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Massive Craniofacial Low-Grade Myofibroblastic Sarcoma with Intracranial Extension: A Case Report and Literature Review

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Case Report

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Abstract

Background

Low-grade myofibroblastic sarcoma (LGMS) is a rare malignant mesenchymal neoplasm with myofibroblastic differentiation that most commonly arises in the head and neck region. Although histologically classified as low-grade, LGMS may demonstrate locally aggressive behaviour, such as extensive craniofacial destruction with intracranial extension, making it commonly misdiagnosed and therapeutically challenging.

Case Presentation

: A 27-year-old man presented with a progressively enlarging right-sided craniofacial mass that had developed over nine years. The lesion gradually expanded from the scalp into the right frontotemporoparietal region, resulting in severe craniofacial deformity, orbital compression, and visual disturbance. Radiology examination revealed extensive destruction of the right fronto-parieto-temporo-occipital calvarium, invasion of adjacent intracranial structures, paranasal sinus infiltration, orbital involvement, and significant mass effect with midline shift. Histopathological examination revealed a spindle-cell sarcomatoid neoplasm with myofibroblastic differentiation. Immunohistochemical findings strongly favored LGMS while excluding several important differential diagnoses. Due to the massive size, hypervascularity, and extensive intracranial involvement, the patient underwent multiple staged debulking procedures combined with reconstructive surgery. Despite initial postoperative favorable outcome, progressive neurological deterioration developed following deeper intracranial resections adjacent to the motor cortex. The patient ultimately developed aspiration pneumonia and respiratory failure, leading to cardiopulmonary arrest.

Conclusions

LGMS can be extremely challenging, and accurate diagnosis requires careful clinicopathological correlation and exclusion of mimicking entities. Management requires a multidisciplinary approach and long-term follow-up because of the risk of local recurrence and progressive neurological deterioration. This case highlights the aggressive local behavior that LGMS may exhibit despite its low-grade histological classification and further expands the limited literature regarding intracranial LGMS with massive craniofacial extension.

BACKGROUND

Myofibroblastic sarcoma is a rare malignant sarcoma composed of myofibroblasts. It is typically classified as a low-grade neoplasm and most commonly arises in the head and neck region, extremities, and trunk. In the 2020 World Health Organization (WHO) classification, only low-grade myofibroblastic sarcoma (LGMS) is recognized under the category of fibroblastic and myofibroblastic tumors [1, 2]. LGMS was first reported in 1978 and is histopathologically characterized by diffusely infiltrating spindle cells arranged in fascicles within a hyalinized collagenous stroma, with a known tendency for local recurrence. Although histologically considered a low-grade malignancy, and despite the rarity of extensive craniofacial and intracranial involvement, the tumor may demonstrate aggressive local invasion and cause significant morbidity depending on its anatomical location. Imaging findings on radiography, computed tomography (CT), and magnetic resonance imaging (MRI), with or without contrast enhancement, are generally non-specific and vary according to tumor location [3].

To date, the diagnostic criteria for LGMS remain controversial. Diagnosis primarily relies on microscopic examination, immunohistochemistry (IHC), and, in selected cases, electron microscopy [4]. However, due to overlapping histopathological features with other spindle-cell tumors, LGMS is often misdiagnosed [5]. Furthermore, there is still no consensus regarding the optimal surgical approach or standardized treatment strategy for LGMS [6]. Given the rarity of this disease, a literature review of previously reported head and neck LGMS cases was performed to compare clinical presentation, surgical management, and outcomes. To the best of our knowledge, no previously reported case has demonstrated such massive craniofacial expansion with extensive scalp involvement. This case highlights the exceptionally advanced disease burden and the significant diagnostic and management challenges associated with it, underscoring the importance of early recognition, meticulous clinicopathological correlation, and careful exclusion of histological mimickers.

CASE PRESENTATION

A 27-year-old man presented with a progressively enlarging right-sided craniofacial mass that had been developing over the previous nine years. The lesion initially arose in the right scalp region and gradually extended into the right frontotemporoparietal area. Over time, the mass became extremely large, measuring more than 50 cm in diameter, resulting in marked facial and nasal deformity with compression of the right orbit. The patient complained of progressive heaviness of the head and worsening visual disturbance in the right eye. He had previously undergone two surgical procedures at another institution; however, detailed operative records and pathological documentation were unavailable. The patient was subsequently referred to our institution due to continued enlargement of the mass.

On admission, he was fully conscious with a Glasgow Coma Scale (GCS) score of 15, stable vital signs, and preserved independent ambulation. Physical examination revealed a giant right-sided craniofacial mass involving the scalp and facial region (Fig. 1). Further diagnostic workup was performed, including contrast-enhanced CT, CT angiography, consultation for transarterial chemoembolization (TACE), and planned biopsy. Contrast-enhanced brain CT demonstrated a massive heterogeneous right craniofacial mass measuring approximately 27 × 18 × 27.6 cm, with irregular margins, necrotic and calcified components, extensive destruction of the right fronto-parieto-occipital calvarium, and intracranial extension with invasion of adjacent intracranial structures, producing significant mass effect (Fig. 2).

