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Case report

Orbital meningeal melanocytoma: Histological, immunohistochemical and molecular characterization of a case and review of the literature

Andrea Palicelli (M.D.)[a,*], Maria Giulia Disanto (M.D.)[a], Gabriele Panzarasa (M.D.)[b], Claudia Veggiani (B.Sc.)[c], Gabriella Galizia (M.D.)[d], Stefano Dal Cin (M.D.)[d], Elisa Gruppioni (Ph.D.)[e], Renzo Boldorini (M.D.)[a,c]

[a] Department of Health Science, School of Medicine, "Maggiore della Carità" Hospital, University of Eastern Piedmont "Amedeo Avogadro", via Solaroli 17, 28100, Novara, Italy
[b] Neurosurgery Unit, "Maggiore della Carità" Hospital, Corso Mazzini 18, 28100, Novara, Italy
[c] Department of Pathology, "Maggiore della Carità" Hospital, University of Eastern Piedmont "Amedeo Avogadro", Corso Mazzini 18, 28100, Novara, Italy
[d] Diagnostic Radiology Unit, "SS Trinità" Hospital, Viale Zoppis 10, 28021, Borgomanero, Italy
[e] Laboratory of Oncologic and Transplantation Molecular Pathology, Department of Pathology, S.Orsola-Malpighi Hospital, University of Bologna, Via Pietro Albertoni, 15,40138, Bologna, Italy

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ABSTRACT

Aims: We provide morphological, immunohistochemical and molecular characterization of the 3rd "intermediate-grade" orbital meningeal melanocytoma, testing for the first time Vysis Melanoma FISH Probe Kit. We reviewed the literature in order to discuss the main differential diagnoses and to provide a better molecular description of these unusual tumors of difficult diagnosis and controversial management.

Methods: Histochemical stains (Haematoxylin and Eosin, Perls, reticulin), immunohistochemistry (HMB45, p16, Melan-A, S100, EMA, Ki67, CD68), polymerase chain reaction amplification and sequence analysis (BRAF, exon 15; NRAS exons 2 and 3; c-KIT, exons 11, 13, 17, 18; GNAQ, exons 4 and 5; GNA11, exons 4 and 5) and fluorescent in situ hybridization (RREB1, 6p25; MYB, 6q23; CCND1, 11q13; CEP 6, 6p11.1-q11.1) were performed on paraffin-embedded, formalin-fixed material.

Results: Histological diagnosis of "intermediate-grade" melanocytoma was supported by zonal necrosis and increased Ki67-index (12%). Immunophenotype: HMB45 + (strong, >75%), Melan-A + (strong, >75%), p16 + (~20%), S100 −/+ (<5%), EMA −/+ (<5%), CD68 − (positive histiocytes). No gene mutations nor copy-number alterations were identified. The patient was asymptomatic and disease-free 3 years after total surgical excision.

Conclusions: Adequate sampling and accurate immunohistochemical characterization are important for a correct diagnosis. Molecular analysis could provide important additional information (especially for "intermediate-grade" tumors), but further data are needed.

1. Introduction

Meningeal melanocytoma (mMC) is a circumscribed primary melanocytic tumor of the central nervous system (CNS), showing "well-differentiated" or "intermediate-grade" histology, slow growth rate and possible local recurrences; meningeal dissemination, malignant transformation and metastases were rarely reported [1–37].

Diagnosis may be challenging as to the existence of a spectrum of diffuse or circumscribed melanocytic lesions with benign, intermediate or malignant histology and clinical course (including diffuse melanocytosis, melanomatosis and malignant melanoma); moreover, non-melanocytic pigmented tumors (such as melanotic schwannoma and pigmented meningioma) should be considered in differential diagnosis [1,12,38–40].

* Corresponding author.
E-mail addresses: andreapalicelli@hotmail.it (A. Palicelli), mariagiuliadisanto@gmail.com (M.G. Disanto), renzo.boldorini@med.unipmn.it (R. Boldorini).

