SARCOMATOID ANAPLASTIC LYMPHOMA ALK NEGATIVE PRIMARY CENTRAL NERVOUS SYSTEM: CASE REPORT

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BACKGROUND: Primary lymphoma of the central nervous system (CNS) is an uncommon variety of non-Hodgkin lymphoma (NHL) who constitute about 4% of CNS tumors and 1-2% of all non-Hodgkin's malignant lymphomas. Among them, a rare variety is anaplastic lymphoma, representing 5% of all NHL and which can be divided as anaplastic lymphoma ALK positive(+) and ALK negative(-).

The case of a patient with sarcomatoid anaplastic lymphoma ALK- is presented, being a rare pathology.

METHODS: The clinical file of patient with anaplastic ALK- lymphoma SNC was analyzed including its respective lab and image studies.

RESULTS: Female, 47 years old, presented ocular retro pain and right pulsatile frontotemporal headache, a magnetic resonance imaging (MRI) of the head, showed extra axial tumor dependent middle third of the lesser wing of the right sphenoid, with axial compression of the temporal fossa and orbit radius. Right orbito-fronto-temporal craniotomy with durotomy was performed, as well as resection of the tumor; histopathological report of anaplastic large cell lymphoma sarcomatoid variety, CD30+, CD45+, CD 20-, ALK-. MRI with postoperative persistence dependent lesion of the greater wing of the right sphenoid with extension to intraconal portion of the right orbit and involucre top and lateral rectus muscles. Treatment was initiated with radiotherapy, methotrexate and based chemotherapy vorinostat and cytarabine.

CONCLUSION: ALK- anaplastic lymphoma is a rare disease that occurs in people around 65 years and has a worse prognosis (survival 12.5% at 5years) than ALK+. In the case of our patient's poor response to treatment was observed.