Giant Temporal Bone Angiosarcoma: Case Report and Literature Review

Angiossarcoma do osso temporal: Relato de caso e revisão da literatura

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Abstract

The authors describe a very rare case of an angiosarcoma originating from the petrous portion of the temporal bone that evolved as an indolent lesion until it became a giant retroauricular mass. A biopsy demonstrated that it was an angiosarcoma. A presurgical embolization from the left occipital and left parietal branches of the left middle meningeal artery was performed, followed by a total resection of the tumor. The patient developed a transient dysphasia during early follow-up, from which, subsequently, she fully recovered. There were no signs of recurrence in the current 3 years of follow-up. Free margins can be achieved even in some giant tumors and remain the most important prognostic factor for soft tissue malignant tumors with intracranial infiltration.

Keywords

► sarcoma
► skull base
► temporal bone
► case report

Resumo

Os autores descrevem um raro caso de angiossarcoma proveniente da porção petrosa do osso temporal, que evoluiu como uma tumoração indolente até tornar-se uma volumosa lesão retroauricular. Foi realizada uma biópsia incisional, cujo diagnóstico foi de angiossarcoma. Foi realizada embolização pré-operatória da lesão pela artéria occipital esquerda e pelo ramo parietal da artéria cerebral média esquerda, seguida de ressecção do tumor. No pós-operatório imediato, a paciente evoluiu com disfagia transitória, recuperando-se completamente durante o seguimento. Não houve sinais de recidiva nos três anos de seguimento pós-operatório. A ressecção com margens livres de tumores de grande volume segue sendo o fator prognóstico mais importante para tumores malignos de partes moles com infiltração intracraniana.

Keywords

► sarcoma
► base de crânio
► osso temporal
► relato de caso

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Introduction

The temporal bone is an unusual site of tumor pathologies. When they occur, they are more likely to be secondary to advanced periauricular skin cancer or to parotid gland tumors. 

Primary tumors that affect the temporal bone are rare. The most typical histology varies with age, since younger patients are likely to have sarcomas while older patients are likely to have carcinomas. Angiosarcoma is a malignant soft tissue neoplasm (accounting for <2% of sarcomas) that arises most often from the skin (~60% of the cases), particularly in the head and the neck, but that can also originate from the soft tissue, viscera, and bones.

Angiosarcoma of the skull is a rare entity with <20 cases reported in the literature by the year of 2013, of which most commonly involving the frontal and parietal bones. Only seven cases involving the temporal bone have been reported. The authors report the case of a patient who presented with an indolent swelling in the left posterior temporal region that became a giant mass whose pathological diagnosis was angiosarcoma. The tumor was completely resected, and the patient evolved with an excellent outcome with no signs of tumor recurrence during 3 years of follow-up. The patient authorized the authors to publish the present case report by signing an informed consent form.

Case Report

A 56-year-old, previously healthy woman, was referred to the department of onco- logical neurosurgery from a general hospital presenting with a huge tumor in the left temporo-occipital region of the skull. She had no personal or family history of cancer. The patient had noticed the lesion 10 months before the medical evaluation, and during that period it grew progressively. The neurological exam of the patient was normal, and she reported no pain in the tumor area. The patient was admitted to the hospital and submitted to imaging investigation. A computed tomography (CT) exam showed an exophytic lesion in the temporal bone, posterior to the external acoustic meatus, displacing the pinna, eroding the petrous portion of the left temporal bone, and presenting an intense but heterogeneous enhancement after contrast infusion (Fig. 1). Magnetic resonance imaging (MRI) demonstrated an isointense pattern of the tumor (compared to brain parenchyma) on T1 images, with a heterogeneous but predominantly hypointense signal on T2 images with small areas of enhancement after gadolinium infusion (Fig. 2). Furthermore, the MRI confirmed that most of the tumor was exophytic; however, there was an intracranial component with dural and parenchymal invasion of the tumor with no signs of edema. The patient was submitted to an incisional biopsy whose pathological diagnosis was angiosarcoma. Surgical resection of the tumor was planned and accepted by the patient and her relatives. Preoperative angiography and embolization were programmed for the patient. An angiography study demonstrated that the tumor had no vascularization from the left internal carotid or from the left vertebral arteries, and its vascularization was exclusively from branches of the left external carotid artery. The main arterial feeders (left occipital artery and parietal branch of the left meningeal artery) were embolized with microparticles (Fig. 3).

