Operative case of Langerhans’ cell histiocytosis of the skull with dual invasion. An immunohistochemical study of ki-67 expression of eosinophilic granuloma: case report and review of the literature

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Eosinophilic granulomas (EG), Letterer-Siwe disease and Hand-Schüller-Christian disease are collectively called Langerhans-cell histiocytosis (LCH). While the latter two are systemic diseases, the former is a localized form of histiocytosis. Solitary EG of the skull are rare lesions characterized by a natural history not well defined yet. In this context, we report a case of a 23-year-old male suffering from a recurrent and progressive right parietal headache. On computed tomography (CT), it was observed an ovoidic lesion which on magnetic resonance imaging (MRI) appeared as an hypointense soft mass on both T1 and T2 weighted images. The lesion showed a marked and heterogeneous enhancement after gadolinium administration. The surgical excision was complete and the severe headache disappeared. Immunohistochemical analysis of the specimen indicated an eosinophilic granuloma characterized by Ki-67 nuclear antigen expression with a labeling index of 20%. In the pertinent literature we have found two aggressive cases of EG showing the Ki-67 expression with a respectively 6.2% (cortical bone granuloma) and 10% (parietal bone granuloma) labeling index. That high proliferative activity suggests a local Langerhans’ cell proliferation along with an exuberant inflammatory response and also explains the aggressive clinical course and the rapid expansion of the lesion observed in some rare cases of solitary EG. This is the third case-report of calvarial EG characterized by Ki-67 nuclear antigen expression.