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Review article

Primary undifferentiated sarcoma of the meninges: A case report and comprehensive review of the literature

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ABSTRACT

Background and importance: Sarcomas make up 1% of all cases of adult cancer, with 5–10% of those classified as undifferentiated pleomorphic sarcomas (UPS/PUS) and 0.1–4.3% primary intracranial sarcomas. Intracranial undifferentiated sarcoma is characterized by an earlier age of onset and generally poorer prognosis compared to extracranial undifferentiated sarcomas. Current therapies involve surgical excision with wide margins and radiotherapy, with minimal data available regarding the efficacy of chemotherapy.

Case description: A 79-year-old man with a history of remote superficial bladder cancer presented with a large frontal scalp lesion. A biopsy was initially attempted by a dermatologist in the outpatient setting, but a follow-up CT scan revealed a skull-eroding, enhancing soft tissue lesion. Neurosurgical treatment revealed an undifferentiated sarcoma. The patient underwent adjuvant radiation therapy of 59.4 Gy fractionated over 45 days following surgery. Follow-up brain MRIs at 1-, 6-, 9-, 12-, 15-, 21-, and 27 months after surgery have not shown any indications of local recurrence or tumor metastasis. Despite the high propensity that undifferentiated sarcomas have for recurrence and metastasis and the patient's advanced age, this patient remains uniquely disease-free.

Conclusion: We provide a description of an unusual case and comprehensive literature review of UPS to clarify the hallmarks of the disease, identify the difficulties in diagnosis, and provide a summary of therapies employed in the literature with their corresponding patient outcomes.

1. Background and importance

Tumors of the skull are comprised of numerous entities distinguishable primarily by immunohistochemical analysis (IHC) [1–6]. Undifferentiated pleomorphic sarcomas (UPS/PUS), also known as Malignant Fibrous Histiocytoma (MFH), make up 5–10% of skull sarcomas in adults >40 years [7] with high-grade UPS, the most common variant (90.2%) [8].

Primary intracranial undifferentiated sarcomas, from mesenchymal cells of the meninges or leptomeninges, are exceedingly rare, with <36 total reported cases representing 0.1–4.3% of all cases of sarcoma [9,10] and affecting a much younger demographic (mean 38.7 years). Prognosis and survival are typically poor, with many patients dying within 1-year of diagnosis [9].

Diagnostically, on CT or MRI these lesions can resemble high-grade gliomas, demonstrating strong contrast enhancement, central necrosis, and edema [9] with increased uptake at the tumor site [11]. Histologically, highly cellular pleomorphic, round and spindle shaped cells with eosinophilic cytoplasm and a high mitotic rate, growth occurs in a storiform or whorling pattern surrounded by a collagenous matrix with an inflammatory or giant cell infiltrate [7,9,11,12]. However, since other CNS tumor types also exhibit these same histological characteristics [13], IHC identification of vimentin, desmin and a high Ki67/M1B1, along with absence of other markers, is essential for a definitive diagnosis [7,9,12,14].

As curative treatment requires complete surgical excision with negative margins [8] and adjuvant radiation and chemotherapy are reserved for exceptions and metastases [8,9], there are limited options for successful treatment.

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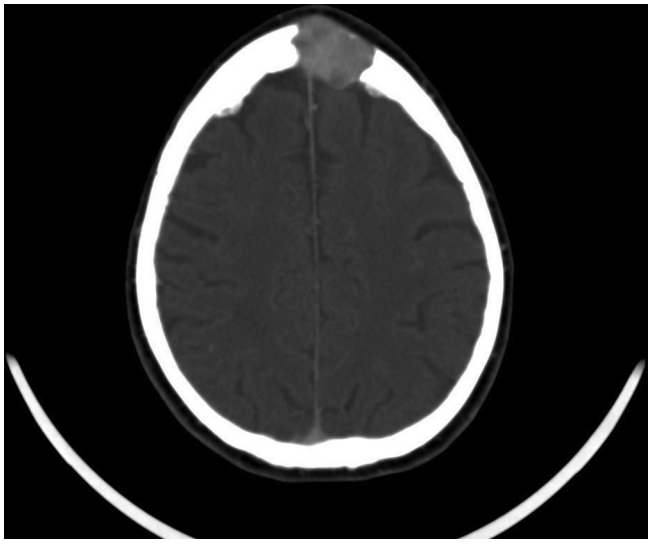

Fig. 1. Pre-operative Imaging. CT with contrast axial view. Note the enhancing lesion eroding through the frontal bone.

