Immunoglobulin G4-related Disease in the Head and Neck Region – Pictorial Review with Radiological-Pathological Correlation

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Objective:

Immunoglobulin G4-related Disease (IgG4-RD) is an immune-mediated fibroinflammatory systemic disease affecting every organ system. It is characterized by enlargement of affected organs caused by lymphoplasmacytic infiltration with a predominance of IgG4-positive plasma cells. This pictorial review presents the radiological and pathological features of IgG4-RD in the head and neck region.

Materials and Methods:

A total of 10 cases of IgG4-related disease with head and neck involvement from the database of our center from 2015 to 2019 were included. Retrospective review of the radiological and histopathological features was performed.

Radiological Features

The main radiological features of IgG4-RD in the head and neck region are divided into orbital and salivary gland involvement.

Salivary Glands

Submandibular Gland

In our review, five patients had submandibular gland involvement, four of whom demonstrated evidence of chronic sialadenitis (Fig. 1a) and one had submandibular nodules (Fig. 1b). Histology sections of chronic sialadenitis are shown in Fig.2.

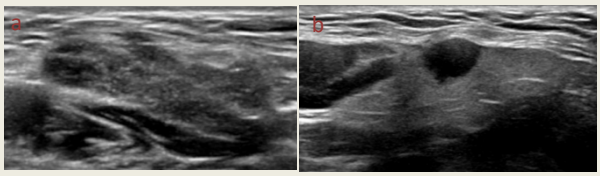

Fig 1. (a) Ultrasound scan (USG) of a submandibular gland, showing lobulated outline and heterogeneous parenchymal echo, compatible with chronic sialadenitis. (b) USG showing a submandibular nodule.

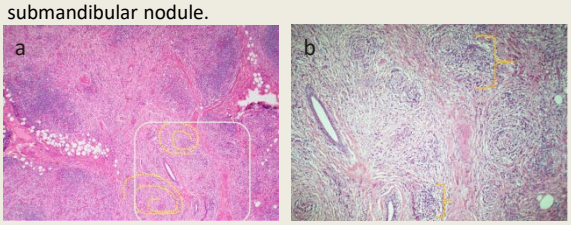

Fig 2. Histology sections of a left submandibular gland showing (a) prominent fibrosis in vague storiform pattern (orange dotted lines). H&E, x 4. (b) dense plasmacytic infiltrate (orange brackets). H&E, x 10.

Parotid Gland

Seven patients had parotid involvement, six of whom demonstrated nodules in the parotid gland (Fig. 3), and two had parotid enlargement.

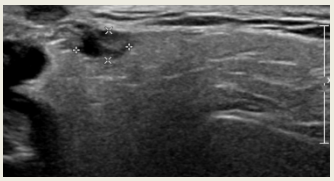

Fig 3. USG showing a nodule in the right parotid gland remnant of a patient who presented with right parotid mass. Excision of the right parotid mass was performed, with the histology shown in Fig.4

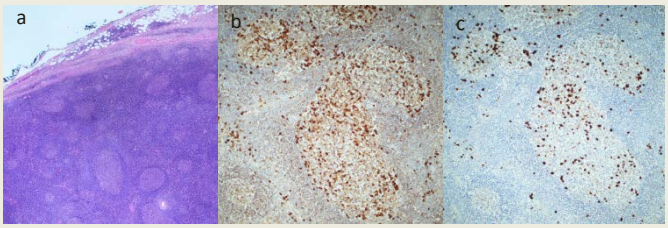

Fig 4. (a) Sections of a right parotid mass show a lymph node with reactive lymphoid follicles. Tingible body macrophages and mild increase of plasma cells are noted in the germinal centers. H&E, x 22.5. (b & c) Absolute count of IgG4 positive cells is 80 per high power field and the majority of IgG-positive plasma cells (b) appear positive for Ig4 (c). (b) IgG immunostaining and (c) IgG4 immunostaining, both at x 100.

Orbit

Lacrimal Gland

The lacrimal glands are the most commonly affected by IgG4-related disease in the orbit. There were 5 cases of orbital involvement, all of them demonstrated bilateral lacrimal gland enlargement (Fig. 5 &6).

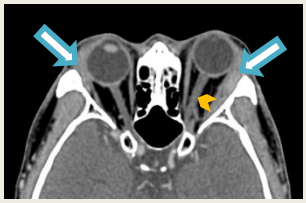

Fig. 5. Orbital CT demonstrates enlargement of the bilateral lacrimal glands (blue arrows). Left optic nerve (orange arrowhead) is also slightly prominent in caliber compared to the right side.

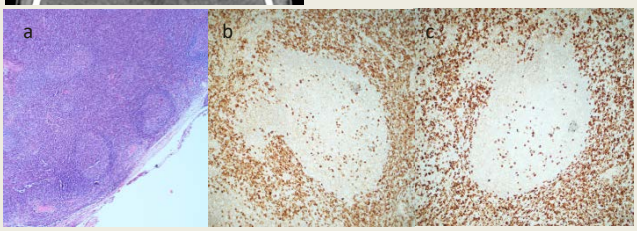

Fig 6. (a) Sections of a left orbit mass show numerous reactive lymphoid follicles with germinal centers containing tingible-body macrophages and prominent plasmacytosis between the follicles. H&E, x 50. (b & c) IgG4 positive cells are 200 per high power field and 90% of IgG-positive plasma cells (b) appear positive for Ig4 (c). (b) IgG immunostaining and (c) IgG4 immunostaining, both at x 100.

Extraocular Muscles, Infraorbital Nerve and Optic Nerve

Two patients displayed extraocular muscles enlargement (Fig. 7a-b), one of whom had bilateral involvement. One patient also had bilateral infraorbital nerves (Fig. 7a & c) and left optic nerve thickening (Fig. 5). Infraorbital nerve involvement has been reported to be a specific sign for IgG4-RD.

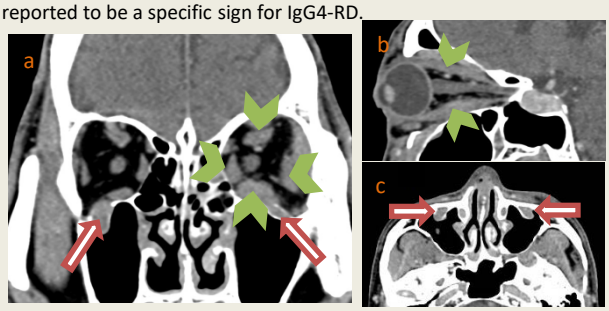

Fig 7. Orbital CT demonstrates enlargement of the left inferior, superior, lateral and medial rectus muscles (green arrowheads). Infraorbital nerves are also thickened bilaterally in this patient (red arrows).

CONCLUSION:

Familiarization and timely recognition of the imaging features of IgG4-RD in the head and neck region can facilitate early diagnosis and treatment.