



(CASE REPORT)



UNDIFFERENTIATED PLEOMORPHIC SARCOMA OF THE SCALP WITH CERVICO-FACIAL EXTENSION MIMICKING SEVERE PYOGENIC CELLULITIS: A CASE REPORT HIGHLIGHTING THE COST OF DIAGNOSTIC DELAY

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ABSTRACT

Background: Undifferentiated pleomorphic sarcoma (UPS), formerly malignant fibrous histiocytoma, is the most common soft tissue sarcoma of late adulthood but only rarely arises in the head and neck. Because of its nonspecific inflammatory appearance, cutaneous fistulization and secondary bacterial colonization, cervico-facial UPS can closely mimic a pyogenic soft-tissue infection, delaying histological diagnosis until the disease has become locally advanced or metastatic.

Case presentation: We report a 69-year-old woman with a two-month history of a progressive left fronto-parieto-temporal scalp mass and left cervico-facial edema, initially treated abroad as a refractory abscess with surgical drainage and broad-spectrum intravenous antibiotics (imipenem and vancomycin), without improvement. On referral, cervico-facial computed tomography (CT) demonstrated a large (62 × 80 × 160 mm), locally invasive soft-tissue mass of the scalp and neck with multilevel necrotic cervical lymphadenopathy, extension to the parapharyngeal space, compression of the internal jugular vein, and infiltration of the sternocleidomastoid muscle. Thoraco-abdomino-pelvic CT showed bilateral iliac and vertebral osteolytic lesions, a small pericardial effusion, and two indeterminate hepatic hypodensities. Histopathology of the scalp mass and cervical/supraclavicular lymph nodes revealed a highly pleomorphic spindle- and giant-cell tumor. Immunohistochemistry showed absence of AE1/AE3, EMA, CD3, CD20, CD34, desmin, myogenin and S100, with preserved nuclear INI-1 and moderate diffuse nuclear TLE-1 positivity, supporting undifferentiated pleomorphic sarcoma. Three of four excised lymph nodes were tumor-positive with capsular rupture, confirming nodal metastasis. Given the extent of locoregional and probable distant disease at diagnosis, the multidisciplinary tumor board oriented the patient toward palliative care; she died a few weeks later.

Conclusion: This case illustrates how cervico-facial UPS can convincingly mimic a non-resolving pyogenic infection, and how the resulting diagnostic delay can allow an initially localized tumor to progress to inoperable, metastatic disease within weeks to months. Failure to respond to adequate antibiotic therapy and drainage, a progressively enlarging mass, and necrotic lymphadenopathy on imaging should prompt early biopsy. Earlier recognition might have

allowed treatment while the disease was still surgically resectable, underscoring that timely histological diagnosis, not the biological aggressiveness of UPS alone, was the modifiable determinant of this patient's outcome.

Keywords: Undifferentiated pleomorphic sarcoma; Malignant fibrous histiocytoma; Scalp tumor; Cervico-facial cellulitis; Necrotic lymphadenopathy; TLE-1; Diagnostic delay; Case report

1 INTRODUCTION

Undifferentiated pleomorphic sarcoma (UPS), previously classified as malignant fibrous histiocytoma (MFH), is a high-grade mesenchymal neoplasm of exclusion, diagnosed only after a systematic immunohistochemical panel has ruled out a definable line of differentiation (epithelial, lymphoid, melanocytic, muscular, vascular, or a specific sarcoma subtype). It accounts for a substantial proportion of adult soft tissue sarcomas of the extremities and retroperitoneum, but head and neck involvement is distinctly uncommon, most often affecting the scalp and facial skin of elderly patients.

Because the scalp and cervico-facial region are richly vascularized and frequently colonized or traumatized, an ulcerated or fistulized UPS in this location can be clinically indistinguishable from a pyogenic abscess or necrotizing cellulitis, particularly once the lesion becomes secondarily infected. This overlap creates a substantial risk of diagnostic delay: patients may undergo repeated incision and drainage procedures and prolonged empirical antibiotic courses before a malignant etiology is even considered, allowing local progression and regional or distant metastatic spread. We report an illustrative case of scalp UPS with cervico-facial extension and metastatic cervical lymphadenopathy that was initially managed for two months as a refractory infectious process. The patient's disease had already progressed beyond curative options by the time of histological diagnosis, and she died within weeks of presentation to our institution — an outcome we use here to argue for a lower threshold for biopsy in atypical, treatment-refractory cervico-facial masses.