Histopathological examination revealed a sarcomatoid spindle-cell neoplasm infiltrating the adjacent bone tissue, consistent with a low-grade spindle-cell neoplasm with myofibroblastic differentiation. IHC analysis demonstrated findings consistent with a spindle-cell tumor showing myofibroblastic differentiation, strongly favoring the diagnosis of LGMS, although the results were not entirely specific and therefore did not definitively confirm LGMS as a standalone diagnosis (Additional File Table A1). Nevertheless, the immunoprofile helped exclude several important differential diagnoses, including malignant peripheral nerve sheath tumor (MPNST), neurofibroma, desmoid-type fibromatosis, solitary fibrous tumor, leiomyosarcoma, and meningioma. Other differential diagnoses were considered less likely, including myofibroma, due to the unusually aggressive behavior characterized by destructive infiltrative recurrence and intracranial extension; inflammatory myofibroblastic tumor, due to the absence of reported ALK expression; and unclassified spindle-cell sarcoma with myofibroblastic differentiation, which remained a possible consideration, particularly if atypical morphological features were present.

Subsequent surgical intervention was performed; however, due to the massive size, hypervascularity, and extensive intracranial involvement of the tumor, multiple staged debulking procedures were required between August 2025 and January 2026. Reconstructive procedures with free flap reconstruction were performed in collaboration with the plastic surgery team. Contrast-enhanced MRI was obtained after three debulking procedures (Fig. 3). Serial postoperative brain CT scans are presented in Fig. 4, while detailed radiologic findings for each imaging study are provided in the Additional File Figure A1.

Initially, the patient demonstrated a relatively stable postoperative recovery following the early debulking procedures. However, as subsequent resections involved deeper intracranial components adjacent to the motor cortex, the patient progressively developed neurological deficits, particularly reduced motor strength in the left upper and lower extremities, accompanied by spasticity and stiffness. Ophthalmologic examination also revealed right-sided lagophthalmos with a 3-mm eyelid closure defect, while the remaining cranial nerve functions were preserved. During the course of treatment, the patient developed progressive ventriculomegaly requiring multiple ventriculoperitoneal shunt procedures. Given the advanced disease progression and declining functional status, the patient was eventually transitioned to palliative home care with rehabilitation therapy twice weekly to improve functional capacity. Staged surgery timeline is provided in the Additional File Table B1.

Approximately 11 months after being transferred to our institution, the patient was brought back to the emergency department with acute respiratory distress and deterioration of consciousness. According to the family, the patient had previously remained minimally communicative through head movements but had become progressively less responsive since the previous night. The patient was diagnosed with aspiration pneumonia, progressive intracranially infiltrative myofibroblastic sarcoma, decreased consciousness possibly secondary to hypoxic-ischemic encephalopathy, a hypercoagulable state, and right-sided lagophthalmos with corneal exposure. Despite intensive supportive management, the patient's condition progressively deteriorated. Over the following days, he developed worsening hypoxemia, severe respiratory distress with recurrent desaturation episodes, and hypotension requiring fluid resuscitation. On the third day of admission, the patient developed cardiopulmonary arrest secondary to respiratory failure caused by pneumonia in the setting of an advanced intracranially infiltrative low-grade myofibroblastic sarcoma of the right temporoparietal region.

DISCUSSION

LGMS is a rare soft tissue sarcoma currently classified by the WHO as a low-grade malignant mesenchymal neoplasm with myofibroblastic differentiation and fibromatosis-like features [2]. LGMS most commonly arises in the head and neck region of adults (51.5%), typically during the fourth to sixth decades of life, with a slight male predominance. However, the tumor remains extremely rare overall, accounting for approximately 0.6% of all soft tissue malignancies, with only a limited number of cases reported in the literature [5].

Despite its histologically low mitotic activity, LGMS may demonstrate aggressive local behavior. Nevertheless, intracranial and craniofacial involvement with extensive calvarial destruction is exceptionally uncommon. Most reported cases of head and neck LGMS are localized lesions without significant intracranial extension (Table 1). This case highlights the potentially devastating local aggressiveness of LGMS despite its low-grade histological classification. In the present patient, several unusual and aggressive features were observed, including giant tumor size, infiltrative craniofacial extension with destruction of the frontotemporoparieto-occipital bone, orbital compression, and intracranial extension.

Table 1
Summary of clinical presentation, surgical management, and outcomes of previously reported head and neck L

