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A RARE ORBITAL PRESENTATION OF P-ANCA-ASSOCIATED VASCULITIS SUCCESSFULLY TREATED WITH METHOTREXATE AND RITUXIMAB

RIJETKA ORBITALNA PREZENTACIJA P-ANCA-VEZANOG VASKULITISA USPJEŠNO LIJEČENA RITUKSIMABOM I METOTREKSATOM

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Background: ANCA-associated vasculitides (AAV) can occasionally present with atypical and organ- specific manifestations, posing diagnostic and therapeutic challenges. Orbital involvement, particularly mimicking a space-occupying lesion, is a rare manifestation.
Case Presentation: We present the case of a 61-year-old female previously treated for p-ANCA-positive vasculitis with cyclophosphamide pulses. She was referred for ophthalmologic and neurological evaluation due to new-onset oculomotor nerve palsy, left-sided eyelid ptosis, and unilateral vision loss. Orbital MRI revealed a poorly demarcated mass lesion in the region of the left annulus of Zinn, measuring 14 × 11 mm, with proptosis, indistinct borders from the optic nerve, and extension toward the cavernous sinus. Initial management with multiple courses of intravenous methylprednisolone led to transient clinical improvement, but subsequent imaging demonstrated lesion progression. Neurosurgical biopsy was performed, and histopathological analysis was consistent with an inflammatory lesion. The patient was subsequently treated with methotrexate and rituximab, resulting in complete clinical and radiological remission.
Conclusion: This case underscores the importance of a multidisciplinary approach in the evaluation and management of rare orbital manifestations of AAV. Timely collaboration between ophthalmologists, neurologists, neurosurgeons, and rheumatologists/immunologists is crucial for accurate diagnosis and optimal treatment outcomes.

Keywords: p-ANCA vasculitis, orbital mass, oculomotor palsy, rituximab, methotrexate, multidisciplinary approach, case report
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