Management of IgG4-related orbital disease: when not in doubt, cut it out?

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Most retrospective reviews of the efficacy of treatments for orbital inflammation, whether for idiopathic orbital inflammation (IOI) or for IgG4-related ophthalmic disease (IgG4-ROD) are hampered by the fact that many patients receive multiple treatments, frequently overlapping, and often at different dosages and durations. The natural course of the disease is thereby modified in myriad ways, making an accurate assessment of the treatment modalities difficult. Once a treatment has been accepted as the best first-line therapy (corticosteroids in this case), the physician becomes inclined to his purpose to the extent that, to date, there are no well-designed prospective studies of these two conditions where a single treatment has been assessed with defined treatment, dosage duration and outcomes.

Although a retrospective study, Ominato et al. present 15 cases (8 unilateral and 7 bilateral) of IgG4-ROD treated solely with surgical debulking. The study is of interest because no other treatment before, during or after surgery was administered to any of the patients, thereby allowing the assessment of the results with the natural history of the disease unaltered in any other manner. Twelve of their cases were classified as dacryoadenitis IgG4-ROD and three as orbital fat tissue IgG4-ROD. They removed 70%–100% of the lesion in these cases with a relapse rate of 13.3% after such debulking surgery (with a follow-up range of 6–84 months). Seven of their 12 cases of dacryoadenitis had bilateral disease, and 6 of them underwent bilateral debulking surgery. Specifically, they noted that in the 12 operated dacryoadenitis cases involving 18 operated sides, they removed 90% or more of the lesions in 17 sides and 70% on the remaining one side. In the three cases involving the orbital fat, they claimed to ‘completely extirpate the lesion’.

Preoperative tear function measurements were not measured in their patients; they noted that seven of their patients with dacryoadenitis developed dry eyes postoperatively, controllable with drops and with no impairment of vision. They noted no complications with the three orbital cases. There was no difference in serum IgG4 levels between patients with and without recurrence. Two cases had a relapse after a resection of the lesions: one had recurrence of swelling at 6 months after surgery with CT scan evidence of an increase in the lacrimal gland lesion. Further resection, which again confirmed the presence of IgG4-ROD, led to further recurrence. Subsequent management is not presented. The second case did not develop any symptoms or demonstrate signs, but they noted that on a follow-up CT scan 17 months later, there was further lacrimal gland swelling. They noted that there was spontaneous regression of this swelling without further treatment. Since there was no other treatment administered in these cases, the study gives a glimpse into the natural history of these lesions following surgery alone. Since the two most common causes of orbital inflammation are IOI and IgG4-ROD, and even though they may not be entirely cognate, the questions raised by this paper should be discussed in relation to both this conditions:

1. Why is surgical resection of an inflammatory mass (IOI dacryoadenitis and IgG4-ROD dacryoadenitis or IgG4-ROD orbital disease) an option?
2. What are the results of surgical removal of lesions in IOI dacryoadenitis?
3. What other evidence is there to support surgical resection of lesions in IgG4-ROD dacryoadenitis or IgG4-ROD orbital disease?
4. How much of the inflammatory mass should be removed?
5. Why and how does such debulking surgery help?
6. How much dryness do patients have when presenting with either type of dacryoadenitis?
7. How much lacrimal gland tissue is necessary to prevent dryness after surgery, assuming that the ductules are protected during such surgery?
8. When both lobes of the lacrimal gland are affected, is it necessary to extirpate both lobes?
9. What is the natural course of IOI dacryoadenitis and IgG4-ROD dacryoadenitis?
10. Is there evidence of functional recovery of the lacrimal gland after successful medical treatment of IOI dacryoadenitis and IgG4-ROD dacryoadenitis?
11. How does one assess a case of IOI dacryoadenitis and IgG4-ROD dacryoadenitis so as to allow a reliable way of determining outcomes of treatment?

Answers to some of these questions may be gleaned from a careful and detailed analysis of published studies. Others await properly designed studies by inquiring minds.

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