

SOLITARY ORBITAL PLASMOCYTOMA

A CASE REPORT

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ABSTRACT Solitary orbital plasma cell neoplasm in the absence of multiple myeloma is very rare (Meyer et al. 2018). We present here a case report of extramedullary plasmacytoma of the left orbital soft tissue in a 50-year-old woman whose thorough diagnostic workup was negative for multiple myeloma. The patient presented with rapidly increasing proptosis, swelling, and visual disturbances of the left eye for past 15 days. MRI of the orbit revealed an extra conal well-circumscribed homogeneously enhancing masse filling the superior and the temporal part of the left orbit without bone destruction Or intracranial extension. The patient underwent a subtotal resection and the pathological report revealed a plasma cell neoplasm. So, normal skeletal survey, absence of bone marrow involvement, absence of Bence-Jones protein, no anemia, no hypercalcemia, no renal disease, normal level of immunoglobulin's all exclude the diagnosis of multiple myeloma. Since the visual acuity of the patient is well-preserved, so we have planned to treat her with local radiotherapy alone with 50 Gy.

Key words : Orbit, Plasmacytoma, Radiotherapy.

INTRODUCTION

Plasmocytoma can be manifest as systemic disease, namely, multiple myeloma and its variants (such as indolent myeloma, smoldering myeloma, osteosclerotic myeloma, plasma cell leukemia and non-secretory myeloma) or localized disease represented as solitary plasmacytoma of bone (SMB) or extramedullary plasmacytoma (EMP) (Uceda-Montañés et al. 2000). Involvement of the orbit can occur in either of the two forms. Solitary orbital plasma cell neoplasm in the absence of multiple myeloma is very rare with less than 50 cases reported in the literature (Matos et al. 2020) .

OBSERVATION

A 50 year-old female; with a biliary cirrhosis history was referred from an ophthalmologist with rapidly progressive left proptosis for past 15 days and moderate visual disturbances.

Physical examination showed with left-sided proptosis, swelling, limited ocular movement (an offense of abduction) with sever impairment of vision. The visual acuity was 8/10 of the left eye.

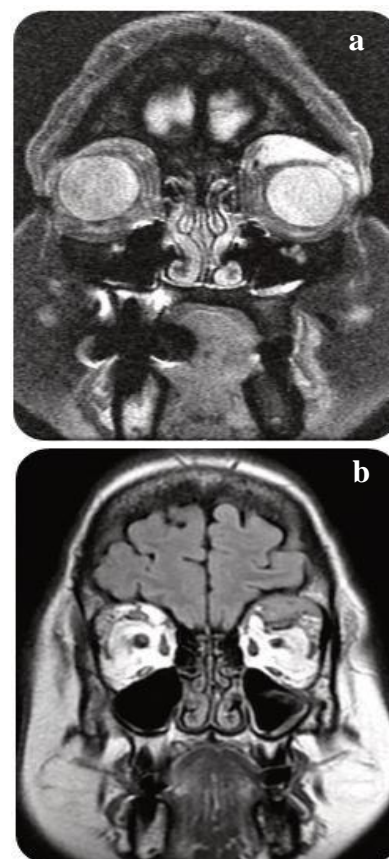


Figure 1: The patient orbital MRI showing, an extra conal well-circumscribed homogeneously enhancing masse filling the superior and the temporal part of the left orbit without bone destruction.

The patient underwent subtotal resection to protect and improve ocular function by preserving the surrounding anatomical structures through a sub frontal approach with removal of upper orbital rim, the result of the pathological report revealed a plasma cell neoplasm.

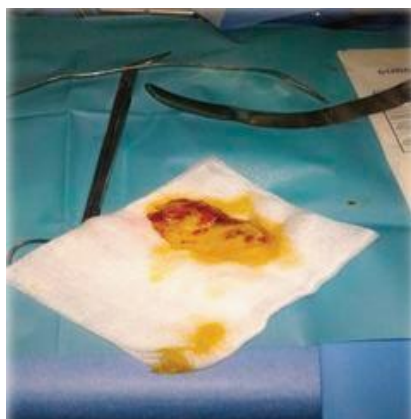


Figure 2 : mono-block fronto-orbital bone flap.



Figure 3a : Exposed orbital contents.

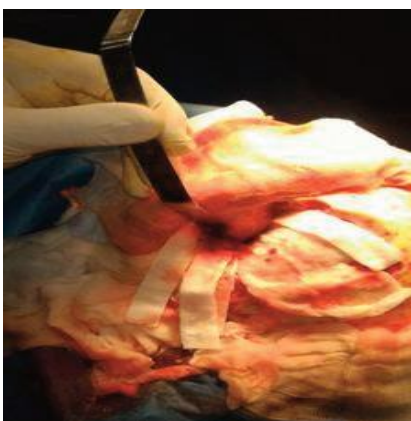


Figure 3b : Image of the lesion at the moment of being resected through a sub frontal supra orbital craniotomy.

