

Complete Treatment Response of Isolated Lacrimal IgG4-Related Disease on ¹⁸F-FDG PET/CT Imaging

İzole Lakrimal IgG4 İlişkili Hastalıkta 18F-FDG PET/BT'de Tam Tedavi Yanıtı

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Abstract

We describe a case of a 57-year-old woman with isolated immunoglobulin G4-related disorder affecting her lacrimal glands. Pre-treatment ¹⁸F-fluorodeoxyglucose-positron emission tomography/computed tomography (¹⁸F-FDG PET/CT) showed diffuse enlargement and increased FDG uptake in both lacrimal glands. After immunosuppressive treatment, ¹⁸F-FDG PET/CT showed no significant FDG uptake, and the sizes of both glands returned to normal.

Keywords: Lacrimal, IgG4, FDG, PET, treatment

Öz

57 yaşında, izole lakrimal immünoglobulin G4 ilişkili hastalık tanısı almış bir kadın olgusu sunuyoruz. Tedavi öncesi yapılan, ¹⁸F-florodeoksiglukoz pozitron emisyon tomografisi/bilgisayarlı tomografi (¹⁸F-FDG PET/BT), her iki lakrimal bezde de yaygın boyut artışı ve FDG tutulumu göstermiştir. İmmünosüpresif tedavi sonrası yapılan ¹⁸F-FDG PET/BT'de anlamlı bir FDG tutulumu izlenmemiş olup her iki bezin boyutları normale dönmüştür. **Anahtar Kelimeler:** Lakrimal, IgG4, FDG, PET, tedavi

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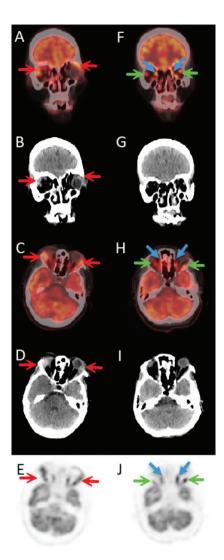


Figure 1. Immunoglobulin G4-related disease (IgG4-RD) is an immune-mediated, chronic inflammatory disorder that can affect multiple organ systems (1,2). The pathogenesis involves lymphocytic infiltration of tissues with subsequent secretion of IgG4 (3). The most commonly affected sites include the orbits, lacrimal and salivary glands, pancreas, lungs, biliary ducts, kidneys, aorta, and retroperitoneum (4,5,6). Due to its varied presentation, IgG4-RD can be mistaken for malignancy, infection, or other autoimmune diseases, necessitating a comprehensive evaluation encompassing clinical, radiological, and pathological data for accurate diagnosis (6).

¹⁸F-fluorodeoxyglucose-positron emission tomography/computed tomography (¹⁸F-FDG PET/CT) is a valuable imaging modality for identifying increased metabolic activity in affected organs, guiding biopsy sites, and assessing therapeutic response (7,8,9). Various studies have demonstrated the utility of ¹⁸F-FDG PET/CT in evaluating treatment response in IgG4-RD (8,9,10,11). Rare manifestations, such as IgG4-related sclerosing mesenteritis and sacroiliac involvement, have also been documented in the literature (12,13). ¹⁸F-FDG PET/CT is particularly beneficial in identifying the extent of multiorgan involvement.

We report a case of IgG4-RD diagnosed by lacrimal gland biopsy. The patient presented with eyelid swelling and elevated inflammatory markers.

18F-FDG PET/CT was performed to assess systemic involvement, revealing increased FDG uptake and enlargement of both lacrimal glands, more pronounced on the right, without evidence of involvement of other organs.

The patient underwent corticosteroid and rituximab therapy. A follow-up ¹⁸F-FDG PET/CT conducted six months post-treatment demonstrated normalization of the lacrimal gland size and the absence of FDG uptake, indicating a favorable therapeutic response. To our knowledge, this is the first ¹⁸F-FDG PET/CT image of isolated lacrimal IgG4-RD demonstrating its complete response to treatment.

This case underscores the significance of ¹⁸F-FDG PET/CT in evaluating treatment response in IgG4-RD. Pre-treatment coronal fusion (A), coronal CT (B), axial fusion (C), axial CT (D), and axial PET (E) images demonstrate increased size and FDG uptake in the bilateral lacrimal glands, more prominent on the right (red arrows). Post-treatment coronal fusion (F), coronal CT (G), axial fusion (H), axial CT (I), and axial PET (J) images show normalization of lacrimal gland size and absence of pathological FDG uptake. Physiological uptake in lateral rectus (green arrows) and medial rectus (blue arrows) muscles can also be seen.

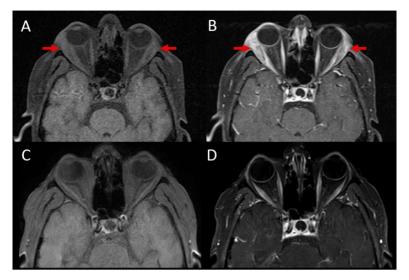


Figure 2. On pre-treatment magnetic resonance images, T1-weighted non-contrast images (A) demonstrated enlargement of both lacrimal glands (red arrows), while contrast-enhanced images (B) revealed contrast enhancement of the lacrimal glands. On the post-treatment T1-weighted images (non-contrast, C; contrast-enhanced, D), regression was observed.

Ethics

Informed Consent: Informed consent was obtained from the patient for the use of their imaging data.

Footnotes

Authorship Contributions

Surgical and Medical Practices: A.K., S.A., Concept: A.K., S.A., O.E.Ş., L.U.B., S.S., K.S., H.B.S., Design: A.K., S.A., Data Collection or Processing: A.K., S.A., Analysis or Interpretation: A.K., S.A., O.E.Ş., L.U.B., S.S., K.S., H.B.S., Literature Search: A.K., Writing: A.K.

Conflict of Interest: No conflicts of interest were declared by the authors.

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