Authors	Study Design	Year	Case no.	Gender	Age	Site of Tumor	Size (cm)	Intracranial / Bone Extension	Treatment	Loc Rec
Head region										
He et al.[1]	CR	2025	1	F	23	Intracranial (right fronto-parieto-occipital)	5.2 × 4 × 6.1	Yes (extensive calvarial and intracranial extension)	Surgery + RT	(−)
Zhang et al.[7]	CR	2017	2	F	2	Orbit	2.1 × 1.3	Yes (orbital bony involvement possible)	LE	(−)
Oromaxillofacial region										
Nivedhan et al.[13]	CR	2026	5	M	7	Maxilla	NA	Yes (maxillary bone involvement)	CTx → surgery planned	NA
Flores et al.[14]	CR	2025	4	F	30	Mandible	7.5	Yes (cortical bone expansion/perforation)	Block resection + reconstruction	NA
Emanuelli et al.[16]	CR	2022	6	M	11	Right mandible	5.2 × 3.9 × 5	Yes (mandibular bone involvement)	Embolization + radical excision + free flap	(−)
Schwerzmann et al.[9]	CR	2022	7	F	45	Mandible	1.2 × 2	Yes (mandibular bone destruction)	Segmental mandibular resection	(−)
Rahman et al.[4]	CR	2022	8	M	73	Tongue + mandibular alveolus	4 × 1 and 2 × 1	Yes (mandibular alveolar involvement)	LE	NA
	CR	2022	9	NA	NA	Maxilla	Extensive osseous lesion	Yes (maxillary bone involvement)	LE	NA
Jayasooriya et al.[17]	CR	2021	10	F	7	Mandible (intra-osseous)	3 × 4	Yes (mandibular bone involvement)	LE	NA
	CR	2021	11	F	12	Gingiva	NA	No bone extension reported	LE	NA
	CR	2021	12	M	13	Gingiva	NA	No bone extension reported	LE	NA
Jay et al.[23]	CR	2007	1	M	41	Tongue (left lateral tongue/intrinsic lingual musculature)	Primary: 1.5–1.7 cm; recurrence: 0.5 × 0.5 cm	No intracranial or bone extension	Initial excisional biopsy → re-excision for recurrence → wide local excision	**(+)** yea
Neck region										
Sun et al.[24]	CR	2023	13	M	40	Pharynx/parapharyngeal space	7.5 × 7.5 × 3	No	Surgery + pembrolizumab + anlotinib	(+)
Dinker et al.[20]	CR	2023	14	F	15	Left neck	23 × 25	No	Surgery + CTx + RT	(+) tim
Covello et al.[25]	CR	2011	15	F	69	Larynx (true/false vocal cords)	NA	No	LE	(−)
Ni et al.[26]	CR	2011	16	F	41	Epiglottic–glossal surface (larynx)	3 × 2.3 × 2.1	No	Extended excision + partial epiglottis resection	(−)
Coyne et al.[27]	CR	2007	1	M	44	Piriform fossa	Initial: 3 × 2 × 1.3; recurrence: 2.5 × 2.5 × 2.5	No	Initial pharyngeal excision; recurrent tumor treated with radical cervical excision	(+) yea
Takahama et al.[8]	CR	2006	18	NA	NA	Parapharyngeal space	NA	No	WR	NA

CR, Case report; CTx, Chemotherapy; FU, Follow-up; LE, Local excision; LGMS, Low-grade myofibroblastic sarcoma; NSR, No sign of recurrence; LN, Lymph n retrospective series; RT, Radiotherapy; WR, Wide resection

Authors	Study Design	Year	Case no.	Gender	Age	Site of Tumor	Size (cm)	Intracranial / Bone Extension	Treatment	Loc Rec
Head region										
Mentzel et al.[28]	RS	1998	19	F	29	Right supraclavicular	8	No	LE	(−)

CR, Case report; CTx, Chemotherapy; FU, Follow-up; LE, Local excision; LGMS, Low-grade myofibroblastic sarcoma; NSR, No sign of recurrence; LN, Lymph no retrospective series; RT, Radiotherapy; WR, Wide resection

Diagnostic Approach and Differential Diagnosis Workup

The diagnosis of LGMS is challenging because both the clinical and radiological features are nonspecific and frequently mimic several other spindle-cell tumors [5]. In the present case, a definitive diagnosis could not initially be established, and several differential diagnoses were considered. The patient initially presented with a slowly progressive craniofacial mass that had enlarged over approximately nine years, initially suggesting a relatively indolent lesion. However, serial imaging findings increasingly raised suspicion for a locally aggressive mesenchymal neoplasm rather than a benign soft tissue tumor. The diagnostic approach should involve a systematic evaluation of the clinical presentation, imaging characteristics, histopathological findings, and IHC profile [7]. Most patients with LGMS present with a painless swelling or a gradually enlarging mass [8]. Previous studies have reported that LGMS may mimic a broad spectrum of both benign and malignant spindle-cell lesions, morphologically as well as immunophenotypically. Schwerzmann et al. (2022) reported that LGMS may mimic benign lesions such as myofibroma or nodular fasciitis, while Rahman et al. (2022) emphasized the challenges in diagnosing spindle-cell and myofibroblastic tumors in the head and neck region, underscoring the importance of correlating histopathological and IHC findings [4, 9]. However, progressive enlargement over time, recurrence after prior surgery, neurological deficits, infiltrative growth patterns, destruction of adjacent bone, orbital involvement, and intracranial extension should raise suspicion for a malignant soft tissue neoplasm [9–11].

Radiographic features of LGMS generally appear as nonspecific soft tissue masses and may vary depending on the site of involvement. On CT, some LGMS lesions may demonstrate calcification or ossification, and the lesions have been described as osteolytic and destructive masses. On MRI, the lesions are typically hypo- to isointense relative to muscle on T1-weighted imaging, while T2-weighted imaging often demonstrates heterogeneous high signal intensity. Post-contrast T1-weighted imaging may show homogeneous to heterogeneous, predominantly peripheral enhancement [12]. Nivedhan et al. (2025) and Flores et al. (2025) reported that aggressive LGMS lesions may infiltrate and destroy adjacent bone structures, which may explain the extensive calvarial destruction observed in the present case [13, 14]. In addition to massive calvarial destruction, this patient also demonstrated sinus infiltration, cerebellar involvement, and orbital extension.

