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Case Report

Hemangiopericytoma/Solitary Fibrous Tumor of the Parietal Bone: A Case Report

Ana Rita Dias Neto[a] Regina Leite[a] David Rothwell[a] Domingos Roda[b] Claudia Sousa[c] Mónica Henriques[a] Paula Alves[a]

[a]Department of Radiotherapy, Portuguese Institute of Oncology of Coimbra, Coimbra, Portugal; [b]Department of Radiotherapy, Algarve Radiation Oncology Unit – Joaquim Chaves Saúde (JCS), Faro, Portugal; [c]Hospital of the Divine Holy Spirit, Ponta Delgada, Portugal

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Abstract
Hemangiopericytoma/solitary fibrous tumor (HPC/STF) is a rare tumor arising from Zimmerman's pericytes and it is characterized by an aggressive malignancy, with a high tendency for local and distant recurrence. The authors report the case of a middle-aged woman with HPC/SFT of the right parietal bone, which is an extremely rare primary location of involvement. The patient presented with a painful deformity of insidious growth at the right parietal region. Assessment with cranial computed tomography scan and magnetic resonance imaging revealed an expansive lesion at the right parietal bone, with exocranial extension and 27 mm of maximal diameter. Craniotomy with gross tumor removal, duraplasty, and cranioplasty was performed, and the diagnosis of HPC/SFT, WHO grade III, was established by pathological and immunohistochemical analysis. The patient was then evaluated for adjuvant radiation therapy and received a dose of 60 Gy (2 Gy/fraction) with 3D conformal radiotherapy to the surgical bed. The adjuvant treatment was uneventful and, after 8 months of follow-up, there was no suspected local or distant recurrence. The rarity of this diagnosis, its aggressive behavior, and the lack of published data posed several challenges for the treatment management.

Correspondence to:
Ana Rita Dias Neto, anardneto@gmail.com

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Introduction

Hemangiopericytoma (HPC) is a rare cancer of the mesenchymal lineage that arises from the modified smooth muscle cells within the walls of capillaries and postcapillary venules called Zimmerman's pericytes [1]. First described in 1942 by Stout and Murray [1], it has shown to most frequently affect the soft tissues of limbs and retroperitoneal space; however, it is also frequently reported amongst primary intracranial tumors. The World Health Organization's (WHO) classification for soft tissues and bone tumors re-termed HPC and solitary fibrous tumors (SFT) as a single entity due to molecular similarities [2]. They both present 12q13 inversions which result in NGFI-A binding protein 2 (NAB2) and signal transducer and activator of transcription 6 (STAT6) gene fusions and STAT6 nuclear expression detectable by immunochemistry (IHC) [3]. The underlying mechanisms for its development remain poorly understood, and there are no known risk factors. Clinical presentation is dependent on primary tumor location, and it is usually characterized by mass-related signs and symptoms. Diagnostic workup is not well defined. Magnetic resonance imaging (MRI) and computed tomography (CT) lack diagnostic precision, with pathological analysis and IHC always being necessary to establish the diagnosis [4, 5]. Also, there are no consensus guidelines for treatment management. Surgery is usually the first approach and is considered the standard of care. Nonetheless, it is recognized that HPC/SFT have a high tendency for local and distant recurrences, and additional treatment strategies have not been shown to be beneficial [6]. Systemic therapies, for instance, have not demonstrated clinical benefit in the adjuvant setting, and there is even evidence of a detrimental effect on survival in certain cases [4]. They are usually reserved for nonoperable patients with locally advanced disease and metastases [4, 7]. On the other hand, despite some conflicting results in the literature, adjuvant radiation therapy (RT) is believed to provide more beneficial outcomes [4, 5, 8]. The authors present a rare clinical case of HPC/SFT of the parietal bone with emphasis on diagnostic workup and treatment management.