The first surgical step was to remove the soft tissue components infiltrated by the tumor (en bloc resection of the muscle and the skin on the tumor topography). Cranietomy was performed by drilling the bone with normal aspect, respecting a margin of 2 cm from the infiltrated bone. Mastoidectomy with skeletonization of the sigmoid and transverse sinuses were performed. The intracranial portion of the tumor was removed as well as the infiltrated dura mater adjacent to the tumor and the intracranial component. The left transverse and sigmoid sinuses infiltrated by the tumor were resected. The left Labbé vein was also infiltrated and had to be sacrificed to remove the tumor with free margins. Watertight duroplasty was performed using fascia latta graft. The skin and subcutaneous soft tissue reconstruction was performed with a trapezious myocutaneous flap (Fig. 4). The postoperative CT scan showed a gross total tumor resection, but the patient developed a left temporal edema and intraparenchymal hematoma in the territory of Labbé drainage secondary to vein infarction (Fig. 5). During the early postoperative period, the patient had a sensitive dysphasia, from which she recovered gradually in four weeks with full recovery. In the 7th day of postoperative follow-up,
the patient was discharged from the hospital, sustaining excellent wound healing and complete neurological recovery. The patient was referred to adjuvant radiotherapy. However, she did not attend to the medical neurosurgery and radiotherapy appointments during the follow-up. In 2020, the authors performed an active search of the patient. She remained asymptomatic and with no signs of tumor recurrence in 3 years of current follow-up.

Discussion

Angiosarcoma is a rare and highly aggressive malignant tumor, originating from lymphatic or vascular endothelial cells. A recent study analyzed 1,250 cases of angiosarcoma exclusively from the head and neck regions; among those patients, there was no case of angiosarcoma involving the temporal bone. Endothelial-derived neoplasms of the temporal bone may range from self-limited benign hemangiomas to tumors of intermediate malignancy, such as hemangioendotheliomas, or highly malignant and aggressive angiosarcomas.

Primary skull angiosarcoma is twice more common in men, with a median age of 32 years old. Previous radiation therapy, arsenic exposure, chronic lymphoedema, and history of trauma are known risk factors for extracranial angiosarcomas. However, primary skull angiosarcoma has no known risk factors. Temporal bone tumors present nonspecific symptoms that are often attributed to inflammatory ear diseases. Therefore, the diagnosis is often delayed, despite the superficial location of the tumor.

The most common presentation of skull base angiosarcoma is a swelling in the affected region. Otorrhea, otalgia, and hearing loss make up a classic triad for temporal bone cancer, but this classic triad is seen only in 10% of the patients who have temporal bone cancer, and it is related to the topography of the tumor, not to its specific histopathology.
Radiographically, skull angiosarcomas usually demonstrate a well-demarcated lytic, hypervascularized, and hemorrhagic mass. However, these neuroradiological features are not pathognomonic, being also seen in other sarcomas or metastasis to the skull. On MRI, angiosarcoma of the temporal bone usually shows an isointense mass compared to cerebral parenchyma on T1-weighted images, and hyperintense on T2-weighted images.

The histologic features of skull angiosarcomas range from well-differentiated to poorly-differentiated tumors. Well-differentiated tumors are seen as abnormal endothelial cells retaining some degree of well-differentiated vascular...
architecture. On the other hand, poorly-differentiated tumors present sheets of abnormal cells with significant hemorrhage and necrosis. Immunohistochemistry is crucial for the diagnosis of poorly-differentiated tumors. Positive staining for the erythroblast transformation specific related gene (ERG) endothelial marker, factor VIII-related antigen, CD31, FLI-1, CD99, S-100 protein, signal transducer and activator of transcription 6 (STAT6) and smooth muscle actin (SMA) are characteristic of angiosarcomas. Among these markers, some are related to vascular and endothelial origin and proliferation (such as CD31, CD34, FLI-1, and ERG), with CD31 being the most specific marker of angiosarcoma.3,11

Because of the rarity of skull angiosarcomas, a gold standard management has not been defined. Therefore, at present, the treatment of angiosarcomas is often individualized. The medical literature suggests that complete surgical excision followed by adjuvant radiotherapy is the most effective treatment.2 This association has demonstrated local control benefit particularly in cases with close surgical margins16 and, based on some retrospective studies, can prolong survival.3

As with any traditional oncologic resection, a free margin excision is the main aim of temporal bone oncological surgery,17 especially an en bloc resection when feasible.

