

Pleomorphic dermal sarcoma of the scalp with intracranial space involvement: management of a rare entity. Illustrative case

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BACKGROUND Exophytic tumors of the calvaria (ETCs) remain a challenging pathology because of their complex management. The authors discuss the case of a woman with a large exophytic mass of the right frontotemporal region and underline their decision-making process on the management of this unique case and possible similar ones.

OBSERVATIONS Neuroradiological findings showed a calvarial tumor with both epicranial and intracranial extension involving the frontotemporal bone with a mixed component (lytic and sclerotic) and dural infiltration with a pseudonodular pattern. A wide en bloc excision from the skin to the dura mater was performed. The compound 5-aminolevulinic acid (5-ALA) was not very useful in identifying the tumor boundaries. One-step cranioplasty and a skin pedicle flap were used to reconstruct the anatomical defect. Acellular dermal matrix was used to repair the uncovered calvaria. Pathological examination confirmed the diagnosis of pleomorphic dermal sarcoma (PDS).

LESSONS In the evaluation of an ETC, PDS should be considered. Wide en bloc excision, if achievable, should be considered the gold standard. The 5-ALA was not helpful, and a pedicle skin flap could be considered as a less invasive alternative to microsurgical reconstruction. The use of an acellular matrix implant on the contralateral exposed galea increases the rate of reconstruction success.

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KEYWORDS pleomorphic dermal sarcoma; exophytic tumors of the calvaria; metastasis; primary intraosseous meningioma; 5-aminolevulinic acid; 5-ALA; acellular dermal matrix

Exophytic tumors of the calvaria (ETCs) remain a challenging pathology because of their complex management. ETCs require a multidisciplinary discussion regarding the differential diagnosis and treatment in order to plan the reconstruction of the anatomical site after resection and adjuvant therapy after surgery.¹ It is mandatory to consider the oncological necessity of achieving radical excision as well as a satisfying skull reconstruction in order to protect the brain while also taking into account the aesthetic implications of the procedure. The reconstruction phase is crucial, especially for ETCs arising from

soft tissues. In these cases, the first phase of surgery must involve en bloc excision of the lesion, including the skin. The most common histotypes classified as ETCs are basal cell carcinoma, squamous cell carcinoma, melanoma, and pleomorphic dermal sarcoma (PDS). PDS is a rare, malignant spindle cell tumor of mesenchymal origin, arising predominantly in the sun-damaged skin of older patients. Here, we discuss the case of a woman with a large exophytic mass in the right frontotemporal region, highlighting our decision-making process.

ABBREVIATIONS 5-ALA=5-aminolevulinic acid; AFX=atypical fibroxanthoma; CT=computed tomography; ETC=exophytic tumor of the calvaria; GTR=gross-total resection; IHC=immunohistochemistry; MRI=magnetic resonance imaging; PDS=pleomorphic dermal sarcoma; PIM=primary intraosseous meningioma; PM=plasmocytoma; TBCT=total-body CT.

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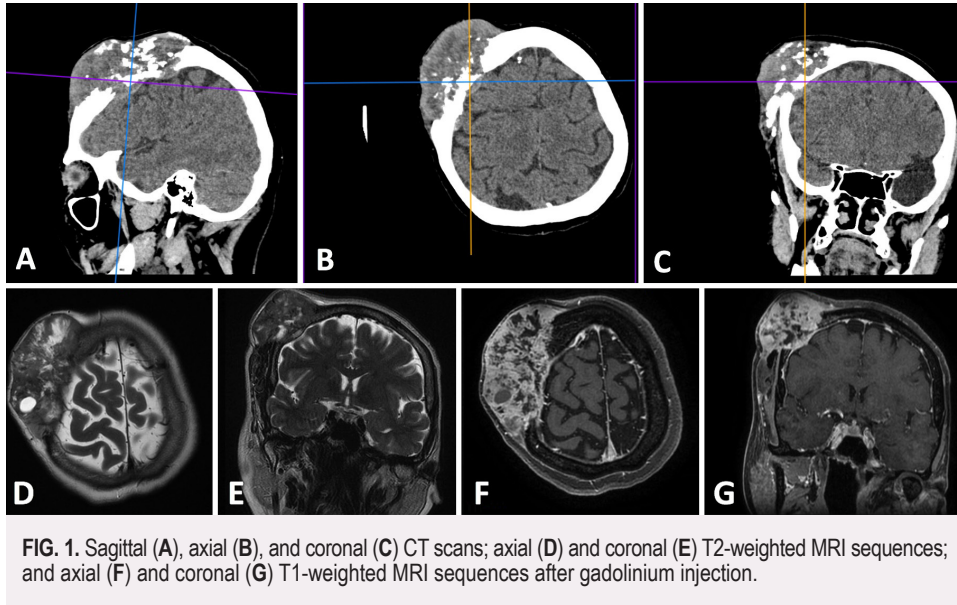


