Sixth nerve and superior division of third nerve palsy due to intracranial extension of multiple myeloma. A "stroke mimic" diagnostic challenge.

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INTRODUCTION

Isolated acute ophthalmoparesis (iAO) may result from several intracranial vascular, infective, neoplastic or inflammatory diseases. Neuro-ophtalmologic signs have an important clinical value for localizing the pathologic process. Among rare cause of iAO, intracranial extension of multiple myeloma (MM) should be taken into consideration for its severity and the possibility of effective treatment.

CASE REPORT

A 78-year-old man was referred to our department for the sudden onset of horizontal diplopia, lasting two hours and spontaneously remitting and consistent right ptosis. No headache, rhinorrhea neither orbital pain occurred. His anamnesis showed arterial hypertension and 2-years-history of immunoglobulin-k-type MM, treated with bortezomib, melphalan, prednisone for 7 months and followed by 1-year-stability. Latter protein electrophoresis was unremarkable until 1 month before neurological symptoms. Daily low-dose of acetylsalicylic acid was administrated because of polyglobulia and relating hyperviscosity syndrome since 1 month. Neurological examination showed right ptosis and functional limitation of the superior rectus and ipsilateral abducens nerve palsy. No visual field deficit, symmetric pupil diameter, bilateral normal light-reagent pupil. The lesion of the superior subdivision of the 3th cranial nerve, including fibers for the superior rectus and levator palpebrae, associated with sixth nerve palsy indicated a localization within the anterior cavernous sinus or superior orbital fissure. Blood tests revealed monoclonal band in γ region with Ig-K-paraproteinaemia. CT showed soft tissue occupying the sphenoid sinus, with osteolysis of the clivus and the vault of the skull. Brain MRI confirmed a sphenoid sinus bone lesion, extending into the right cavernous sinus. The patient was referred to hematologist for therapy with bortezomib and dexamethasone, and radiotherapist, to combine radiotherapy, with complete disappearance of symptoms. After 4 months of therapy, brain MRI showed marked reduction of the mielomatous mass, with absence of enhancement and with central necrosis.

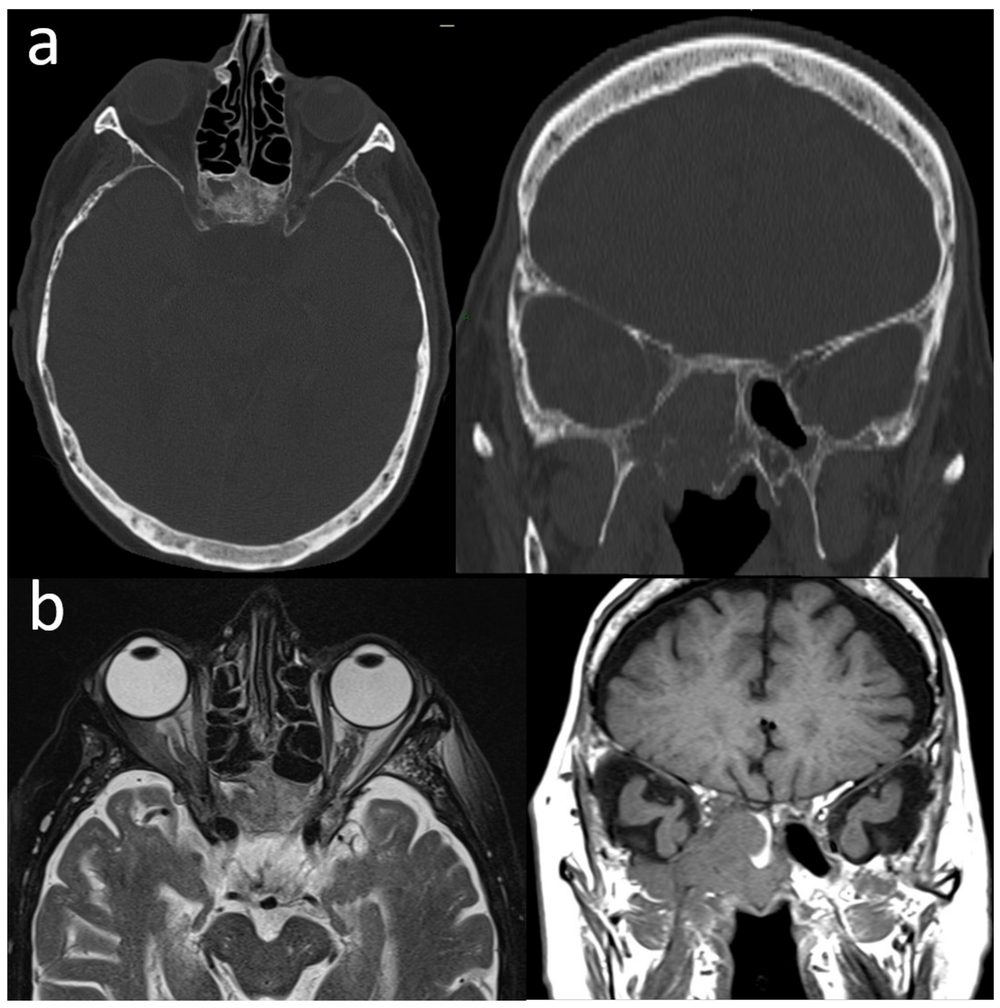

Fig1a CT scan: axial and coronal reformatted images. Fig. 1b, MRI T2-weighted axial and T1-weighted coronal images.

CONCLUSIONS

Multiple oculomotor nerve involvement, presence of absence of pain, proptosis, conjuntival chemosis and other peculiar neuro-ophalmological signs have an important clinical value. In our patient, considering his cerebrovascular risk factors (age, hypertension, hematological hyperviscosity), painless transient diplopia appeared more typical for a vascular attack than for an infiltrative lesion. Increasing knowledge of differential diagnosis about "stroke mimic" is fundamental. Although MM is a rare cause of cranial nerves palsies, an major awareness of clinical presentation of intracranial MM has be achieved for an early diagnosis and therapy.

BIBLIOGRAPHY
1. Cerase A, Tarantino A, Gozzetti A, Muccio C, Gennari P, Monti L, Di Blasi A, Venturi C, Intracranial involvement in plasmacytomas and multiple myeloma: a pictorial essay, Neuroradiology, (2008) 50:665–674.
2. Gozzetti et al, Multiple myeloma involving the cavernous sinus: a report of 3 cases and response to bortezomib, Clinical Lymphoma and Myeloma, 2007, (5), 376-378.
3. Gozzetti A., Cerase A., Novel agents in CNS Myeloma treatment, Central Nervous System Agents in Medicinal Chemistry, 2014, 14, 23-27.
4. Panda BB et al, Oculomotor nerve palsy as a rare presentation and first sign of multiple myeloma, J. Clin. Diagn. Res. 2016.
5. Gozzetti A, Cerase A, Tarantino A, Fabbri A, Bocchia M, Pirrotta MT, Lauria F, Multiple Myeloma involving the cavernous sinus: a report of 3 cases and response to bortezomib, Clinical Lymphoma and Myeloma, (2007) 7, 376-378.