Definitive diagnosis requires a tissue biopsy. Histopathologically, LGMS is characterized by spindle-shaped myofibroblastic cells arranged in fascicles with infiltrative margins, mild-to-moderate nuclear atypia, and low mitotic activity, supporting the diagnosis of a low-grade spindle-cell neoplasm [4, 7]. The infiltrative growth pattern into osseous structures is also a characteristic feature of LGMS and helps distinguish it from more circumscribed benign myofibroblastic lesions. Furthermore, the relatively low mitotic activity supports a low-grade sarcomatous process rather than a high-grade pleomorphic sarcoma [15].

Several spindle-cell neoplasms share overlapping morphological features with LGMS, including nodular fasciitis, inflammatory myofibroblastic tumor, fibrosarcoma, leiomyosarcoma, malignant peripheral nerve sheath tumor (MPNST), and spindle-cell variants of meningioma. Therefore, the major differential diagnoses in this case included LGMS, MPNST, fibrosarcoma, atypical or malignant meningioma, solitary fibrous tumor, inflammatory myofibroblastic tumor, nodular fasciitis, and other spindle-cell sarcomas [2, 5].

Immunohistochemically, LGMS commonly expresses smooth muscle actin (SMA) and vimentin, supporting myofibroblastic differentiation. However, due to its indolent growth pattern, diagnosis is often delayed until the lesion becomes locally advanced. In this patient, diffuse SMA positivity supported myofibroblastic differentiation and a smooth muscle/myofibroblastic phenotype commonly observed in myofibroblastic tumors, including LGMS. Focal CD99 positivity and weak nuclear TLE1 positivity were considered nonspecific findings that may be observed in various spindle-cell tumors and soft tissue sarcomas. However, negative epithelial membrane antigen (EMA) expression made synovial sarcoma less likely, while also arguing against meningioma and epithelial neoplasms. Negative desmin staining helped exclude true smooth muscle tumors such as leiomyosarcoma. Negative S100 expression helped exclude nerve sheath tumors, including schwannoma, MPNST, and plexiform neurofibroma. CD34 positivity was observed in this patient, which may occasionally be expressed in myofibroblastic tumors but is more commonly associated with solitary fibrous tumor, dermatofibrosarcoma protuberans, spindle-cell lipoma, certain fibroblastic tumors, and vascular tumors. However, negative STAT6 expression made a solitary fibrous tumor less likely. The absence of Ki-67 expression suggested low proliferative activity, supporting a low-grade biological behavior. Negative beta-catenin expression helped exclude desmoid-type fibromatosis. In addition, immunohistochemical staining for fibronectin to detect fibronexus junctions and fibronectin expression may also assist in establishing a more accurate diagnosis without the need for electron microscopy [2].

Multidisciplinary Management, Surgical Strategy and Intraoperative Challenges

Management of giant craniofacial LGMS with intracranial extension requires a multidisciplinary approach because of the complexity of the disease and the involvement of multiple anatomical structures. The neurosurgical team plays a central role in the management of intracranial pressure, decompression of intracranial structures, tumor debulking, and treatment of complications such as hydrocephalus. Plastic surgeons are essential for scalp and soft tissue reconstruction following extensive tumor resection, particularly in cases requiring free flap reconstruction. Ophthalmologists contribute to the evaluation and

management of orbital compression and lagophthalmos in order to prevent further corneal injury and vision loss. Rehabilitation specialists are important for long-term functional recovery, while palliative care teams play a role in optimizing symptom control and quality of life in advanced-stage disease [1, 5, 6, 13, 16].

Complete surgical excision with clear margins is generally considered the primary treatment for LGMS [17]. Kim et al. (2021) described the use of a negative surgical margin strategy with a routine resection margin of approximately 3 cm, adjusted according to tumor location and surrounding anatomical structures [18]. Emanuelli et al. (2022) suggested preoperative arterial embolization prior to surgical resection to reduce intraoperative bleeding risk [16]. However, achieving gross total resection in tumors involving the head and neck region is particularly challenging because of the complex anatomy and potential invasion into critical neurovascular and functional structures [15].

Therefore, preoperative planning should include contrast-enhanced MRI to evaluate soft tissue and intracranial extension, CT scanning to assess calvarial destruction, and CT angiography or digital subtraction angiography to evaluate tumor vascularity. Preoperative embolization should also be considered in highly vascular tumors to reduce intraoperative hemorrhage. In the present case, preoperative imaging demonstrated infiltration of multiple anatomical compartments, including the scalp, calvarium, dura mater, cerebral parenchyma, cerebellum, paranasal sinuses, orbit, and mastoid region. The tumor received its vascular supply predominantly from branches of the right middle cerebral artery (MCA) and anterior cerebral artery (ACA), with involvement of the cortical dural sinuses, transverse sinus, and sigmoid sinus. These findings indicated extensive local invasion and significant tumor vascularity, thereby increasing the risk of massive intraoperative hemorrhage. Consequently, lesions such as the one in this patient often require staged or debulking procedures rather than complete radical excision, as total resection may not be feasible without causing catastrophic neurological injury. Multiple staged debulking procedures were therefore performed over several months in an attempt to reduce tumor burden. One of the resections resulted in the removal of approximately 1400 g of tumor tissue, illustrating the extraordinary tumor burden encountered intraoperatively.