Case Report/Case Presentation

A 58-year-old woman presented with a painful mass of insidious growth at the right aspect of the head. No constitutional symptoms, cognitive impairment, or signs of neurological focalization were noted. She took medication for non-insulin-dependent diabetes and had no other relevant medical or surgical history. Upon physical examination, a painful, adherent mass was palpable in the right parietal bone region. A cranial CT scan revealed an intraosseous lesion, with expansive behavior and ill-defined limits centered at the posterior aspect of the right parietal bone (shown in Fig. 1). The lesion projected onto the adjacent parietal cerebral parenchyma with no signs of invasion or vasogenic edema. No other relevant abnormalities were reported. Diagnosis of multiple myeloma, meningioma, or a metastatic lesion was considered. Blood tests, mammography, a full body CT scan, and bone scintigraphy did not reveal any further relevant abnormalities. A cranial MRI confirmed an expansive lesion with exocranial extension and isointense signal in T1 and T2-weighted images, measuring 27 mm (shown in Fig. 1). The patient underwent surgery with right parietal craniotomy and removal of the gross tumor disease, duraplasty, and cranioplasty with titanium. Pathological analysis (shown in Fig. 2) revealed a densely cellular and highly vascularized mesenchymal neoplasm, consisting of elongated cells with sparse and slightly eosinophilic cytoplasm, round to oval nuclei, chromatin of variable density, and occasional evident nucleoli, in a towel-like format, parallel bundles, crisscrossing, or swirling arrangements. Cellular mitosis was observed in some areas with a count of 7–8 per 10 high-power fields, as well as evidence of apoptosis. The tumor had a prominent vascular network with thin, branching

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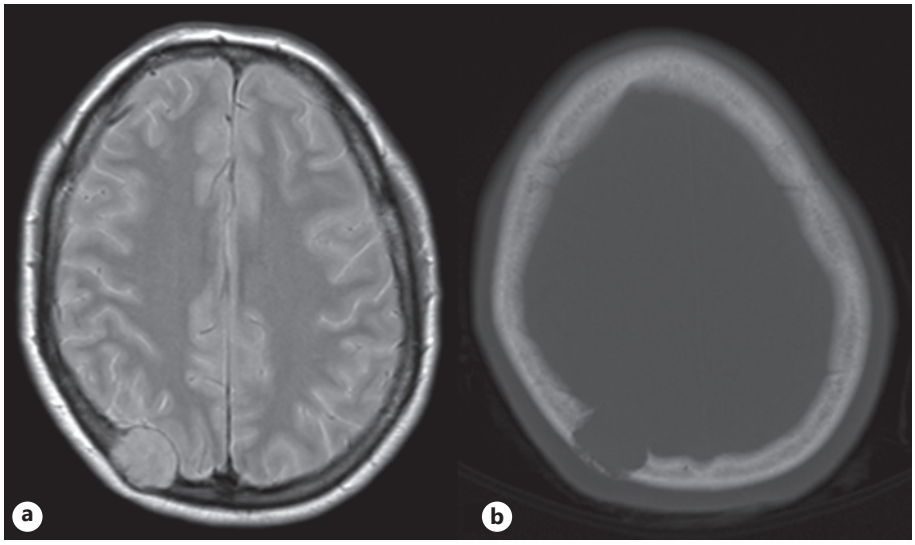

Fig. 1. a Preoperative cranial MRI (T1 sequence) revealing a well-defined, isointense lesion at the right parietal bone with endo and exocranial extension. **b** Preoperative cranial CT scan (bone window) showing a lytic lesion at the right parietal bone with erosion of both cortical layers.

