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Ophthalmology | Case report

Orbital Extramedullary Plasmacytoma: Case report and literature review

Lilian Brito Gomes[1*], Jonathan Dos Santos Lima[2], Valéria Camargo Silveira[3], Jacqueline Stephanie Fernandes Do Nascimento[4], Nicolle Dos Santos Moraes Nunes[5], Marco Orsini[6], Gilberto Canedo Martins Jr[7], Jorgete Haiddar Sessim David Cosendey[8], Roberta Araújo De Arruda Camara[9], Carlos Henrique Melo Reis[10], Antonio Marcos Da Silva Catharino[11]

[1]Physician of the Internal Medicine Service of Nova Iguaçu General Hospital
[2]Medical Student of Universidade Iguaçu. Universidade Iguaçu- UNIG-RJ, Nova Iguaçu - RJ, Brasil.
[3]Professor of Neurology - Universidade Iguaçu- UNIG-RJ, Nova Iguaçu - RJ; Physician of the Neurology Service of Nova Iguaçu General Hospital
[4]Medical Student of Universidade Iguaçu. Universidade Iguaçu- UNIG-RJ, Nova Iguaçu - RJ, Brasil. Orcid: https://orcid.org/0000-0003-1847-3829.
[5]Medical Student of Universidade Iguaçu. Universidade Iguaçu- UNIG-RJ, Nova Iguaçu - RJ, Brasil. Orcid: https://orcid.org/0000-0003-0336-2261.
[6]Professor of Neurology, Universidade Iguaçu- UNIG-RJ, Nova Iguaçu - RJ, Brasil. Orcid: https://orcid.org/0000-0002-8526-6937.
[7]Professor of Neurology - Universidade Iguaçu- UNIG-RJ, Nova Iguaçu - RJ; Physician of the Neurology Service of Nova Iguaçu General Hospital
[8]Physician of the Neurology Service of Nova Iguaçu General Hospital
[9]Physician of the Radiology Service of Nova Iguaçu General Hospital; Professor of Universidade Iguaçu- UNIG-RJ, Nova Iguaçu - RJ
[10]Universidade Iguaçu- UNIG-RJ, Nova Iguaçu - RJ; Physician of the Neurology Service of Nova Iguaçu General Hospital
[11]Professor of Neurology - Universidade Iguaçu- UNIG-RJ, Nova Iguaçu - RJ; Physician of the Neurology Service of Nova Iguaçu General Hospital. Orcid: https://orcid.org/0000-0002-5736-1486

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Address for correspondence: Lilian Brito Gomes - Rua Maria José Pacheco, 101 - Bairro: Dom Rodrigo, Nova Iguaçu - Rj, Cepl: 26.260-370 – Phone (21) 99926-6136 - email: lihbrito@yahoo.com.br

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Abstract

Extramedullary plasmacytoma is a rare disease, characterized histopathologically by infiltration of plasma cells of different maturities and producing monoclonal immunoglobulin outside the bone marrow. J.R.G., 52 years old, male, sought medical assistance due to amaurosis and blocked eye movements. We performed Magnetic Nuclear Resonance of the skull that showed discrete intermediate signal tissue, next to the optic nerve canal and homolateral superior orbital fissure, with slight extension to the lower orbital fissure, associated with thickening of the corresponding meninges, involving the proximal portion of the optic nerve, measuring 1.3 x 1.1 x 2.5 cm. Non-specific aspect, but suggestive of lymphoproliferative lesion. Right and extraorbital tumor excision was performed, in which it was not possible to completely resect the tumor. The anatomopathological examination of the piece showed a neoplasm of plasma cells (plasmacytoma). The diagnosis is made from the exclusion of Multiple Myeloma. The treatments of choice are radiotherapy, due to the high radiosensitivity in 80–100% of cases, and surgery for localized lesions.

Keywords: Extramedullary plasmacytoma; Orbital tumor.

Introduction

Plasma cells are cells differentiated from B lymphocytes and specialized in the production of antibodies. They develop in Organs lymphoid organs and in the sites of the immune response. The plasmacyte differentiation process involves morphological changes and, mainly, changes in gene expression, in order to promote the synthesis of antibodies [1].