FIG. 1. Sagittal (A), axial (B), and coronal (C) CT scans; axial (D) and coronal (E) T2-weighted MRI sequences; and axial (F) and coronal (G) T1-weighted MRI sequences after gadolinium injection.

Illustrative Case

A 54-year-old female patient presented with a rapidly growing tumor of the head, with clear deformity of the right frontotemporal region. No oncological history was reported. Neurological examination findings were unremarkable. A computed tomography (CT) scan revealed a calvarial tumor with a 10-cm diameter and both epicranial

and intracranial extension involving the frontotemporal bone with a mixed component (lytic and sclerotic). Magnetic resonance imaging (MRI) with and without gadolinium was performed, confirming the presence of a large tumor of the calvaria with intense neovascularization, cystic and necrotic areas, clear enhancement after gadolinium injection, and dural infiltration with a pseudonodular pattern (Fig. 1).

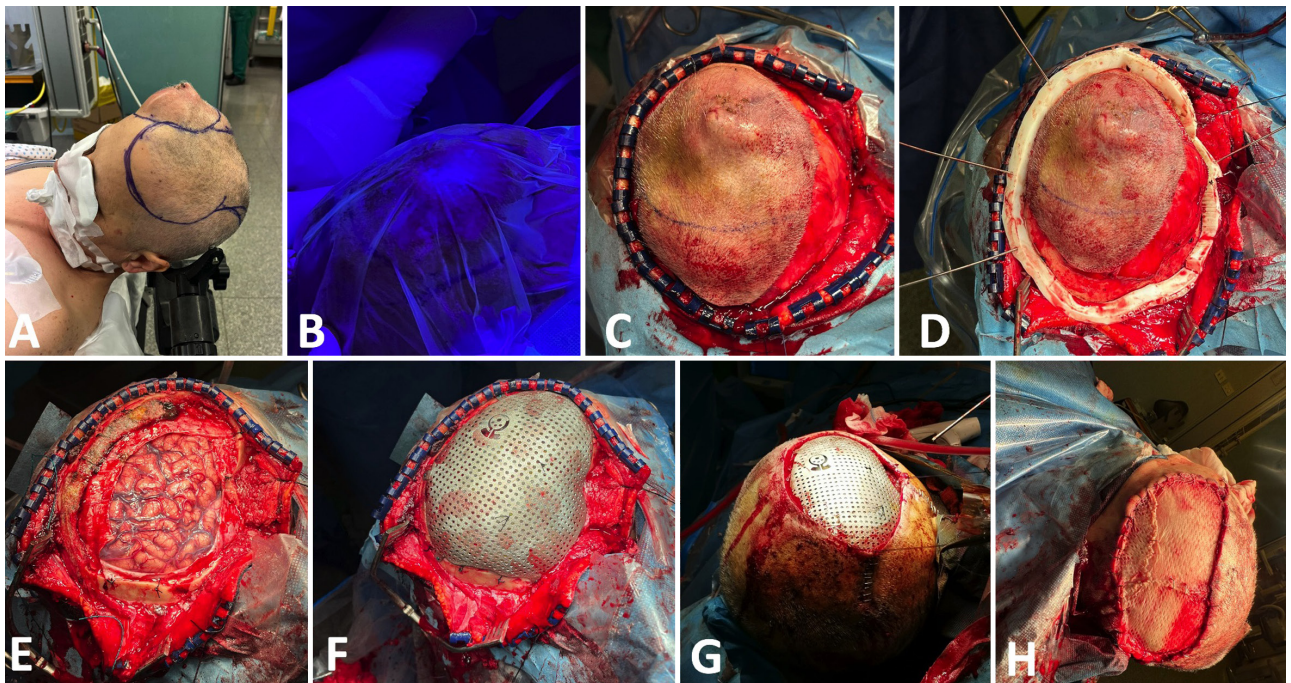


FIG. 2. A: Surgical planning with the contralateral frontoparietal pedicle flap. B: Faint fluorescence of the skin on the tip of the tumor. C: Lozenge incision. D: The craniotomy frame fixed on the healthy bone with K-wires. E: Brain after en bloc tumor removal. F: Cranioplasty. G: Skin defect after lozenge resection. H: Rotation of the contralateral pedicle skin flap and skin graft on the uncovered galea.

We considered atypical primary intraosseous meningioma (PIM), plasmacytoma (PM), and metastases in the differential diagnoses. We proceeded with a total-body CT (TBCT) scan and a punch biopsy of the subcutaneous component of the tumor with local anesthesia. The histological examination confirmed the diagnosis of a sarcoma with a Ki-67 labeling index of 80%. TBCT excluded other tumor localizations but revealed a bilateral peripheral pulmonary microembolism. The patient was started on anticoagulant therapy, and we planned for surgery with a plastic surgeon after 30 days, as suggested by the anesthesiologist and pulmonologist. A titanium cranioplasty (MtOrtho) for one-step resection and reconstruction was planned.