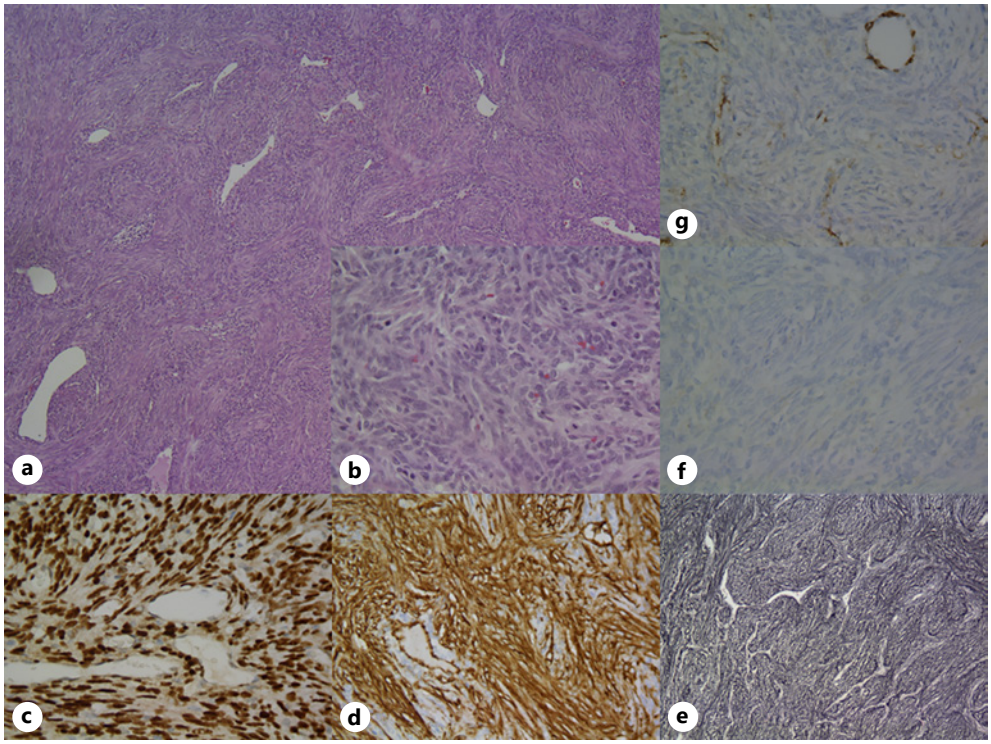

Fig. 2. Pathological and immunohistochemical analysis. **a** Patternless architecture with hypercellular and hypocellular areas and "stag-horn" vessels (hematoxylin and eosin; original magnification, ×50). **b** Bland, ovoid to spindle nuclei cells with hyaline collagen deposition (hematoxylin and eosin; original magnification, ×400). **c** Diffuse nuclear STAT6 expression (original magnification, ×400). **d** Positive immunostaining for CD34 (original magnification, ×400). **e** Gordon and Sweet's silver impregnation showing demonstration of reticulin fibers (original magnification, ×100). **f** Negative immunostaining for EMA (original magnification, ×400). **g** Negative immunostaining for somatostatin 2A receptors (original magnification, ×400).

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vessels, sometimes with a "stag-horn" pattern, and an abundant network of reticulin fibers delimiting isolated cellular elements. It exhibited an expansive, intraosseous growth, destroying the bone throughout its thickness, not reaching any of the surgical resection margins. The IHC study demonstrated positivity of tumor cells for CD34, STAT6, and Bcl-2; negativity for EMA, somatostatin 2A receptors, and desmin; and a proliferative index of 4–5% was estimated with Ki-67. These analyses led to the diagnosis of HPC/SFT, WHO grade III. Residual disease could not be ruled out on MRI due to image artifacts caused by the cranioplasty. The patient was then evaluated for adjuvant RT and received a dose of 60 Gy (2 Gy/fraction) with 3D conformal radiotherapy to the surgical bed. No radiation-induced acute toxicities or complications were reported beyond transient alopecia. After 8 months of follow-up, there was no clinical or radiological evidence of local or distant recurrences.

Discussion/Conclusion

The primary involvement of the bony skeleton in HPC/SFT, as observed in this patient, is extremely rare. It accounts for 4.7% of all primary vascular bone tumors [9]. The etiology is not yet fully understood, although the specific gene fusion NAB2-STAT6 has been shown to play a role by promoting cell proliferation and by activating the early growth response (EGR) gene [5]. The clinical scenario is defined by the location and size of the lesion, and CT scan and MRI play an important role in providing essential information about its local extension. Although there are no distinctive radiological characteristics, focal sclerosis, lytic, and honeycomb patterns have been described [10]. The patient presented a lytic lesion with erosion of both cortical layers of the right parietal bone, which was highly suggestive of malignant behavior. The definitive diagnosis was ultimately ascertained by the pathological analysis and IHC of the resected lesion.

The natural history of this disease is poorly defined. Even though patients may remain asymptomatic until advanced disease, HPC/SFT is known for its aggressive behavior with a tendency for both local and distant recurrences. There is no consensus evidence regarding optimal treatment management. Current knowledge is mostly based on case reports, case series, and retrospective cohorts, with little prospective data published so far [7]. A first approach with surgery has demonstrated significant improvements in overall and cancer-specific survival in patients with intracranial primary disease [4]. According to a recent review, tumor grade and degree of resection are the main determinants of local recurrence (LR). It has also been demonstrated that LR and tumor grade are associated with distant metastasis formation and, hence, lower survival rates [6]. More than 30% of HPC/STF patients experience tumor recurrence after surgery, but recurrence rates of up to 90% at 12 years have been reported in patients with intracranial disease [6, 11]. Recurrences have a strong tendency to occur at the surgical bed and are not uncommonly seen together with distant metastases. Several adjuvant treatment strategies have been explored, but there is as yet no clear role for any of them. The combination of surgery and chemotherapy has been associated with worse clinical outcomes when compared to surgery alone. Previous studies suggested that systemic treatments with anthracycline-based regimens, combinations of temozolomide and bevacizumab or other anti-angiogenic drugs in monotherapy, such as sorafenib, sunitinib, or pazopanib, could be effective in patients with locally advanced and metastatic stages, but their benefit in the adjuvant setting remains to be further explored [4, 7].

