an equal, and possibly superior, alternative to the external approach.

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