Plasmacytomas are lymphoproliferative disorders in soft tissues where the plasmocyte clone occurs in different stages of differentiation, increasing the production of immunoglobulins. This neoplasm is divided into three types: extramedullary plasmacytoma (PEM), solitary bone plasmacytoma (PCOS) and Multiple Myeloma (MM), which can be primary, or secondary to the evolution of solitary bone plasmacytoma [2,3]. Plasma cell disorders are characterized by the presence of monoclonal proteins (antibodies or altered antibody fragments) in serum or urine and/or by the presence of plasma cells in the bone marrow or, rarely, in other tissues [4].

In the period from 2008 to 2017, a total of 29,168 patients were treated at SUS with a diagnosis of disorders of plasma origin. Of these, 27,100 were diagnosed with Multiple Myeloma (93% of the total), 1,030 with Extramedullary Plasmacytoma (3.5%), 541 with Monoclonal Gammopathy of Undetermined Meaning (1.8%) and 497 with Plasmacytic Leukemia (1.7%). The state with the highest number of patients seen was São Paulo (7,460 patients), followed by Minas Gerais (3,648 patients) and Rio Grande do Sul (2,345 patients). Multiple myeloma and related disorders were more prevalent in male patients (52%), while the most common race/color in all disorders was white (44.5%), followed by no information (26%) and by race / brown color (23%). The median age of patients seen was 63 years for patients with Multiple Myeloma and Monoclonal Gammopathy of Undetermined Meaning, 62 years for patients with Plasmocytic Leukemia and 60 years for patients with Extramedullary Plasmacytoma [1].

Extramedullary plasmacytoma is a rare disease, characterized histopathologically by infiltration of plasma cells of different maturities and producing monoclonal immunoglobulin outside the bone marrow. It may be the initial manifestation of multiple myeloma, with an average progression of 30% of the situations [3].

After treatment, three situations may occur: local recurrence of the disease, recurrence in regional lymph nodes and/or progression to multiple myeloma. The most common is the progression to myeloma after an average of 2 to 2.5 years. The probability of local control after treatment is 80% to 90%, being the same after surgery or radiation therapy [6].

Clinical case

JRG, 52 years old, male, with systemic arterial hypertension and type 2 diabetes mellitus, sought medical assistance due to amaurosis and blocked eye movements, manifesting at first by visual turbidity on the right and progression to bilateral amaurosis in up to 2 weeks. We performed Magnetic Resonance of the skull that showed abnormal tissue of intermediate signal, next to the right optic nerve canal and homolateral superior orbital fissure, with slight extension to inferior orbital fissure, associated with thickening of the corresponding meninges, involving the proximal portion of the optic nerve, measuring 1.3 x 1.1 x 2.5 cm (Figure 1). Non-specific aspect, but suggestive of lymphoproliferative lesion. Right and extraorbital tumor excision was performed, in which it was not possible to completely resect the tumor. The anatomopathological examination showed a neoplasm of plasma cells (Plasmacytoma). The result of protein electrophoresis was normal and the Bence Jones test was negative. The patient was referred to the National Institute of Oncology for further treatment.