Complete skeletal survey and bone marrow examination report were normal. So, normal skeletal survey, absence of bone marrow involvement, absence of Bence-Jones protein, no anemia, no hypercalcemia, no renal disease, normal level of immunoglobulin's all exclude the

diagnosis of multiple myeloma. The diagnosis of solitary orbital plasmacytoma is retained. And have planned to treat her with local radiotherapy alone with 50 Gy to stabilize the tumor volume after 04 months of evolution.

DISCUSSION

Orbital plasmacytoma can be primary or secondary. Side is multiple myeloma events (Sen, Kashyap, et Betharia 2003a); more frequent and tend to be more aggressive. The diagnosis of plasmacytoma requires a biopsy to distinguish several closely related pathologies, including plasma cell granuloma, plasmacytoid lymphoma and large cell lymphomas (immunoblastic type). The prognostic importance comes from a higher response rate to chemotherapy or radiotherapy, and a prolonged overall survival compared with those with the sole diagnosis of multiple myeloma. Radiotherapy as the sole treatment option, as given in this case is adequate in the absence of systemic involvement.

Plasmacytomas have been classified as low grade, intermediate or high grade using criteria originally intended for the grading of multiple myelomas. Chemotherapy was recommended after primary radiotherapy for the later two grades as these lesions had only 17% local control rate with radiation alone. However, EMPs with low-grade histologies were controlled in 83% cases external radiation only. Our case was a low-grade EMP of orbital soft tissue and well-preserved visual acuity treatment with radiation alone was justified.

CONCLUSION

An orbital plasma cell neoplasm with no systemic involvement at present may be the first of its kind to be reported to the best of our knowledge. However, the risk of progression to multiple myeloma is not yet documented, should always be kept in mind, mandating a close follow-up of this patient.

REFERENCES

- 01] ADKINS, JEFFERY W., JERRY A. SHIELDS, CAROL L. SHIELDS, RALPH C. EAGLE, JOSEPH C. FLANAGAN, ET PETER C. Campanella. 1997. « Plasmacytoma of the eye and orbit ». *International ophthalmology* 20(6):339–343.

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- 02] DANG, SABINA, NAUMAN F. MANZOOR, HANNAH HARMSSEN, ALEJANDRO RIVAS, ET JOSEPH M. AULINO. 2019. « Skull Base and Orbital Solitary Plasmacytoma Mimicking a Meningioma ». *JAMA Otolaryngology-- Head & Neck Surgery*.
- 03] EZRA, ERIC, GEVA MANNOR, JOHN E. WRIGHT, ET GEOFFREY E. ROSE. 1995. «Inadequately Irradiated Solitary Extra medullar Plasmacytoma of the Orbit Requiring Exenteration ». *American journal of ophthalmology* 120 (6) : 803–805.
- 04] JACKSON, A., J. KWARTZ, J. L. NOBLE, ET M. J. REAGAN. 1993. « Multiple myeloma presenting as bilateral orbital masses : CT and MR appearances ». *The British Journal of Radiology* 66 (783) : 266–268.
- 05] MATOS, ANDREIA, ANDRÉ GOULART, ANDRÉ RIBEIRO, RICARDO FREITAS, CATARINA MONTEIRO, ET PAULO MARTINS. 2020. «Orbital Plasmacytoma, An Uncommon Presentation of Advanced Multiple Myeloma ». *European Journal of Case Reports in Internal Medicine* 7 (3) : 001149.
- 06] MEYER, HANS JONAS, SEBASTIAN ULLRICH, GORDIAN HAMERLA, ET ALEXEY SUROV. 2018. « [Extramedullary Plasmacytoma] ». *RoFo: Fortschritte Auf Dem Gebiete Der Rontgenstrahlen Und Der Nuklearmedizin* 190(11):1006-9.
- 07] NA'ARA, SHOROOK, MORAN AMIT, ZIV GIL, ET SALEM BILLAN. 2016. « Plasmacytoma of the Skull Base: A Meta-Analysis ». *Journal of Neurological Surgery. Part B, Skull Base* 77(1):61-65.
- 08] PANDA, BIJNYA BIRAJITA, SUCHETA PARIJA, JYOTIRANJAN MALLICK, ET SUSANTA PUJAHARI. 2016. « Oculomotor Nerve Palsy as a Rare Presentation and First Sign of Multiple Myeloma ». *Journal of Clinical and Diagnostic Research: JCDR* 10(5):ND01-03.
- 09] SEN, SEEMA, SEEMA KASHYAP, ET SUBHASH M. BETHARIA. 2003a. «Primary orbital plasmacytoma: a case report». *Orbit* 22(4):317–319.
- 10] SEN, SEEMA, SEEMA KASHYAP, ET SUBHASH M. BETHARIA. 2003b. « Primary orbital plasmacytoma: A case report ». *Orbit* 22(4):317-19.
- 11] UCEDA-MONTAÑÉS, A., G. BLANCO, M. A. SAORNIL, C. GONZALEZ, J. L. SARASA, ET J. CUEVAS. 2000. « Extramedullary plasmacytoma of the orbit ». *Acta Ophthalmologica Scandinavica* 78 (5) : 601-603.
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