Giant Cell Tumor in a Pediatric Patient: Rare Case Report

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Relato del caso: Paciente pediátrico con diagnóstico de sarcoma de Ewing (SE) y posteriormente de TCG, cuando procuró atención médica por aumento de volumen en arco cigomático y región preauricular derecha. Los exámenes de imagen mostraron una lesión expansiva en la porción cigomática y escamosa del cráneo, con componente intracraneal extradural en la fosa media, sin evidencias de infiltración. Desde el punto de vista anatomo-patológico, se observó neoplasia fusocelular. El marcador CD68 fue positivo y el marcador S100, negativo. Se realizó microcirugía para resecar el tumor. El paciente permanece en seguimiento con exámenes de imagen en los servicios de neurocirugía y ortopedia, con las últimas resonancias magnéticas de cráneo mostrando una reducción del tamaño de la lesión. Además, no se somete a terapia adyuvante y actualmente no tiene quejas.

Conclusión: Este es un caso importante por su rareza y datos clínicos que agregan informaciones sobre las patologías presentadas, permitiendo, en el futuro, nuevas investigaciones para optimizar el tratamiento de la neoplasia referida, y el pronóstico de estos pacientes.

Palabras clave: tumor óseo de células gigantes; fosa craneal media; neoplasias craneales; informes de casos.
INTRODUCTION

A giant cell tumor (GCT) is usually a benign tumor, which can be locally aggressive. They account for 5% of bone tumors, only 2 to 5% of these cases occur in the pediatric group whose age-range is 13-19 years. The treatment of choice is surgical1-3. Monitoring the patient after the end of treatment is essential, as GCTs have high recurrence rates4.

Case report about a GCT in a child localized in the zygomatic arch and in the squamous portion of the right temporal bone.

The Institutional Review Board of “Centro de Projetos de Ensino e Pesquisa (CEPEP)” of “Hospital Erasto Gaertner”, Curitiba, Brazil approved the study, CAAE number 87548518.2.0000.0098. The study patient signed the Informed Consent Form (ICF).

CASE REPORT

Male patient, 12 years old, previously diagnosed and treated for Ewing’s sarcoma in the left hip, returns to the pediatric oncology service of Hospital Erasto Gaertner, Curitiba, PR, complaining of swelling in the zygomatic and right preauricular region, detected by the family after one month of a local trauma. The lesion was difficult to delimit upon unpainful palpation.

A skull computed tomography (CT) showed a solid destructive lesion, compromising the right zygomatic arch, extending to the ipsilateral temporal bone, where it was determined intracranial compartment invasion. Anatomopathological analysis of a biopsy specimen showed gigantocellular proliferation associated with reactive osteoid neoformation.

Magnetic resonance imaging (MRI) was performed for surgical planning (Figure 1), showing an expansive bone lesion affecting the zygomatic arch and squamous portion of the right temporal bone, measuring about 4 cm in its longest axis, with a small extradural intracranial component in the middle cranial fossa, with no evidence of infiltration or dural thickening. Therefore, the patient underwent microsurgery for total tumor resection.

The anatomopathological diagnosis of the excised lesion showed fusocellular neoplasia with giant cells with foci of bone neoformation. Immunohistochemistry showed positivity for CD68 (Figure 2) and negativity for S100, closing the diagnosis of GCT. One month after surgery, the control skull CT scan showed two lesions, one on the inner face of the middle third of the zygomatic arch, measuring 9x8x6 mm, and another lesion in the transition between the posterior portion of the zygomatic arch and squamous portion of the right temporal bone, measuring 13x7x5 mm, with suspicion of recurrent or residual injury.

The patient returned to a new MRI in 3 months, which showed the same irregular expansive lesion in the proximal portion of the right zygomatic arch, measuring about 30x13x14 mm, which may represent an area of confluence of the lesions identified in the previous examination. Another skull MRI (Figure 3) 9 months after surgery showed a slight reduction of the lesion, now measuring 19x5x19 mm. A possible relapse of the injury was again suggested.

The patient continues to be monitored. Until the beginning of the year 2020, he had no complaints, without recurrence signs of the ES and monitoring of the dimensions of the GCT of the zygomatic arch.
DISCUSSION

GCTs are considered primary bone neoplasms, and, for pediatric cases, the most common implantation sites are the longest bones. There is suggestion that involvement of the temporal bone occurs in approximately 1 in 546 cases of GCT, the extension to the infratemporal fossa is extremely uncommon. It is important to note that GCT has its peak of incidence between 30-40 years old, more common in females. Even though the incidence of cranial GCT is rare in children, it’s still more typical than in a male adult in this site (the ratio is 2:1 female in the first and second decade of life for 1 male adult). The clinical condition is variable. In cases of involvement of the axial skeleton, there may be complaints of pain, edema, deformities, and restricted mobility. When present in the temporal bone, a patient may complain of retroauricular pain, hearing loss, and weakness of facial muscles.

Some differential diagnoses should be considered, such as giant-cell reparative granuloma, aneurysmal bone cyst, chondrodysplasia, metastatic lesions, and Ewing’s sarcoma. The imaging exams were important to determine the extent of the lesion and support the diagnosis. Histologically, it is a heterogeneous tumor, presenting giant multinucleated cells resembling osteoclasts and spindle cells. In immunohistochemical analysis, CD68 and CD163 markers may be present.

The first line of treatment for cranial GCT is surgical resection, with subsequent reconstruction. Adjuvant treatment can be associated to reduce the risk of neoplasm recurrence. The patient must be monitored after the end of treatment, as GCTs have recurrence rates of 40-60%.

According to the few information available in articles, the first and only treatment line implemented in the case was surgical. After being discharged, he continued in follow up with regular skull MRI and other imaging tests, the most recent MRIs showing a reduction of the lesion’s dimensions, which had previously increased. A possible recurrence continues to be questioned.

A few similar cases were found while investigating the case. One of them, according to Elder et al., was a 2-year-old girl diagnosed with GCT of the right temporal bone, affecting the dura mater as well. The treatment of choice was microsurgery, similar to the current case. After 13 months of follow-up, there were no signs of recurrence. In the same article, the authors reported the case of a 7-week-old female infant diagnosed also with a GCT at the left temporal bone, involving the left external and internal auditory canal, the confluence of sinuses, parotid gland and cerebellopontine angle without metastasis. The treatment was a gross-total resection of the mass. In a 11-month post-operative follow-up, there were no signs of recurrence, but a persistent left facial palsy was detected.

CONCLUSION

This is an important case due to its rarity, new clinical data about the pathologies and the patient’s survival. Possibly, future studies may discuss the optimization of the treatment of these neoplasms, mainly of GCTs of the zygomatic arch, and consequently, the prognosis of these patients.

CONTRIBUTIONS

Jhulia Farinha Maffini and Leonardo Cesar Ferreira Antunes contributed for the article drafting and data acquisition; Maria Eduarda Turczyn De Lucca contributed for the article drafting, review and editing; Felipe Antonio Torres Mazzo contributed for the article drafting, review, editing and figures; Carlos Eliseu Barcelos contributed for the study conception and investigation; Rodrigo Leite de Moraes and Eduardo Talib Bacchi Jaouhari contributed with data collection and investigation; Milena Massumi Kozonoe contributed with data collection; Rosângela Stadnick Lauth de Almeida Torres contributed for the supervision. All the authors reviewed, read and approved the final version to be published.

CONFLICTS OF INTEREST

There is no conflict of interest to declare.

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REFERENCES

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