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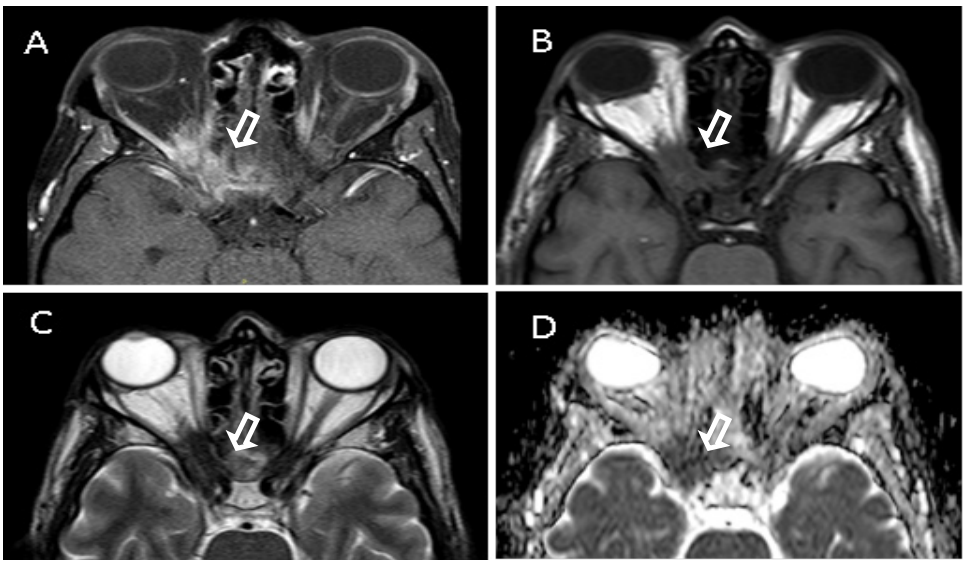

Figure 1: Magnetic Resonance showing abnormal tissue of intermediate signal, next to right the optic nerve canal and homolateral superior orbital fissure, with slight extension to inferior orbital fissure (arrows) - A) T1 WITH GADOLINIUM; B) T1; C) T2; D) ADC.

Discussion

Plasmacytomas are mainly divided into single medullary / bone (solitary myeloma), extramedullary or one of the components of multiple myeloma. They are made up of plasma cells showing malignant degeneration, producing a specific immunoglobulin molecule. The incidence of PEM is higher in males than in females, with a ratio of 3–4: 1, more frequent around 50–60 years of age. It is estimated that they correspond to 2–4% of plasma cell-derived neoplasms, in which the greatest representative is multiple myeloma, the latter constituting up to 1% of the total of general malignancies [5].

Approximately 80–90% of PEM cases occur in craniocervical structures (upper aerodigestive tract, larynx, nasopharynx, tonsils, nasal and paranasal cavities), however, their numbers do not reach 1% of the head and neck neoplastic lesions. Other sites such as the gastrointestinal and urogenital tracts, central nervous system, thyroid, parathyroid glands, salivary glands, lymph nodes, skin, lungs and breasts are uncommon [2].

Histologically, they do not originate directly from the bone marrow and are indistinguishable from multiple myeloma, as well as the differentiation of plasma cell granulomas and other inflammatory reactions, making immunophenotyping essential. The diagnosis of PEM occurs after a rigorous investigation to exclude multiple myeloma, highlighting its histological confirmation using immunohistochemistry, bone marrow biopsy / puncture showing less than 5% of plasma atypia, discarding the presence of osteolytic lesions, dosage and electrophoresis of serum and urinary proteins (exclude the presence of M and Bence-Jones proteins, respectively) and no anemia. Nuclear Magnetic Resonance is the exam of choice to show lytic lesions [4].

The treatments of choice are radiotherapy, due to the high radiosensitivity in 80–100% of cases, and surgery for localized lesions. In these alternatives, recurrence and dissemination rates of approximately 20–40% are observed, with a 70% survival in up to 10 years [2,3]. Adjuvant chemotherapy can be used to prevent progression to MM; however, its role is still controversial. Several studies suggest that Chemotherapy increases the clearance of M proteins and reduces the progression to MM, while others have found no benefit [7].

As PEM can be the first evidence of a plasma cell neoplasia and its progression to MM occurs in up to 30% of cases, after the initial diagnosis, a thorough investigation must be carried out in this regard [6].

Conclusion

Orbital extramedullary plasmacytoma is a rare disease and should be considered in the evaluation of an orbital tumor, with special emphasis on symptoms such as proptosis, exophthalmos, diplopia; that allow an early diagnosis and a better interventionist approach within a multidisciplinary team, considering that an appropriate treatment can lead to the cure of the patient or, at least, to a significant tumor response, with delay in the conversion to multiple myeloma [8,9].

Conflict of interest

The authors declare that there is no conflict of interest.

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