2 CASE PRESENTATION

A 69-year-old Moroccan woman, with a past medical history of arterial hypertension and hysterectomy performed 20 years earlier for a uterine fibroid, presented with a two-month history of a progressively enlarging left fronto-parieto-temporal scalp swelling accompanied by local inflammatory signs and a deterioration of her general condition.

While abroad, she underwent surgical drainage of a presumed scalp abscess and received a prolonged course of broad-spectrum intravenous antibiotics (imipenem and vancomycin) without clinical improvement; the mass continued to enlarge despite therapy. She was subsequently referred to the Department of Plastic and Reconstructive Surgery at Mohammed VI University Hospital, Tangier, for further management.

2.1 Clinical Findings

On admission, physical examination revealed:

- A large left fronto-parieto-temporal mass, violaceous, painful and fluctuant on palpation, with a cutaneous fistula discharging sero-hemorrhagic fluid (Figure 1A).
- Diffuse left facial and cervical edema extending to the supraclavicular region (Figure 1B).
- No facial nerve palsy.
- Normal mouth opening; unremarkable endobuccal examination with patent Stensen's and Wharton's duct orifices and a free oropharynx.
- No stridor or airway compromise.



Figure 1: Clinical presentation at admission. (A) Violaceous, fluctuant fronto-parieto-temporal scalp mass with a central fistulous opening. (B) Diffuse left cervico-facial edema extending toward the supraclavicular region. (Periocular region masked to preserve patient anonymity.)

2.2 Investigations — Imaging

Cervico-facial CT showed a large, locally invasive, heterogeneous soft-tissue mass centered on the left sternocleidomastoid region, poorly limited, lobulated and irregular in contour, measuring approximately 62 × 80 × 160 mm (transverse × antero-posterior × cranio-caudal), with heterogeneous post-contrast enhancement and internal hypodense necrotic areas. Its extension pattern was as follows:

- Medially: extension into the retrostyloid and prestyloid parapharyngeal fat with a discrete mass effect on the oropharynx, and compression of the internal jugular vein, visualized proximal to the jugular bulb, without evidence of thrombosis.
- Anteriorly: contact with the posterior border of the left mandibular ramus and voluminous extension into the superficial lobe of the ipsilateral parotid gland, abutting the masseter muscle.
- Superiorly: contact with the external auditory canal (with loss of the fat plane) and multilobulated infiltration along the outer table of the skull without underlying osteolysis, extending into the temporal and fronto-parietal scalp bilaterally as a bourgeoning, focally ulcerated thickening measuring up to 36 mm.
- Inferiorly: extension along the peri- and paravertebral space to the level of the ipsilateral clavicle, with mild deviation of cervical structures to the right.
- Posteriorly: invasion of the occipital musculature and soft tissues, encasing (without occluding) the occipital branch of the external carotid artery, which remained patent.

Multiple heterogeneous cervical lymphadenopathies were identified in the submandibular, jugulo-carotid, parotid and spinal accessory chains, the majority with necrotic centers, the largest measuring 18 mm in short axis (left submandibular level). Necrotic left axillary lymphadenopathy (17 mm short axis) was also noted, together with tumefaction and heterogeneity of the left submandibular gland. The parotid and right submandibular glands, thyroid, larynx and nasopharynx were unremarkable, and no suspicious osseous lesion was seen on this study.

Thoraco-abdomino-pelvic CT performed for locoregional and distant staging confirmed infiltration of the left sternocleidomastoid muscle with jugulo-carotid and spinal-accessory lymphadenopathy, and additionally demonstrated a pericardial effusion of mild abundance (7.2 mm); two small, difficult-to-characterize hypodense hepatic lesions (segment VII, up to 4.6 mm); a simple left renal cyst (Bosniak I, 9 × 12 mm) and calcified mesenteric lymph nodes, both considered incidental; and, on bone windows, bilateral iliac osteolytic lacunae with an osteocondensing focus of the left pubic ramus, a lytic lesion of the D11 vertebral body and left 11th rib (Lodwick grade Ia), and a lytic lesion of the L3 vertebral body without marginal sclerosis (Lodwick grade Ib) — findings raising concern for osseous metastatic disease.