After 2 weeks, the patient presented with skin ulceration caused by further tumor growth. A new CT scan confirmed an increase in the tumor size, necessitating a revision of the cranioplasty project. At this point, we decided to stop anticoagulant drugs and proceed with the operation. We performed surgery with the aid of 5-aminolevulinic acid (5-ALA). The use of this fluorescence dye stems from its well-known application as an intraoperative marker for tumor cells in high-grade gliomas. The purpose of its use in this high mitotic rate tumor, as demonstrated by the biopsy, was to localize the tumor's boundaries to achieve the best possible resection. The patient was placed supine, with the head fixed with a Mayfield clamp. A faint fluorescence was visible on the skin over the top of the tumor under blue light (400 nm). We performed a wide lozenge skin incision away from the tumor border on healthy nonfluorescent skin and a craniotomy following the guide of the planned cranioplasty frame in healthy bone. This approach allowed us to perform en bloc removal of the skin, subcutaneous tissue, muscle, bone, and dura. There was no macroscopic brain involvement. There was no visible fluorescence on the dura removed. Duroplasty with a heterologous patch was performed, and the custom-made cranioplasty was easily positioned. A cutaneous contralateral frontoparietal pedicle flap was rotated to cover the uncovered galea, and a skin graft was taken from the leg and placed on the uncovered galea (Fig. 2). The specimen showed a macroscopic fatty tumor of the skin involving the skull with free margins (Fig. 3).

The postoperative course was characterized by the onset of atrial fibrillation necessitating reintroduction of the anticoagulant medication. Moreover, the mesial part of the skin graft and galea failed to take hold, and we proceeded with the acellular matrix implant (Integra dual layer; Fig. 4). The final graft healed without complications. Postoperative neuroradiological workups confirmed gross-total resection (GTR)

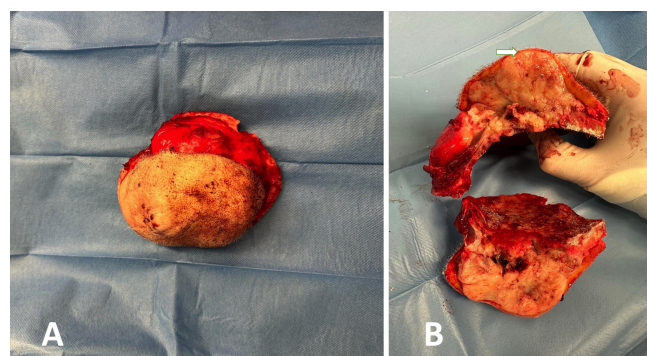


FIG. 3. A: Tumor excised en bloc. **B:** Tumor section highlighting a fatty subcutaneous mass with bone infiltration and erosion. The arrow indicates tumoral infiltration of the skin covering the mass.

