IgG4 related ophthalmic disease of the Caruncle

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Dear Editor,

Immunoglobulin G4 (IgG4)-related disease is a rare systemic disease that affect various organs with elevated serum IgG4 levels and marked IgG4-positive lymphoplasmacytic infiltration. [1] In the field of ophthalmology, IgG4-related ophthalmic disease (IgG4-ROD) is most commonly found in the lacrimal gland, but also found in diverse ocular adnexal tissues such as the extraocular muscles, infraorbital nerve, and eyelid. [2] IgG4-ROD of the caruncle has not been described previously. The authors describe a patient with IgG4-ROD of the caruncle along with literature review.

A 41-year-old male patient presented with a mass in the left caruncle. The lesion was noted to have been present for a period of 2 weeks. Examination revealed a single, asymptomatic, round pinkish protruding mass in the left caruncle (Fig.1). There were no complaints of pain, discharge or bleeding. The patient's medical history and family history were unremarkable and a systemic search found no papules or nodular skin lesions. Magnetic resonance imaging revealed no abnormal mass lesion in the orbit. Under local anesthesia, the lesion was excised completely. On gross examination, it was a 0.3 × 0.3 × 0.4 cm mass. Histopathologic examination revealed nodular lymphoid proliferation with follicular hyperplasia pattern composed of several hyperplastic follicles and increased IgG4 positive cells (>300/HPF, IgG4/CD138 ratio >40%) (Fig.1). Serum IgG4 level was not elevated. It was diagnosed as possible IgG4-ROD of caruncle with progressively transformed germinal center type of lymph nodes. [1,3]

IgG4-related disease (IgG4-RD) is characterized by infiltrate of IgG4 positive plasma cells and elevated serum IgG4 levels, which can affect various organs. [1] In general, histopathological features of IgG4-RD are a dense lymphoplasmacytic infiltrate, storiform-type fibrosis and obliterative phlebitis. However, in organs such as the lymph node, lung, minor salivary glands, and lacrimal glands, storiform-type fibrosis and obliterative phlebitis usually absent. [3] IgG4-ROD is a condition that IgG4-RD occurs in the ocular adnexal organs and most commonly affect the lacrimal gland, followed by trigeminal nerve branch, extraocular muscles, and orbital fat tissue. [2] However, it has never been reported in the caruncle.

The caruncle contains skin elements, conjunctiva and lacrimal glands, which can lead to a variety of disease entity. A case series of the caruncular lesion, lymphoid lesions were reported to be rare, with a frequency of about 3%. [4] However, since many ocular adnexal lesions, previously diagnosed as
lymphoid hyperplasia or idiopathic orbital inflammation, have been re-diagnosed as IgG4-ROD. In conclusion, we present the first case of possible IgG4-ROD in the caruncle. Although rare, IgG4-ROD should be considered in the differential diagnosis of a caruncle mass.

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Reference


Figure legend

Figure 1. Clinical photograph of the left caruncular lesion (arrow) (A). A single, round pinkish protruding mass was observed in the caruncle (B). Histologic features of IgG4-related disease of the caruncle (C-J). Low power view shows nodular lymphoid proliferation with follicular hyperplasia pattern composed of several hyperplastic follicles (H&E) (C). Higher magnification reveals reactive hyperplastic follicles containing frequent plasma cells (D). CD3 and CD20 immunostaining show relatively preserved B and T zones (E) & (F). CD138+ plasma cells are shown within hyperplastic follicles (G), many plasma cells are positive for IgG4 (>300 cells/HPF; IgG4/CD138 ratio >40%) (H). Kappa and lambda light chain staining shows no evidence of light chain restriction with slight kappa-dominance (I) & (J). IgH gene rearrangement study revealed no evidence of B cell monoclonality (not shown).