Figure 2: Evolution of the scalp lesion over the course of hospitalization, from early post-drainage appearance (A) through subsequent reviews (B, C) to the mass at multidisciplinary tumor board presentation (D), illustrating continued local progression despite intervention.

2.3 Laboratory Findings

Table 1: admission laboratory findings

Parameter	Result	Interpretation
Hemoglobin	9.2 g/dL	Anemia
Leukocyte count	12,390 /mm ³	Leukocytosis
Platelet count	Decreased (exact value not available)	Thrombocytopenia
LDH	368 U/L	Elevated
HIV, HBV, HCV serology	Negative	Infectious workup negative
Antinuclear antibodies (ANA)	Negative	Autoimmune workup negative

2.4 Therapeutic Intervention

Given the lack of response to prolonged antibiotic therapy, the patient underwent surgical drainage of the fronto-parietal collection together with excisional biopsy of cervical and supraclavicular lymph nodes, and, subsequently, an incisional biopsy of the left sternocleidomastoid muscle mass to obtain a definitive histological diagnosis.

2.5 Histopathological and Immunohistochemical Findings

Histological examination of the intramuscular tissue sample showed skeletal muscle dissociated by a diffuse, sheet- and lobule-forming mesenchymal proliferation of undifferentiated appearance. Tumor cells were large and markedly pleomorphic, alternating between epithelioid and globoid morphologies, with anisokaryotic, hyperchromatic, nucleolated nuclei (severe atypia) and abundant eosinophilic cytoplasm; the stroma was thin and vascular, and no analyzable tumor necrosis was identified in this sample (Figure 3A–B).

Examination of the lymph node excision specimen identified four lymph nodes, three of which were replaced by tumor (3N+/4N), with images of capsular rupture consistent with extranodal extension.

A broad immunohistochemical panel was performed to exclude alternative lines of differentiation:

Table 2 : Immunohistochemical markers

Antibody	Result
AE1/AE3 (pan-cytokeratin)	Negative
EMA	Negative
CD3	Negative
CD20	Negative
CD34	Negative

Desmin	Negative
Myogenin	Negative
S100	Negative
INI-1 (SMARCB1)	Retained nuclear expression
TLE-1	Moderate, diffuse nuclear expression

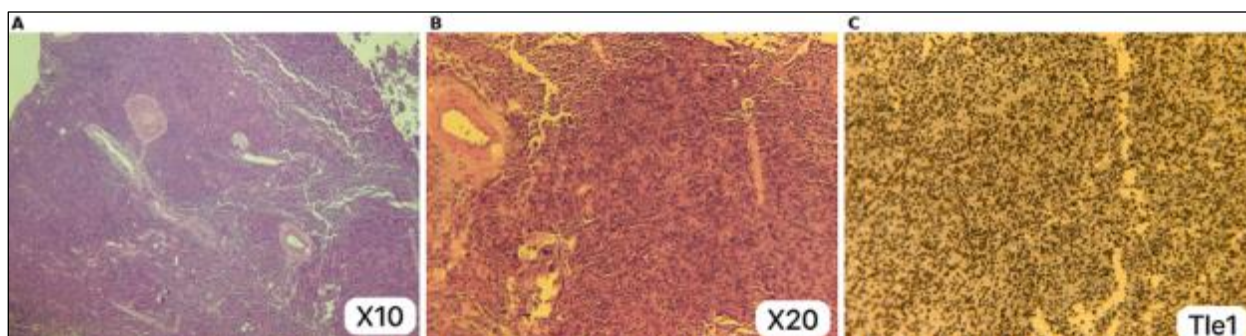


Figure 3: Histopathology of the scalp/intramuscular tumor mass. (A) Low-power view (×10, H&E) showing a diffuse, sheet-like proliferation of pleomorphic mesenchymal cells with a thin vascular stroma. (B) High-power view (×20, H&E) demonstrating marked cytological pleomorphism with large, hyperchromatic, nucleolated nuclei and abundant eosinophilic cytoplasm. (C) Immunohistochemistry showing moderate, diffuse nuclear positivity for Tle-1.

