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Case report and brief review of the literature on high-grade sarcoma in the skullcap

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ABSTRACT

High-grade sarcoma in the skullcap is a rare neoplasm, affecting adults between 30 and 60 years of age, with no gender predilection. Genetic factors and exposure to ionizing radiation may be associated with its development. It has a malignant histological profile, with poorly differentiated cells, rapid growth, and aggressive local invasion. This article aims to describe in detail the case of a 22-year-old patient, with no expected comorbidities, who suffered trauma to the frontal region of the head. After the incident, he developed persistent and daily headache, leading to imaging tests. Computed tomography and magnetic resonance imaging of the skull were performed, showing sarcoma, which was promptly addressed. In January 2023, the patient returned with tumor recurrence, evidencing the need for a second surgical approach. The patient was referred for chemotherapy and radiation therapy afterwards as part of the treatment plan.

Keywords: Sarcoma, Neoplasm, Skull.

1 INTRODUCTION

High-grade sarcoma in the skullcap usually involves adults between 30 and 60 years of age, and there is no significant gender dominance. Specific risk factors have not yet been clearly identified, but genetic factors and exposure to ionizing radiation may be related to the development of this type of neoplasm[1].

The histological profile of high-grade sarcomas in the skullcap usually has features of malignancy. These high-grade sarcomas can be composed of undifferentiated or poorly differentiated cells, with rapid growth patterns and a high rate of mitosis, marked energetic atypical cells, and aggressive local invasion[2].

This high-grade sarcoma in the skullcap is a rare type of cancer and its clinical presentations can vary, but often include localized pain and lump in the head. The diagnosis involves imaging tests, biopsy and clinical evaluation. And as for treatment, they may include surgery, radiotherapy and in some cases chemotherapy2,3.

The present research project aims to report an unusual clinical case, given the rarity and complexity of high-grade cranial sarcoma. This unique malignancy presents a major challenge for healthcare professionals due to its rare condition and the intricate nature of its clinical features. Its importance is justified by the scarcity of articles that contemplate this theme. Thus, a detailed case report followed by a brief review of the literature will contribute to disseminating knowledge about this condition, facilitating early diagnosis and ensuring more appropriate and effective treatments.

2 METHODS

A search was carried out in the patient's medical records, extracting information relevant to the article, such as imaging tests, medical opinions, clinical evolutions and anamnesis. In parallel, a review of recent literature was carried out in databases such as PubMed and Lilacs. The project was approved by the Hospital's ethics committee and followed the guidelines of the National Council for Ethics in Research (CONEP).

3 CASE REPORT

A 22-year-old patient, admitted in June 2022 from Primavera-SP, without comorbidities or continuous use of medication, reports that he suffered mechanical trauma with a soccer ball in the frontal region of the head, without loss of consciousness, but since then he has had headache and dizziness daily, requiring intravenous analgesics.

Due to the patient's chronic complaint, a computed tomography scan of the skull was requested, which revealed expansive lesions in the bilateral frontal parietal region with advanced bone erosion. The investigation was complemented with magnetic resonance imaging of the skull with spectroscopy and venous angiotomography for the study of vessels. The patient reported progressive headache during hospitalization, refractory to the use of analgesic medications.

The case was discussed with the neurosurgery team, which opted for a surgical approach, where it performed resection of the complete lesion in the affected skull region, with reconstruction of the gap by means of methacrylate. As a result of anatomopathological findings, a fusocellular mesenchymal lesion was found, with an immunohistochemical pattern suggesting high-grade sarcoma, requiring revision of the slide.

After the surgical intervention, the patient remained in an outpatient segment, with regular follow-ups and imaging tests to control the condition. There was improvement in headache complaints after recovery from surgery, but not complete remission.

In January 2023, the patient returns to the emergency room with swelling and bulging of the flap of the frontal prosthesis, and a new imaging exam was performed that showed tumor growth, with indication for a new surgical approach, and a large part of the liquefied and amorphous lesion was removed, with extension to the underlying meninges in the frontal region, with a new histopathological analysis confirming a high-grade sarcoma lesion with an infiltrative aspect. Patient referred for chemotherapy and subsequent radiotherapy.