The efficacy of adjuvant RT has not yet been fully established. There are multiple single-institution studies demonstrating increased local control but without a consistent demonstration of improvement in survival outcomes [11]. Several radiation treatment modalities have been used, including 3D conformal RT, intensity-modulated radiotherapy, and stereotactic

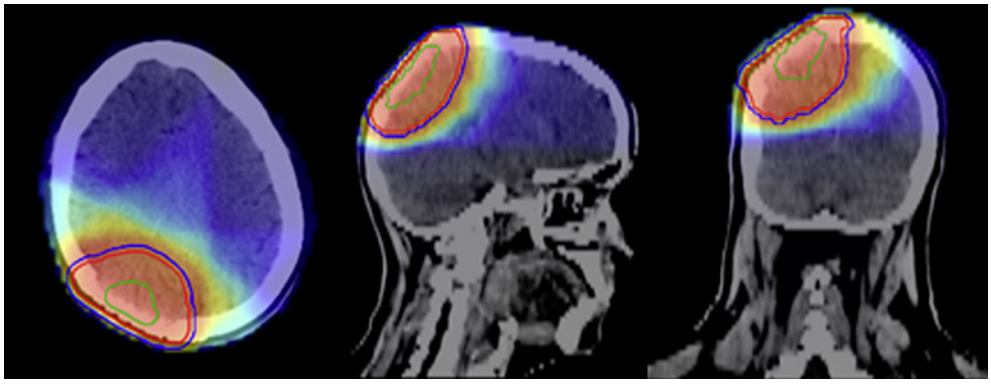

Fig. 3. Treatment volumes contoured for adjuvant RT; tumor bed is represented in green, CTV in red, and PTV in blue. Dose distribution is displayed in color wash.

radiosurgery (SRS). The advantage of dose escalation (≥50 Gy), taking into account the risks and benefits, has been emphasized in terms of local control. In a retrospective analysis, Kim et al. [12] reported that adjuvant RT prevented LR regardless of the degree of resection or histological grade. Patients were treated with doses of 50–60 Gy and no significant radiation-related complications were noted. In a more recent retrospective study, RT failed to improve OS after gross tumor removal, but the improvement in local control was independent of the type of resection. Patients received either daily fractionated doses of up to 54–66.6 Gy or SRS with 18–20 Gy, and a survival benefit was reported in those with subtotal resection or biopsy [11]. The patient reported here received primary surgery with apparent gross tumor removal. Postsurgical MRI could not rule out residual disease, but pathological analysis revealed noninvolved margins. However, due to the expected likelihood of LR of this disease, as well as the age and good performance status of the patient, adjuvant irradiation was planned. SRS was ruled out due to the volume of the tumor bed and because it could not be clearly identified in postsurgical imaging, and a daily fractionated dose of 60 Gy (2 Gy/fraction) with 3D conformal radiotherapy was prescribed. The tumor bed contouring was based on presurgical MRI (T1 sequence) and postsurgical CT-scan. The CTV consisted of a 20 mm margin with editing for anatomical barriers, and an additional 5 mm PTV margin was added to account for setup uncertainties (shown in Fig. 3). No significant radiation-related toxicities were notified, other than transient local alopecia.

This case presents an additional challenge for early LR detection, as MRI, the imaging modality of choice, does not yield an accurate evaluation of the treated volume. CT scans have been used as the alternative modality of assessment. As a local therapy, RT has no role in the prevention of distant recurrence. In case of clinical suspicion, common sites of metastasis, such as the liver, lung, and bone, should be considered for radiologic evaluation. In this case, no evidence of LR or distant metastasis was found after 8 months posttreatment. In order to evaluate the benefit of adjuvant RT, long-term follow-up may be required since high recurrence rates have been reported for up to 12 years. Overall survival rates of 95%, 81%, and 60% were estimated at 1, 5, and 10 years, respectively [13].

Statement of Ethics

The Healthcare Ethics Committee of the Portuguese Institute of Oncology of Coimbra does not require ethical approval to publish an anonymous case report. Written informed consent for publication of this case report and any accompanying images was obtained from the patient.

Conflict of Interest Statement

The authors declare that there is no conflict of interest regarding the publication of this article.

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Author Contributions

Data acquisition: Ana Rita Neto, Regina Leite, and David Rothwell. Drafting of the manuscript: Ana Rita Neto. Critical revision of the manuscript for important intellectual content: Ana Rita Neto, Regina Leite, David Rothwell, Domingos Roda, Claudia Sousa, Mónica Henriques, and Paula Alves. Final approval of the version to be published: Paula Alves.

Data Availability Statement

All data generated and analyzed are included in this article. Further inquiries can be directed to the corresponding author.

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