Surgical resection of the tumor can be achieved by a wide local excision (WLE), en bloc lateral temporal bone resection (LTBR), en bloc subtotal temporal bone resection (STBR), total temporal bone resection (TTBR), or near total temporal bone resection (NTTBR).12,17,18

Besides the tumor, adjacent involved tissue should also be included within the resection specimen. In the medical literature, the discussion about the proper technique that should be used is based on cases of epithelial derived tumors, not angiosarcomas. In standard practice, a key point to decide the surgical technique is the relationship of the tumor with the tympanic membrane (TM). Tumors located lateral to the tympanic membrane can be resected by LTBR, including removal of the cartilaginous and bony external auditory meati (with the eardrum), of periauricular soft tissues, of the parotid (with or without the facial nerve), and of neck lymph nodes. An alternative to LTBR is the STBR, which includes the removal of the petrous bone, of the mastoid, of the bony and cartilaginous canals, of the parotid gland with the facial nerve, of part of the mandible, and neck dissection.17 However, if the malignant lesion invades medially to the TM, NTTBR is the best option for total resection.18

The cavity of temporal bone resection is best reconstructed with free-tissue transfer. The defect can also be repaired with local or regional myocutaneous flaps. The reconstruction should provide proper soft tissue covering for the underlying vasculature, dura, bone, and obliterated dead spaces.18

Even though there is a consensus regarding the importance of systemic chemotherapy in advanced and metastatic cases,19 there is no robust data in the medical literature supporting outcome improvement in terms of recurrence-free period or of overall survival with adjuvant chemotherapy. Besides, in the case of skull angiosarcomas, the effectiveness of chemotherapy has not been substantiated.5

Even with optimal treatment, skull angiosarcoma has been associated with transient responses and a poor prognosis. The outcome is poor regardless of the stage of the disease (localized or metastatic). In patients harboring a localized tumor at diagnosis who were submitted to a large en bloc resection, 40% present with tumor relapse during close follow-up.20 The involvement of the meninges and of the brain and the difficulty to perform a complete surgical resection are factors associated with incomplete resection of the tumor, and, therefore, with higher rates of recurrence and a worse prognosis.5 For giant tumors, en bloc resection can be technically very difficult, but free margins are imperative in order to achieve local control.

The present article has some limitations that must be stated. First, it is a case report, which, in a scientific point of view, does not have the same power of evidence as other

Fig. 5 Postoperative computed tomography showing an excellent tumor resection, but the patient presented with a left temporal edema and intraparenchymatous hematoma in the territory of the Labbé drainage secondary to vein infarction.
most sophisticated study designs. Second, the patient was not submitted to adjuvant radiotherapy as most expert opinions in the literature suggest. Actually, the patient was referred to the treatment; however, she did not accept it. Therefore, optimal treatment was not performed, which may increase the risk of late recurrence of the tumor.

However, the present article has some strengths that deserve to be highlighted. The case report highlights significant information about the case, with illustrations of imaging exams demonstrating the relation of the tumor with intracranial and cervical structures, as well as its vascularization. Key steps of the surgical procedure are demonstrated in Figure 4. The literature review presented embraces important information about skull base angiosarcoma, describing the rare involvement of the temporal bone by this type of cancer. Also, the discussion brings information about suggested investigations and treatments for skull base angiosarcoma, based on specialized opinion (since there is no standard protocol due to the rarity of this lesion in the temporal bone). Finally, the present case report and review have one main note: temporal bone angiosarcoma must be treated aggressively, with resection of all involved structures (when feasible). The accomplishment of this goal is the most important factor for the prognosis of the patient.

Conclusion

The authors describe a giant angiosarcoma of the temporal bone with intracranial extension that was completely resected with the sacrifice of the Labbe vein, which was infiltrated to achieve free margins. The patient presented a transient dysphasia with full recovery and no signs of recurrence in the last 3 years. Free margins can be achieved even in some giant tumors and remain the most important prognostic factor for soft tissue malignant tumors with intracranial infiltration.

Conflict of Interests

The authors have no conflict of interests to declare.

References