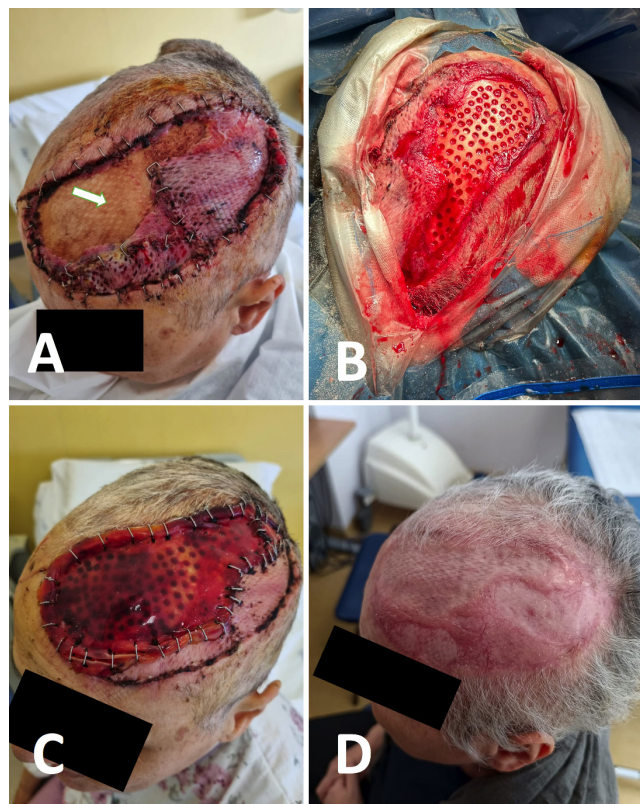


FIG. 4. A: Failure of the frontomesial skin graft to take hold. **B:** Intraoperative view of bone drilled to increase the success rate of the Integra attachment. **C:** Early postoperative result. **D:** Complete healing after skin graft implantation. The arrow indicates the uncovered bone due to the failure of skin graft.

without complications (Fig. 5). Histological examination revealed a PDS with bone and dural infiltration without involvement of the specimen margins confirming the GTR. The patient was still alive 6 months later, with no clinical or radiological signs of recurrence.

Informed Consent

The necessary informed consent was obtained in this study.

Discussion

Observations

ETCs are a rare pathological entity, including PIM, PM, metastases, osteosarcoma, and Ewing sarcoma.¹⁻⁵ In these cases, generally, the skin is not infiltrated by tumor cells and can be spared during excision. However, in cases of skin tumors involving bone and dura, a wide en bloc excision is required.⁶ PDS is a rare mesenchymal tumor that is more common in older people (mean age at presentation 77.6 years) and men. PDS is morphologically similar to atypical fibroxanthoma (AFX) and can be differentiated from it by the presence of subcutaneous invasion, tumor necrosis, lymphovascular invasion, and/or perineural infiltration.⁷ There is no specific immunohistochemistry (IHC) stain to differentiate PDS and AFX. Generally, IHC shows positivity for vimentin, CD10, CD68, and actin and should be negative for pancytokeratins, CD34, and melanoma markers such as melanA, S100 protein, and HMB45. CD10 negativity should suggest the possibility