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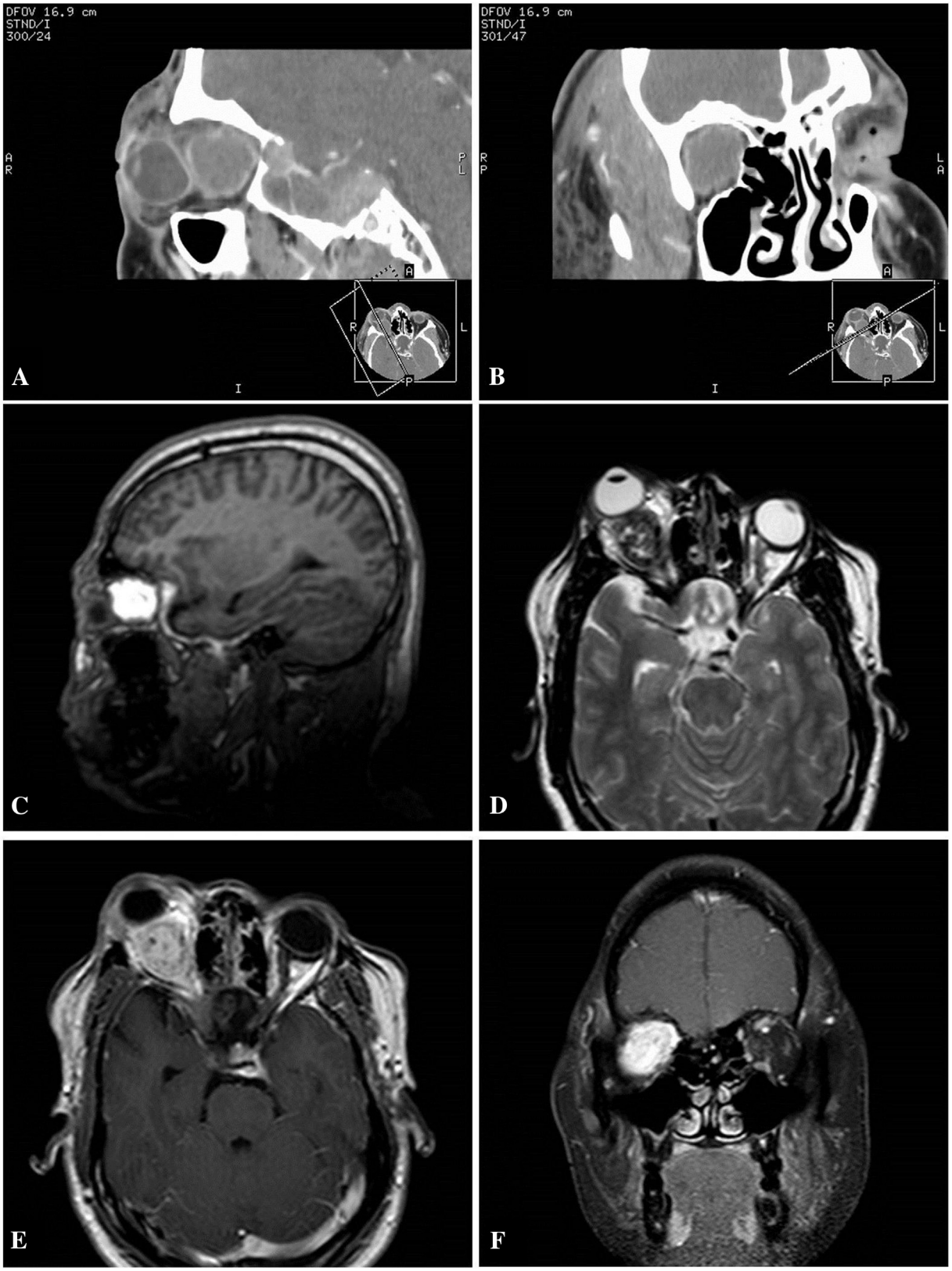

Fig. 1. A,B: Contrast-enhanced CT scans revealed an oval, retrobulbar, inhomogeneously hypodense lesion encasing the right orbital optic nerve and showing a peripheral contrast-enhanced ring (A: oblique sagittal scan; B: oblique coronal scan). C–F: On MRI scans the right intraconal pseudocapsulated lesion appeared hyperintense on T1-weighted scans (C: sagittal scan) and inhomogeneously hypointense on T2-weighted scans (D: axial scan). After administration of the contrast agent, the lesion showed a strong enhancement especially at the periphery (E: axial scan; F: coronal scan).