ORBITAL MYELOID SARCOMA - A CASE REPORT

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INTRODUÇÃO

Myeloid sarcoma (MS), or granulocytic sarcoma, is a tumoral lesion characterized by immature granulocytic series cells, also known as chloroma due to its greenish colour[1,2]. MS are rare extramedullary manifestation of Acute Medullar Leukemia (AML) with a 2,5 - 8% incidence, presenting its most common location the orbital site in pediatric population[3,4]. In case of orbital location, bilateral proptosis is slightly more frequent than unilateral proptosis[5].

Orbital MS report cases are mostly associated with a high misdiagnosis rate, as isolated orbital MS exhibited clinical features that mimicked other conditions[6-11]. Most childhood orbital tumors are benign unilateral diseases, but about 5 - 10% of the malignant ones, rhabdomyosarcoma is the most common, characteristically unilateral[12-15].

This case report shows a bilateral simultaneous orbital MS in a 4-year old patient as a first sign of AML. Although MS is a rare finding among orbital tumors, when a bilateral tumor it becomes a major diagnostic consideration, being crucial to early diagnosis and treatment of AML.

RELATO DO CASO

A caucasian previously healthy 4-year old male patient developed a right earache along unspecific malaise without improvement with antibiotic therapy. After 7 days, a right facial palsy and bilateral painless proptosis appear, being admitted to Mario Covas Hospital for further investigation and treatment. Laboratory examination demonstrated bicytopenia with peripheric blasts, and MRI showed expansive formations on both orbits, infratemporal fossae, pterygopalatine fossae, and anteriorly to S2 and S3. Myelogram is collected presenting over 50% of blasts, CD117, CD34 and CD19 positive, confirming the diagnosis of AML with bilateral orbital, cranial and medullar MS.

The ophthalmic evaluation performed on the first day of admittance demonstrated important bilateral proptosis, worst on the left, with venous palpebrae engorgement, incomplete ocular closing, virtually absent extrinsic ocular mobility and conjunctival hyperemia. The patient denied visual impairment or diplopia, presented good stereopsis, the pupil was round, reactive and regular, with no relative afferent defect on both sides. (Figure 1).

The patient evolved to a worsening of the left proptosis and dense chemosis. Fundus exam realized under sedation was normal. Visual acuity was maintained, as well the absence of pain. The left eye inferior chemosis start to worsen to fold over itself and over the lower eyelid, along increase of superior conjunctival hyperemia and a new linear inferior perilimbal corneal lesion, related to the step between the cornea and the conjunctival lesion, as well the incomplete eye closing. (Figure 2). A biopsy was suggested, but as the patient evolved to important thrombocytopenia and leukopenia, the conservative treatment was maintained due to infection and hemorrhage risks.

After the introduction of chemotherapy, the clinical and ophthalmological conditions started to improve drastically, with the proptosis and chemosis rapidly regressing over the first 48 hours of intravenous treatment. The bloodwork presented a substantially improvement of blasts and the ocular mobility began to return to normality. (Figure 3).

A new myelogram is collected after 15 days of chemotherapy with minimal residue disease of 0,2%, discharging the patient to continue the therapy at home. After numerous readmissions for indutory and intensification protocols, a haploidentical hematopoietic stem cells transplant (HHSCT) is successfully performed after 5 months of the first signs of disease, showing remission after 5 months of HHSCT. (Figure 4).

FIGURAS, TABELAS E GRÁFICOS

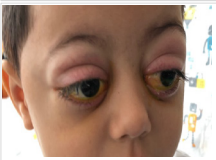

Figure 1: bilateral proptosis.

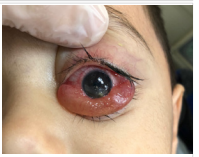

Figure 2: inferior chemosis

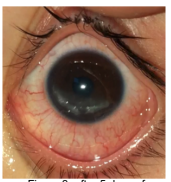

Figure 3: after 5 days of chemotherapy

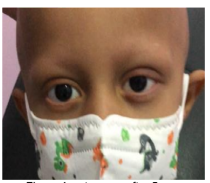

Figure 4: ectoscopy after 5 months of HHSCT

DISCUSSÃO:

MS originates in bone marrow and can spread through the Haversian canals, collecting in soft tissues and sub-periosteum sites forming a tumoral mass[18]. The most frequent manifestation of direct orbital infiltration is proptosis, with other possible signs and symptoms being chemosis, lid edema, intra-retinal or choroid hemorrhages, diplopia, loss of sight, palsies of extraocular muscles and papilledema[12]. The proptosis is related to direct infiltrate of orbital soft tissues, venous blockage, retrobulbar hemorrhages and/or muscle infiltratation[16].

Some orbital MS report cases have been described, most being initially associated with a high misdiagnosis rate, as isolated orbital MS exhibited clinical features that mimicked inflammatory or lymphoproliferative diseases[6-11]. Most childhood orbital tumors are unilateral, as the most frequent benign conditions like dermoid cysts, capillary hemangioma, lymphangioma and optic nerve glioma normally affect one single orbit[12,13]. About 90 - 95% of orbital masses of pediatric population that come to biopsy has shown benign histopathologic findings, and the 5 - 10% that are malignant, rhabdomyosarcoma is the most common disease, invariably unilateral[12,13].The main causes of bilateral orbital masses in children are idiopathic non-granulomatous orbital inflammation, metastatic neuroblastoma, and MS[14,15].

Therefore, when facing an orbital mass in children, the ophthalmologist must evaluate a number of conditions and variations to accomplish a proper diagnosis. Although MS is an uncommon orbital tumor, in setting of bilateral disease it becomes a major, perhaps the first diagnostic consideration, even without any signs of leukemia, as it may be the first sign of this neoplasia. Our reported patient is a 4-year old male child that presented an acute bilateral proptosis associated with a number of unspecific initial signs and symptoms that promptly regressed after the introduction of chemotherapy, leaving no severe consequences. This example shows the importance of undergo evaluation of AML in any child with orbital mass, particularly if bilateral.

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