Following complete wide surgical excision, local recurrence and distant metastasis are generally considered uncommon [15, 19]. However, Dinker et al. (2023) reported recurrence rates of 21.4% for tumors smaller than 3 cm and 46.2% for tumors larger than 3 cm, suggesting that a combination of wide excision, plastic reconstruction, and adjuvant radiotherapy may provide the best chance for prolonged disease-free survival and improved quality of life [20]. Nevertheless, the role of radiotherapy and chemotherapy in LGMS remains unclear, and no standardized treatment guidelines currently exist. Several studies support the use of adjuvant chemotherapy and radiotherapy in cases where complete surgical excision is not feasible because of brain invasion or in cases with lymphatic or hematogenous metastasis [7, 8, 18]. Lang et al. (2025) suggested that radiotherapy followed by toripalimab combined with anlotinib may have therapeutic benefit in recurrent LGMS [6]. In contrast, other previous studies concluded that radiotherapy and chemotherapy play limited roles in LGMS outcomes because myofibrosarcoma is often poorly responsive to radiation therapy [7, 16, 21, 22]. In the present case, the patient did not undergo adjuvant radiotherapy or chemotherapy because of the lack of clear consensus regarding standardized treatment strategies, and management was primarily focused on maximal tumor debulking procedures.

Following tumor resection, reconstruction of large scalp and cranial defects is often necessary to achieve adequate soft tissue coverage and reduce the risk of infection and wound breakdown. Reconstruction may be performed using primary closure, local flaps, skin grafts, or free flap techniques [16, 18]. In this patient, reconstructive procedures using free flap microsurgery were required to restore soft tissue coverage and maintain wound integrity.

Postoperative Progression and Complications

Despite initial postoperative stabilization following the earlier debulking procedures, the patient progressively developed significant neurological and systemic complications. As the surgical planes extended deeper into intracranial compartments adjacent to the motor cortex, progressive neurological deficits became increasingly evident. This represents one of the major neurosurgical dilemmas in advanced infiltrative craniofacial tumors: balancing maximal tumor resection with preservation of neurological function. The patient developed worsening left-sided hemiparesis accompanied by spasticity and progressive functional decline. These deficits likely reflected both direct tumor infiltration and cumulative surgical injury involving eloquent cortical regions. The patient also developed multiple ophthalmologic complications secondary to extensive orbital involvement and facial nerve dysfunction, particularly right-sided lagophthalmos, which likely contributed to corneal exposure injury manifested by conjunctival and ciliary injection, corneal opacity, and corneal infiltrates.

Serial imaging demonstrated progressive postoperative changes, including subdural hygroma, subarachnoid hemorrhage, intraparenchymal hemorrhage, transcalvarial herniation, and progressive ventriculomegaly. The patient subsequently required multiple ventriculoperitoneal shunt procedures for the management of hydrocephalus and ventricular enlargement. During the terminal phase of the illness, the patient developed aspiration pneumonia and progressive respiratory compromise requiring tracheostomy and intensive airway management. Recurrent aspiration events, retained secretions, and prolonged immobilization likely contributed significantly to pulmonary deterioration, ultimately resulting in severe respiratory failure, hypoxic injury, and cardiopulmonary arrest.

Limitations and Future Directions

This study has several limitations. First, its single-case nature limits broader conclusions regarding the prognosis, optimal management, and outcomes of intracranial LGMS. Second, the rarity of the disease and the limited available literature for comparison make standardized diagnostic and therapeutic recommendations difficult. Third, the patient's clinical deterioration and death limited the ability to evaluate long-term recurrence and survival outcomes. Future studies should focus on molecular characterization and emerging systemic therapies, including targeted and immunotherapeutic approaches, which may improve diagnostic accuracy and expand treatment options for patients with unresectable, recurrent, or extensively invasive disease.

CONCLUSION

Low-grade myofibroblastic sarcoma (LGMS) is a rare neoplasm occurs predominantly in adults with a preferred location in the head and neck region. LGMS should be considered in the differential diagnosis, particularly when imaging demonstrates progressive soft tissue invasion and destructive osseous involvement. Because the histological and immunohistochemical features may overlap with other spindle-cell neoplasms and no specific diagnostic marker for LGMS currently exists, the diagnostic workup requires careful clinicopathological correlation and exclusion of mimicking entities. Management of advanced craniofacial LGMS requires a multidisciplinary approach, and long-term follow-up is essential because of the high risk of local recurrence and progressive neurological deterioration. Achieving complete surgical resection in advanced craniofacial LGMS can be extremely challenging due to involvement of the eloquent cortex, orbital structures, dural venous sinuses, and skull base anatomy. This case further expands the currently limited literature regarding intracranial LGMS.

Declarations

Ethics Approval and Consent to Participate

Ethical approval was not required at our institution to publish an anonymous case report. Written informed consent was obtained from the patient for participation in this report and for the publication of anonymized clinical information and imaging data.

Consent for Publication

Written informed consent was obtained from the patient for participation in this report and for the publication of anonymized clinical information and imaging data. All authors have read and approved the final manuscript and agree to its submission and publication.

Competing Interests

The authors declare that they have no competing interests.