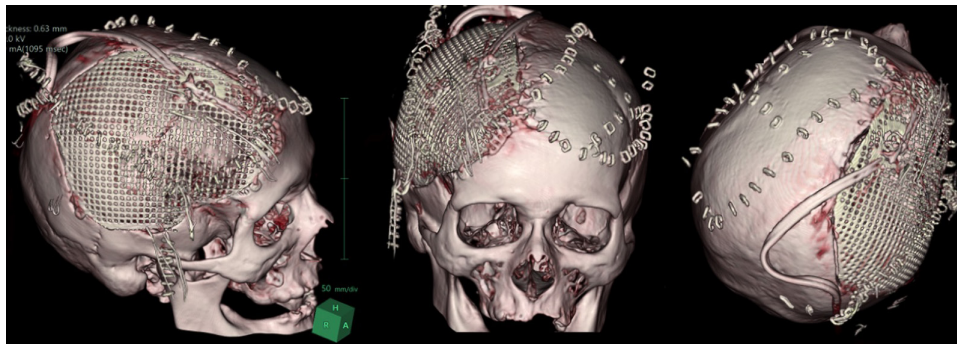


FIG. 5. Postoperative CT scans.

of an alternative diagnosis. The pathological differential diagnosis includes desmoplastic melanoma, spindle cell squamous cell carcinoma, angiosarcoma, and leiomyosarcoma.⁸

The aggressive nature and the high rate of local recurrence of PDS justified surgically removing the skin. The most common PDS locations are the scalp, forehead, and nose. Lesions are typically ulcerated and asymmetric and have poorly defined margins.⁹ At this time, there are no published guidelines on the management and follow-up of PDS.¹⁰ However, general management guidelines for soft-tissue sarcomas can be used as a reference. Surgical treatment with wide local excision remains the first-line therapy for PDS. Although specific guidelines for margin control are not well established, recent data suggest that 95% of cutaneous undifferentiated pleomorphic sarcoma tumors can be cleared with uniform peripheral surgical margins of 3 cm.^{11,12}

To our knowledge, this is the first case of PDS involving the skull and dura. It is also the first time 5-ALA was used to mark the PDS boundaries. In this single experience, however, 5-ALA was not sensitive compared to its use in high-grade gliomas.¹³ Excluding the faint fluorescence on the skin over the tumor, there was no fluorescence on bone and dural layers, despite the clear tumoral infiltration detected by pathologists. The findings in this single case suggest that 5-ALA provided limited assistance in defining tumor margin and guiding the surgery. There is ongoing debate in the literature regarding whether reducing the tumor mass with neoadjuvant radiotherapy or achieving a wide surgical excision should be the first-line treatment. We favored surgery to guarantee and preserve scalp vascularization and achieve the best outcome for the reconstruction phase. Radiotherapy-induced skin atrophy in the presurgical phase could have compromised the vitality of a calvaria pedicle flap and the possibility to reconstruct the patient's anatomical integrity. A microsurgical flap could potentially be an alternative effective reconstruction approach, but in this case we chose to avoid it due to significant patient comorbidities in favor of a less invasive surgery, shorter surgical time, and reduced tissue trauma.

Lessons

When evaluating an ETC, PDS should be considered, especially in cases of a rapidly growing tumor and skin infiltration and/or ulceration. In these cases, a wide en bloc excision should be considered the gold-standard technique. We found that 5-ALA was not useful for identifying tumor boundaries, and a pedicle skin flap could be considered a less invasive alternative to microsurgical reconstruction. In the case of a

pedicle skin flap, we suggest using an acellular matrix implant on the contralateral exposed galea before skin graft in order to increase the rate of reconstruction success.

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Disclosures

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions

Conception and design: Altieri, Limongelli, Massariello, La Rocca. Acquisition of data: Altieri, Massariello, La Rocca, Corvino. Analysis and interpretation of data: Altieri, Massariello, La Rocca. Drafting the article: Altieri, Cuomo, Massariello, La Rocca, Barbarisi. Critically revising the article: Limongelli, La Rocca. Reviewed submitted version of manuscript: Altieri, Limongelli, Barbarisi. Approved the final version of the manuscript on behalf of all authors: Altieri. Study supervision: Vitale, Limongelli, La Rocca, Barbarisi.

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