4 DISCUSSION

High-grade sarcomas represent a category of bone and soft tissue tumors that defy understanding and treatment in the medical community. This article seeks to shed light on the most complex and challenging aspects of these neoplasms, based on a detailed examination of scientific articles that explore their epidemiology, clinical characteristics, and therapeutic strategies2.

The first observation analyzed is the rarity of these tumors, with an incidence that represents less than 1% of all cancers diagnosed. This intrinsic rarity imposes substantial difficulties in conducting scientific studies and obtaining high-quality evidence. However, despite these limitations, the scientific community has made significant progress in understanding the molecular biology of these neoplasms and in developing more effective therapeutic approaches4.

A crucial point discussed in scientific studies is the importance of a multidisciplinary approach in the management of high-grade sarcomas. Collaboration between surgeons, oncologists, radiologists, and pathologists is essential to ensure an accurate diagnosis and proper

therapeutic planning. This approach allows for a comprehensive evaluation of the patient and a careful selection of the most appropriate therapeutic strategies according to each case and its individuality2,4.

About treatment, complete surgical resection of the tumor, whenever possible followed by adjuvant radiotherapy, remains the cornerstone of the management of these neoplasms. However, advances in the understanding of molecular biology have highlighted the heterogeneity of these tumors and the need for personalized therapeutic approaches. Specific biomarkers, such as distinct genetic rearrangements, are essential tools for diagnosis and prognosis, ensuring the selection of the most effective targeted therapies1,3.

It is necessary to emphasize that, despite advances in systemic therapy, such as chemotherapy, many subtypes of high-grade sarcomas demonstrate resistance to these treatments. This underscores the importance of continuous research and the development of new therapeutic strategies, with a focus on targeted therapies and immunotherapy, which have shown promising areas in the treatment of these challenging neoplasms5.

Regarding prognosis, high-grade sarcomas have significant variability, depending on the tumor subtype, location, and stage of the disease. While some subtypes exhibit an aggressive clinical course and unfavorable survival rates, others have a more favorable prognosis, especially when diagnosed early and treated appropriately5,6.

All in all, high-grade sarcomas continue to pose a formidable clinical challenge, but advances in understanding molecular biology and developing new therapeutic strategies are offering hope to patients affected by these devastating conditions. In the context of intracranial neoplasms, high-grade sarcomas affecting the skullcap emerge as clinical and pathological entities that are difficult to manage, characterized by a unique biological complexity and a varied spectrum of presentations.

Analysis of scientific studies has revealed a wealth of information about high-grade sarcomas in the skullcap, highlighting their low incidence, histological diversity, and unique therapeutic challenges. The association between Paget's disease and the occurrence of skull sarcomas highlights the complexity of pathological interactions in this anatomical region, however, there are still doubts about the relationship between trauma and the pathogenesis of these neoplasms, which suggests new directions for future research6

The multidisciplinary approach remains fundamental in the management of high-grade sarcomas in the skullcap, with an emphasis on complete surgical resection of the tumor followed by adjuvant radiotherapy. Advances in understanding the molecular biology of these tumors are

opening up new perspectives for the development of targeted and personalized therapies, offering hope for patients affected by these devastating conditions1,3,6.

However, despite significant advances, many challenges remain. The rarity of these neoplasms and the lack of studies limit our understanding and treatment capacity. Therefore, it is critical to continue investing in translational research and multidisciplinary collaborations to advance knowledge and improve clinical outcomes for patients with high-grade sarcomas in the skull vault5.

Ultimately, this article provides a comprehensive and detailed overview of these pathological entities, highlighting the importance of ongoing research and a holistic approach in the management of these rare tumors. By promoting a deeper understanding of high-grade sarcomas in the skullcap, we hope to catalyze efforts to improve clinical outcomes and quality of life for patients affected by these challenging conditions.

CONFLICTS OF INTEREST

The authors declare that there is no potential conflict of interest that could interfere with the impartiality of this scientific work.

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