This panel excluded carcinoma (AE1/AE3, EMA negative), lymphoma (CD3, CD20 negative), vascular differentiation (CD34 negative), rhabdomyosarcoma (desmin, myogenin negative) and melanoma (S100 negative). Retained INI-1 expression argued against epithelioid sarcoma and other SMARCB1-deficient tumors. Taken together with the markedly pleomorphic, non-lineage-specific morphology, this immunoprofile was considered consistent with undifferentiated pleomorphic sarcoma.

2.6 Multidisciplinary Discussion and Diagnosis

The case was presented at the multidisciplinary head and neck oncology tumor board (Cancérologie – Chirurgie Maxillo-Faciale), where the initial clinical and radiological differential diagnosis had included lymphoma and rhabdomyosarcoma; both were excluded on immunohistochemistry as above. The final diagnosis retained was undifferentiated pleomorphic sarcoma of the scalp with cervico-facial soft-tissue extension and metastatic cervical lymphadenopathy (nodal capsular rupture), in a patient with radiological findings suspicious for additional axillary nodal, hepatic and osseous involvement — a presentation initially masquerading as refractory cervico-facial cellulitis.

2.7 Outcome and Follow-up

By the time histological diagnosis was established, the disease was already locally unresectable, with probable multi-site metastatic spread (axillary nodal, hepatic and osseous involvement on imaging). Curative-intent surgery was no longer a feasible option, and the multidisciplinary tumor board oriented the patient toward best supportive/palliative care. She died a few weeks after diagnosis, approximately three months after the onset of her initial symptoms. This rapid, fatal course — occurring while the tumor was still being treated as an infectious process for a substantial part of that interval — is the central clinical lesson of this report: the window during which surgical cure may have been possible was likely lost during the period of empirical antibiotic therapy and repeated drainage.

3 DISCUSSION

UPS is the diagnosis given, by definition, to a pleomorphic sarcoma that shows no identifiable line of differentiation on a comprehensive immunohistochemical work-up; it therefore remains a diagnosis of exclusion rather than a distinct histogenetic entity. In the head and neck, UPS is rare, and when it does occur, the scalp is one of its more frequent sites, likely reflecting the cumulative effect of prior irradiation, chronic sun exposure, or scarring in some cases, although no such predisposing factor was identified in this patient.

The clinical overlap between cervico-facial UPS and pyogenic infection is a recurring theme in the limited literature on this presentation. Tumor necrosis, secondary bacterial colonization of an ulcerated or fistulized surface, and a fluctuant,

erythematous, tender mass can closely reproduce the semiology of an abscess or cellulitis, particularly in elderly patients in whom malignancy may not be the first diagnostic consideration. In the present case, this mimicry led to surgical drainage and a prolonged course of broad-spectrum antibiotics abroad before the patient was referred for further work-up, resulting in a diagnostic interval of roughly two months during which the tumor progressed from what may initially have been a localized, potentially resectable lesion to one with deep cervical, nodal and probable distant involvement.

This trajectory is, in our view, the most clinically important aspect of the case. UPS is a high-grade sarcoma with an inherently aggressive natural history, but the interval between symptom onset and histological diagnosis was dominated by antimicrobial and drainage procedures directed at a presumed infection rather than by oncologic staging and treatment. By the time the diagnosis was confirmed, the tumor already showed radiological features of locoregional unresectability and probable multi-organ spread, and the patient's disease course culminated in death within weeks, under palliative care. While it cannot be established with certainty that earlier biopsy would have changed this outcome, the sequence of events — rapid clinical deterioration coinciding with a period during which the true diagnosis was not being addressed — is consistent with a missed opportunity for earlier, potentially curative intervention, and reinforces that diagnostic delay, not merely tumor biology, may have determined this outcome.

Several clinical and radiological features should be regarded as red flags for an underlying malignant process rather than simple infection, and should prompt biopsy rather than continued antimicrobial escalation:

- Absence of clinical improvement despite adequate, culture-guided antibiotic therapy.
- Progressive enlargement of the mass despite repeated incision and drainage.
- Necrotic-appearing regional lymphadenopathy on cross-sectional imaging.
- An infiltrative, poorly defined soft-tissue mass with muscular or deep-space extension on CT or MRI, rather than a simple, well-circumscribed fluid collection.