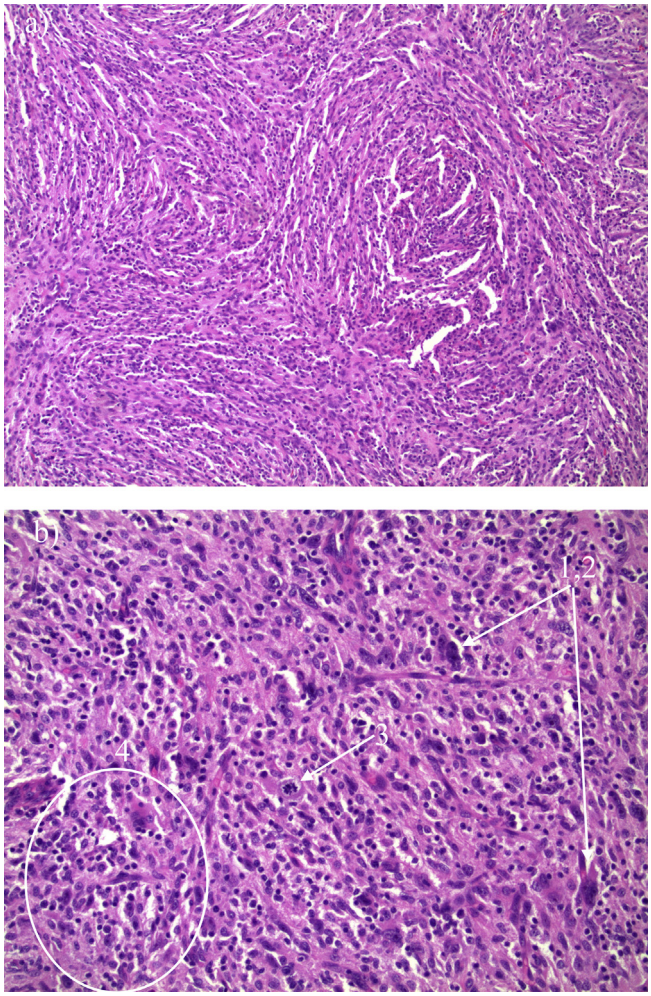

Fig. 2. H&E 40X. a) Spindle cell proliferation with storiform architecture b) Note pleomorphic cells with hyperchromatic nuclei (1) and abundant eosinophilic cytoplasm. Bizarre multinucleated cells (2) are also seen. Many abnormal mitotic figures are present (3). A chronic mononuclear infiltrate example that accompanies the lesion is circumscribed (4).

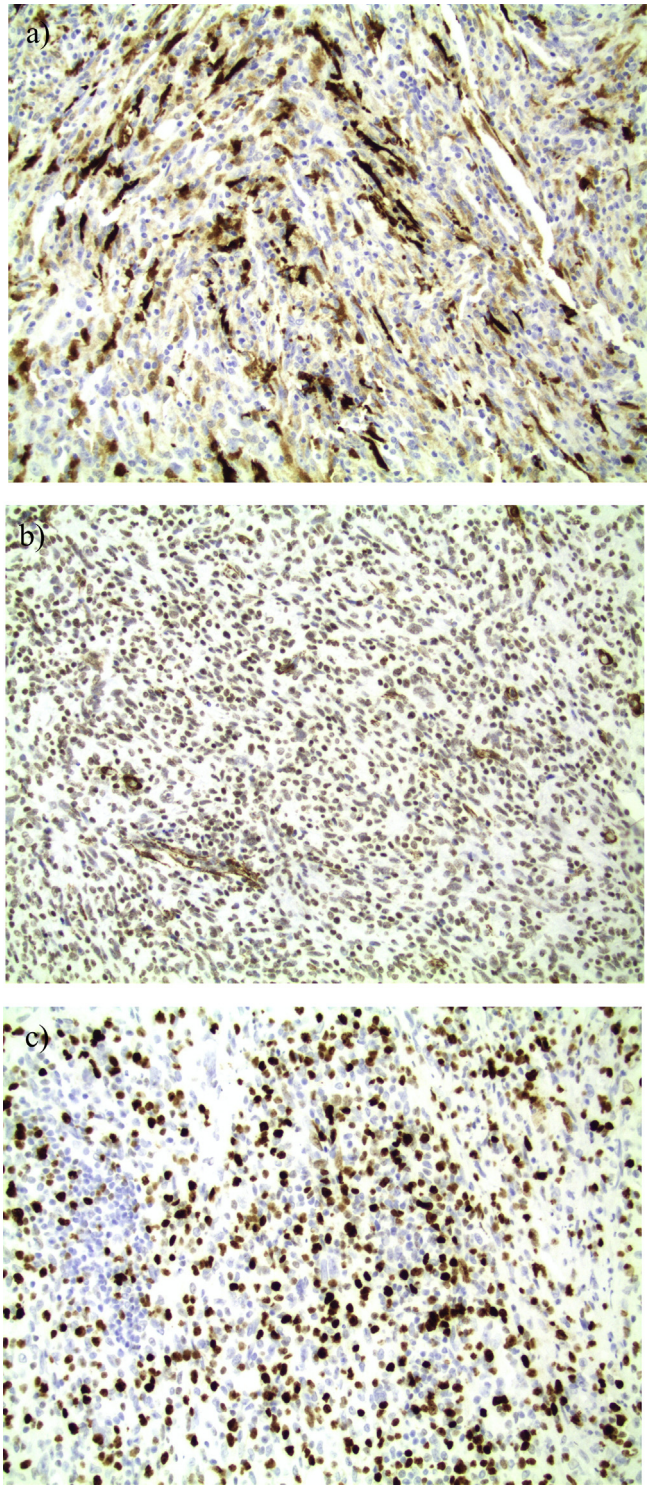

Fig. 3. IHC 40X. a) S100 stain, indicating neuronal differentiation, is focally positive b) SMA, smooth-muscle actin, stains non-specifically c) Ki67 reveals a high proliferation index.

2. Clinical presentation

Following IRB-approval not requiring patient consent, a retrospective analysis of this patient's care was conducted along with a comprehensive review of the literature.

The patient is a 79-year-old male referred to neurosurgery for a large midline scalp mass. Previously, a dermatologist biopsied the

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