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Conjunctival MALT Lymphoma

Utility of FDG PET/CT for Diagnosis, Staging, and Evaluation of Treatment Response

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Abstract: A 67-year-old woman was referred for staging of a mucosa-associated lymphoid tumor lymphoma involving the left conjunctiva. CT scan had shown paravertebral and pelvic masses, and a breast nodule. FDG PET/CT demonstrated moderately increased uptake in the left ocular conjunctiva and confirmed the paravertebral and pelvic masses and the breast nodule. Moreover, abnormal FDG uptake was shown in 2 breast nodules, the flank, the gluteus maximus, and the gastric cardia. The patient received 6 cycles of rituximab-bendamustine chemotherapy with a complete clinical and metabolic response at the 6-month follow-up PET/CT and remained relapse-free without visual acuity problem after a 36-month follow-up.

Key Words: ^{18}F, FDG, PET, ocular lymphoma, MALT, ocular adnexal MALT lymphoma, conjunctival MALT lymphoma

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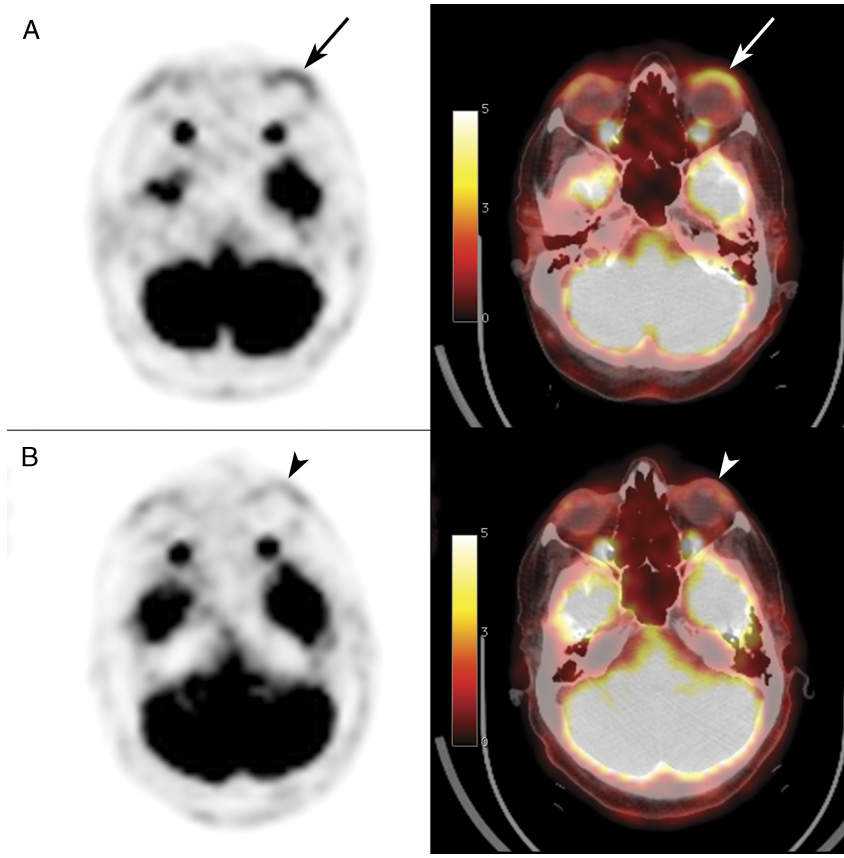

FIGURE 1. **A**, Baseline FDG PET/CT showing increased glucose analog in the left conjunctiva (arrow) as compared with the contralateral side (left-to-right SUV ratio, 3.0). **B**, Control after 3 of 6 cycles of chemotherapy showing an excellent metabolic response with normalized left-to-right SUV ratio to 1.0 (arrowhead).