On immunohistochemistry, the moderate diffuse nuclear expression of TLE-1 observed in this tumor merits specific comment. TLE-1 is classically used as a supportive marker for synovial sarcoma, in which it typically shows strong, diffuse nuclear staining; however, TLE-1 positivity is not entirely specific to synovial sarcoma and has been reported, usually with weaker or more heterogeneous intensity, in a subset of other spindle-cell and pleomorphic sarcomas, including some cases of UPS and malignant peripheral nerve sheath tumor. In this case, the moderate (rather than strong) staining intensity, combined with the markedly pleomorphic — rather than uniformly spindled and monomorphic — cytomorphology and retained INI-1 expression, favored UPS over synovial sarcoma, but this distinction illustrates a genuine diagnostic pitfall: TLE-1 positivity alone should not be over-interpreted without full morphological and immunophenotypic correlation, and, where available, molecular confirmation (e.g., SS18 break-apart FISH) in cases where synovial sarcoma remains a serious differential.

The regional and probable distant spread observed in this patient — necrotic cervical and axillary lymphadenopathy with capsular rupture, and radiological findings suspicious for hepatic and osseous involvement — is also noteworthy, since lymphatic and distant metastasis, while recognized in pleomorphic and epithelioid sarcoma subtypes, are associated with substantially worse prognosis and narrow the therapeutic options toward systemic and palliative strategies rather than curative wide local excision, as ultimately occurred in this patient.

Management of localized UPS is primarily surgical, with wide margins and consideration of adjuvant radiotherapy for high-grade or deep-seated lesions; systemic chemotherapy is generally reserved for metastatic, unresectable, or high-risk disease. In the cervico-facial region, the proximity of critical neurovascular structures often narrows the margin for truly wide resection even when the tumor is localized, which makes early diagnosis — while the lesion is still small and confined — proportionally even more important than in extremity UPS, where more generous margins are typically achievable.

4 CONCLUSION

This case underscores that not every non-resolving, fluctuant cervico-facial mass is infectious in origin, and that the cost of assuming otherwise can be measured in lost time that a high-grade sarcoma does not allow. Failure of appropriate antibiotic therapy and repeated drainage, an infiltrative pattern on imaging, and necrotic regional lymphadenopathy should prompt early tissue biopsy rather than continued antimicrobial escalation. In this patient, the interval spent treating a presumed infection coincided with progression from a presentation that may have been surgically manageable to one that was not; she died within weeks of her eventual diagnosis. Multidisciplinary evaluation, comprehensive immunohistochemical characterization, and a low threshold for biopsy in atypical, treatment-refractory cervico-facial masses are essential to avoid this trajectory in future patients.

Learning Points

- Not every fluctuant, inflammatory cervico-facial mass is infectious; malignancy must remain in the differential, particularly in elderly patients.
- Failure of adequate antibiotic therapy and repeated surgical drainage to control a soft-tissue mass should prompt early biopsy rather than continued antimicrobial escalation.
- Undifferentiated pleomorphic sarcoma can closely mimic aggressive cellulitis or a chronic abscess, especially when ulcerated or fistulized.
- A broad immunohistochemical panel is required to establish UPS as a diagnosis of exclusion; isolated markers such as TLE-1 can be misleading if interpreted in isolation from morphology and the full panel.
- Necrotic regional lymphadenopathy and an infiltrative pattern on cross-sectional imaging are red flags warranting oncologic work-up.

Diagnostic delay in cervico-facial sarcoma can convert a potentially resectable tumor into unresectable, metastatic disease within weeks to months; in this patient, that interval proved fatal, illustrating that early biopsy is not a procedural formality but a determinant of survival.

STATEMENTS ON COMPLIANCE WITH ETHICAL STANDARDS*Acknowledgments*

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Disclosure of conflict of interest

The authors declare that they have no competing interests.

Statement of informed consent

As the patient is deceased, written informed consent for publication of this case report and accompanying clinical images has been obtained from the patient's next of kin, per the target journal's policy. Clinical photographs in this draft have been cropped to remove identifying overlays and the periorcular region has been masked pending final consent

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