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Author Contribution

MF: Conceptualization, Formal analysis, Investigation, Project administration, Supervision, Validation, Writing – review & editing. ES: Conceptualization, Data curation, Formal analysis, Investigation, Methodology, Resources, Software, Visualization, Writing – original draft, Writing – review & editing. AS: Formal analysis, Investigation, Project administration, Supervision, Validation. OAW: Formal analysis, Investigation, Project administration, Supervision, Validation. TJ: Supervision, Validation. R: Conceptualization, Formal analysis, Methodology, Supervision, Validation, Writing – review & editing

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Data Availability

The datasets used and/or analysed during the current study are available from the corresponding author on reasonable request.

ABBREVIATIONS

ACA	Anterior cerebral artery
ALK	Anaplastic lymphoma kinase
CD	Cluster of differentiation
CT	Computed tomography
EMA	Epithelial membrane antigen
FU	Follow-up

GCS	Glasgow Coma Scale
IHC	Immunohistochemistry
LGMS	Low-grade myofibroblastic sarcoma
LN	Lymph node
MCA	Middle cerebral artery
MRI	Magnetic resonance imaging
MPNST	Malignant peripheral nerve sheath tumor
NA	Not available
NSR	No sign of recurrence
RT	Radiotherapy
SMA	Smooth muscle actin
STAT6	Signal transducer and activator of transcription 6
TACE	Tansarterial chemoembolization
TLE1	Transducin-like enhancer of split 1
VP	Ventriculoperitoneal
WHO	World Health Organization
WR	Wide resection

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Figures



Figure 1

Preoperative image showing a massive right-sided craniofacial mass expanded from the scalp into the right frontotemporoparietal region, resulting in severe craniofacial deformity, orbital compression, and visual disturbance.