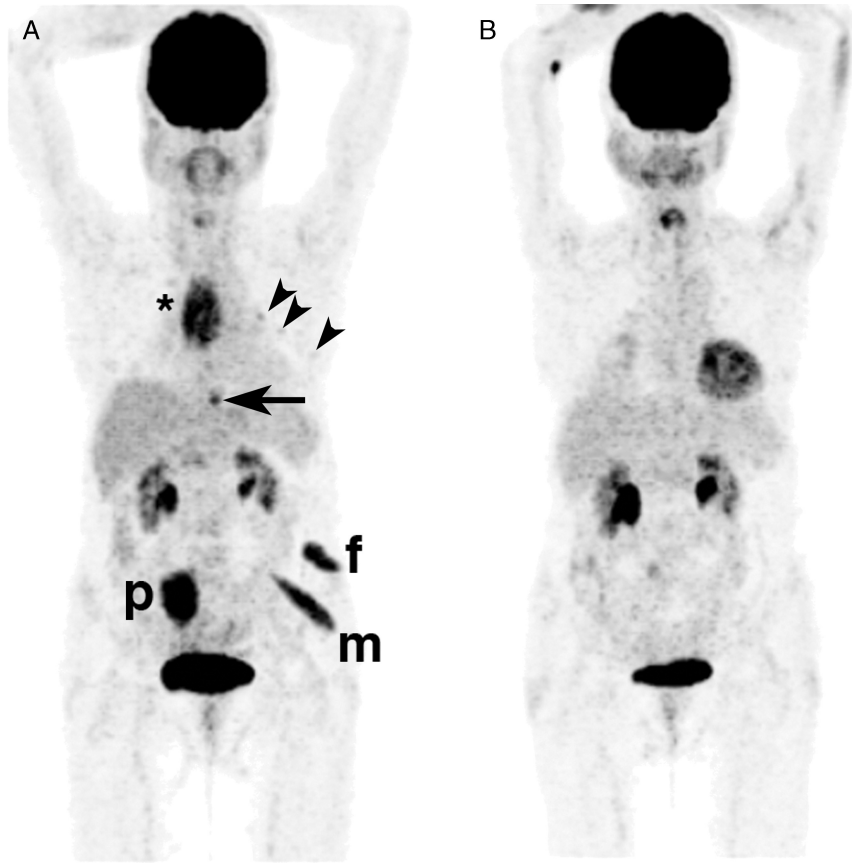

FIGURE 2. **A**, Maximal intensity projection image of the baseline FDG PET showing extraocular lesions with increased pathologic uptake situated in the right paravertebral mass (*), 3 left breast nodules (arrowhead), the gastric cardia (arrow), the left flank (f), the left gluteus maximus muscle (m), and a pelvic paramedian right mass (p). **B**, The same maximal intensity projection image after 3 cycles of chemotherapy showing disappearance of all abnormal FDG uptake sites. The final diagnosis made on a biopsy of the left conjunctiva was a marginal zone lymphoma of mucosa-associated lymphoid tumor (MALT) type (stage III EA). Extranodal marginal zone lymphoma of MALT type is a relatively rare type of B-cell lymphoma accounting for 7% to 8% of non-Hodgkin lymphomas.[1] They arise in the stomach, bowel, lung, ocular adnexa, breast, thyroid, skin, soft tissue, and dura, predominantly in patients older than 60 years.[2] FDG PET/CT has now become part of the diagnostic workup of patients with lymphoma. It is known as being more sensitive than CT for extranodal lesions usually upstaging the disease extent. PET is also now playing a role in the follow-up of lymphoma and during the chemotherapy to assess initial response and determine prognosis. It is known that the conjunctiva is one of the main sites of ocular lymphoma and that lymphomas of conjunctival origin can be detected by FDG PET/CT.[3–5] Likewise, tumoral lesions of the bulbar conjunctiva of other origin can also be seen by FDG PET/CT.[6,7] In the largest up-to-date reported use of PET/CT in ocular adnexal lymphomas, Zanni et al[8] reported 32% of conjunctival lymphomas with FDG PET positivity in only 35% of the cases. In view of the high sensitivity of conventional imaging (CT/MRI) to detect conjunctival involvement, FDG PET does not seem to be particularly useful for establishing the initial diagnosis, besides helping to choose the biopsy site. However, FDG PET can be recommended for staging patients with ocular adnexal lymphomas because of the high frequency of extraocular tumoral sites that can be multiple as in the case of our patient.[3,8] FDG uptake is known to be lower or negative in MALT lymphomas, although this has not been confirmed in ocular adnexal lymphomas.[5] Nevertheless, scintigraphy with somatostatin receptor analogs could be useful in selected cases.[9,10] Indeed, the visualization of a unique lesion directly influences the treatment strategy (surgery, radiation therapy) as compared with a disseminated disease (systemic therapy such as single-agent or combination chemotherapy, immunotherapy with rituximab, monoclonal chimeric anti-CD20 antibodies, or radioimmunotherapy with ^{90}Y ibritumomab tiuxetan [Zevalin]; recently antichlamydial antibiotics therapy have also been proposed as an alternative).[9] The prognosis of MALT lymphoma of conjunctival origin is excellent, and the transformation into a high-grade lymphoma is exceptional.