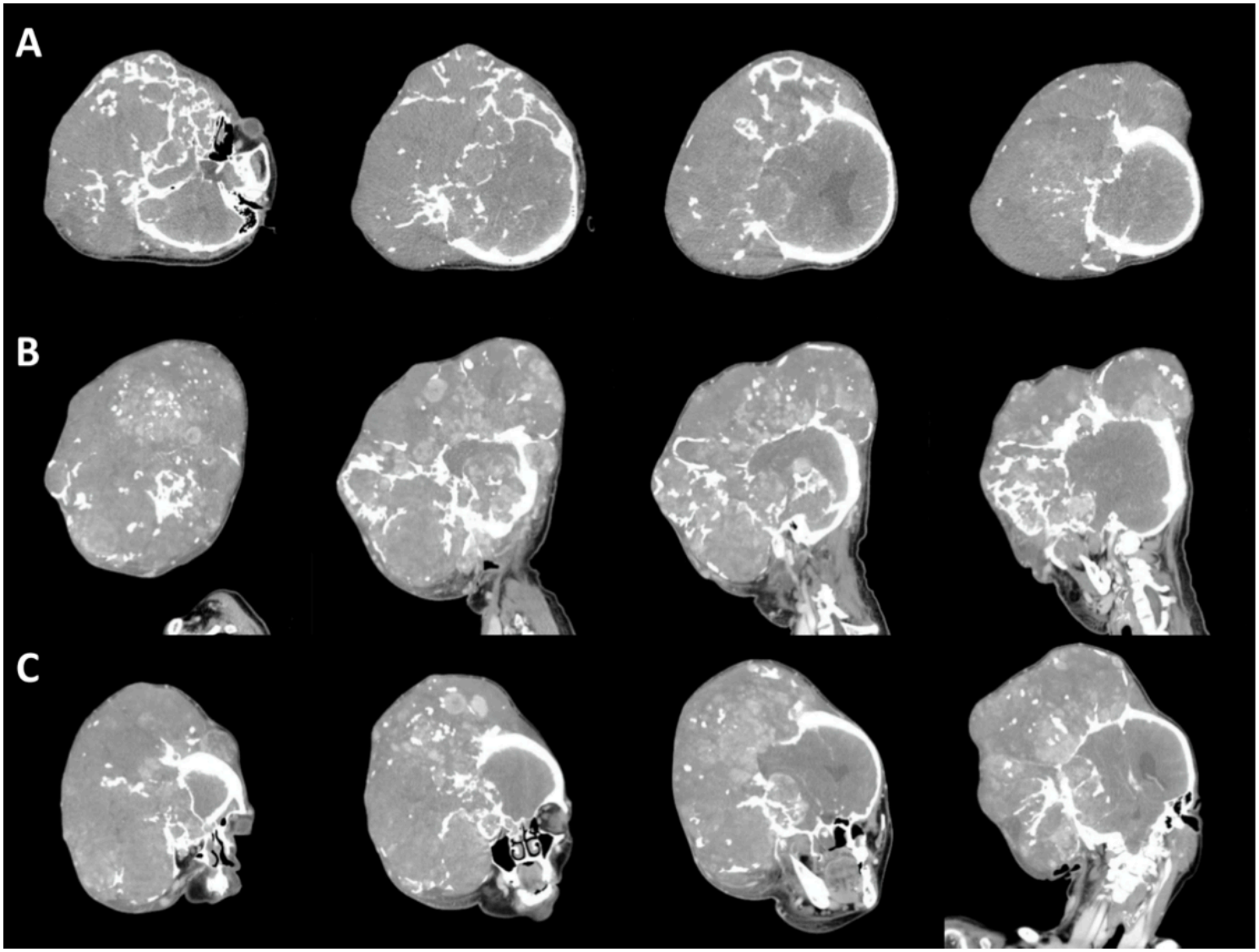


Figure 2

Preoperative contrast-enhanced brain CT in the axial (A), sagittal (B), and coronal (C) planes demonstrated a massive heterogeneous right craniofacial mass measuring approximately 27 x 18 x 27.6 cm with irregular margins, necrotic and calcified components, and extensive destruction of the right fronto-parieto-occipital calvarium. The lesion extended into the right facial-buccal region with invasion of the dura, right frontoparietotemporo-occipital lobes, right cerebellum, paranasal sinuses, mastoid region, and right orbit. Significant mass effect was present, including leftward subfalcine herniation with approximately 0.9 cm midline shift and inferior displacement of the right globe.

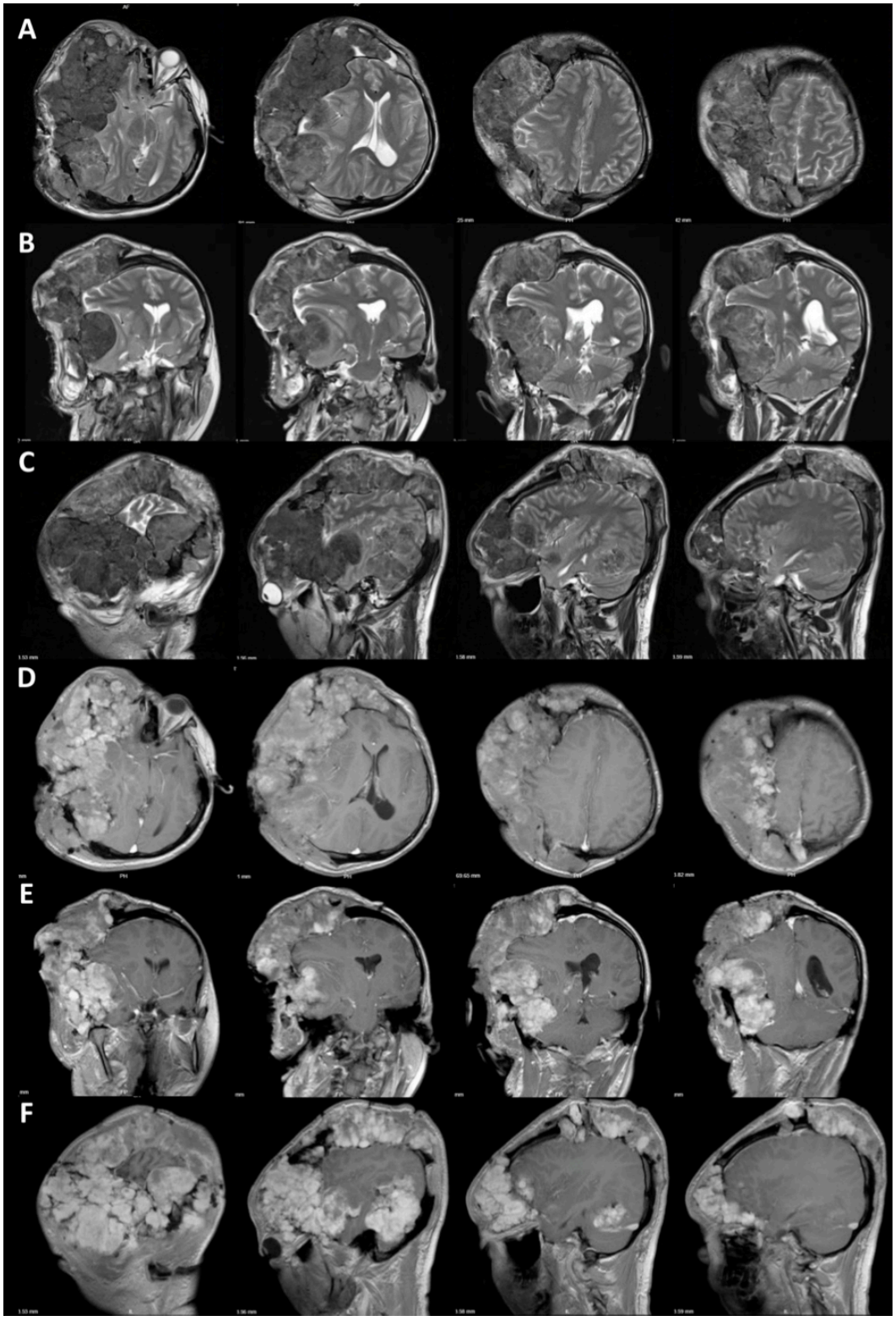


Figure 3

Preoperative brain MRI in T2-weighted axial (A), coronal (B), and sagittal (C) views, as well as contrast-enhanced T1-weighted axial (D), coronal (E), and sagittal (F) views, demonstrating a large inhomogeneous hypodense lesion with ill-defined borders, irregular margins, necrotic and calcified components, and heterogeneous post-contrast enhancement was observed, measuring 20.7 × 13 cm (previously 22.9 × 14.8 cm) in the right cranial region with extension into the right facial-buccal region. The lesion was adherent to the cortical dural sinuses and the right transverse sinus, with destruction of the right fronto-parieto-temporo-occipital calvarium and infiltration into the bilateral frontal sinuses, right ethmoid and sphenoid sinuses, dura mater, portions of the right frontoparietotemporo-occipital lobes and cerebellum, as well as the surrounding right mastoid region and orbit.

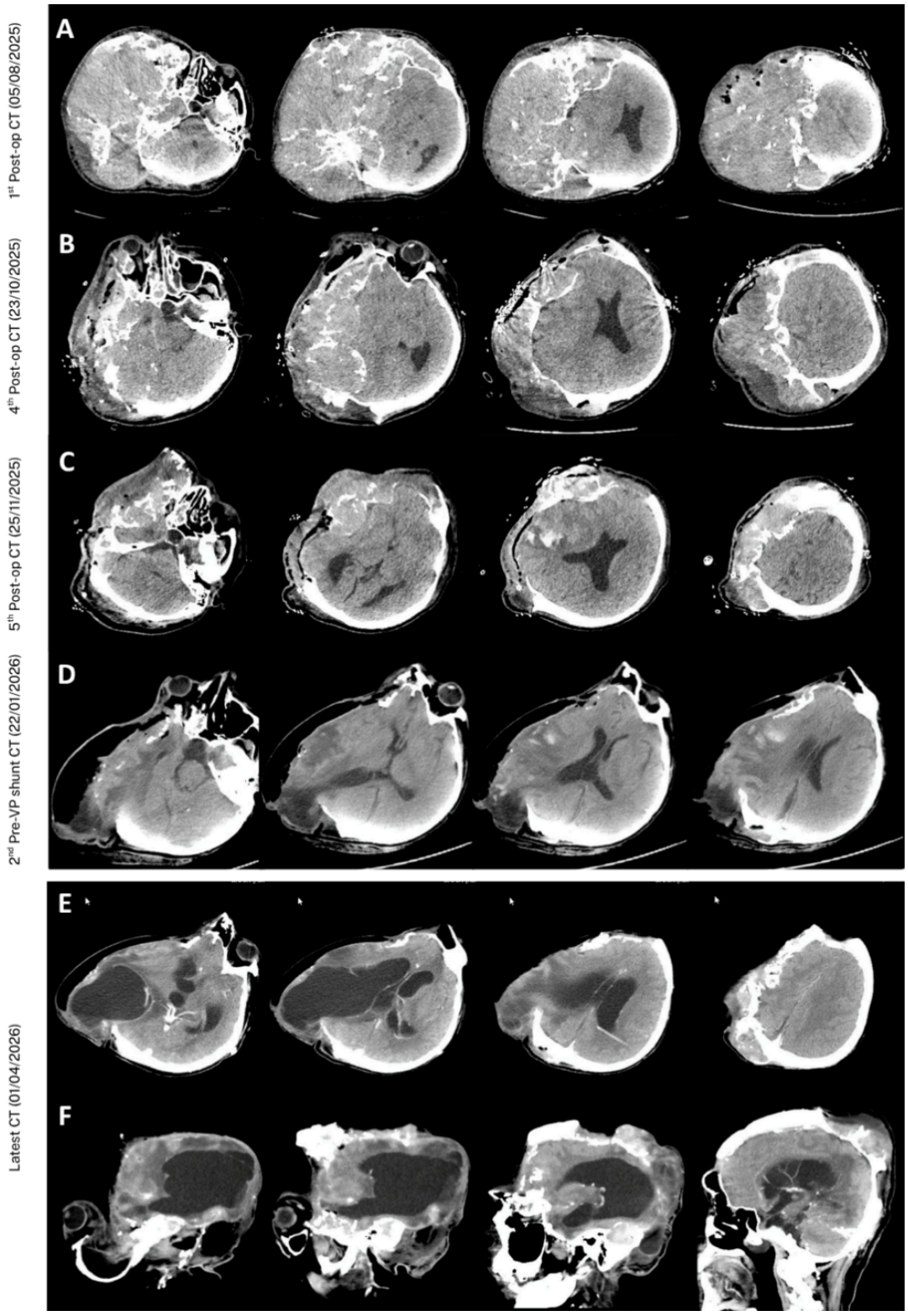


Figure 4

Serial postoperative contrast-enhanced brain CT scans after multiple staged debulking procedures. The first operative CT on 05/08/2025 demonstrated a large heterogeneous necrotic and calcified mass with persistent infiltrative tumor involving the right fronto-parieto-temporo-occipital calvarium, invasion of adjacent craniofacial structures, and marked mass effect (A). CT on 23/10/2025 showed interval reduction in tumor size (B). CT on 25/11/2025 revealed a further decrease in tumor burden, but accompanied by postoperative changes and subarachnoid hemorrhage (C). CT on 22/01/2026 demonstrated partial reduction of the residual tumor and decreased midline shift, but progressive postoperative complications had developed, including transcalvarial herniation, ex-vacuo ventriculomegaly, subdural hygroma, and intraparenchymal hemorrhage (D). The latest CT on 01/04/2026 showed progression of cystic transcalvarial herniation through the right temporoparietal skull defect with worsening ventriculomegaly despite prior surgical interventions (E and F).

Supplementary Files

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- Table1.docx
- Additionalfile1.docx