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CASE REPORT

Pleomorphic liposarcoma originating from intracranial dura mater

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Abstract Liposarcomas are the most common soft tissue sarcoma found in adults; however, disease involving or spreading to the head and neck is extremely rare. To our knowledge, we present the first case of primary pleomorphic liposarcoma originating intracranially. A 56-year-old man presented with new weakness and imaging findings confirming a right frontal mass. After resection, histological analysis confirmed the diagnosis of pleomorphic liposarcoma. The patient underwent radiation treatment and surveillance imaging, which revealed no other areas of disease. Results reported previously in the literature indicate that pleomorphic liposarcoma is very aggressive in nature. The authors review the few cases of primary or metastatic pleomorphic liposarcoma that have been reported involving the head.

Keywords Pleomorphic liposarcoma · Sarcoma · Dura mater · Neurosurgery

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Introduction

Liposarcomas are the most common soft tissue sarcoma found in adults, most often arising from the lower limbs or retroperitoneum. They can be divided into five histologic subtypes: well-differentiated, dedifferentiated, myxoid, round-cell, and pleomorphic. The pleomorphic subtype accounts for approximately 8% of liposarcomas [1]. To date, there have been very few reports of intracranial involvement of liposarcomas [2–4]. We present a case of a 56-year-old man who presented with a pleomorphic liposarcoma originating intracranially. To our knowledge, this is the first case reported in which a pleomorphic liposarcoma originated from the intracranial dura mater without previous known disease or treatment with radiation or surgery.

Case report

The patient is a 56-year-old man with a history of intermittent weakness and numbness over the course of 3 years. Initially, these symptoms were thought to be secondary to an autoimmune process because the patient's weakness improved dramatically after steroid therapy. The patient underwent a thorough evaluation at an outside hospital including physical examination, computed tomography and magnetic resonance imaging, and analysis of blood and cerebrospinal fluid, with the only remarkable finding being a small 3-mm enhancing lesion over the posterior right frontal convexity that appeared to be dural based. A specific cause for the autoimmune process could not be diagnosed. The lesion was monitored with serial imaging and remained stable over the course of 2 years. Treatment initiated at the outside institution involving a combination

of steroids, mycophenolate, and intravenous immunoglobulin provided symptomatic improvement.

Approximately 2 years after his initial presentation, the patient presented to our institution with a 1-week history of new left-sided weakness. Imaging studies revealed a significant increase in the size of the right-sided mass, which now measured approximately 5.3 × 4.6 × 4.3 cm (Fig. 1). Further workup including positron emission tomography–computed tomography revealed no other lesions.

The patient underwent stereotactic craniotomy for resection of the lesion. Although imaging characteristics had suggested an origin from the dura mater, no definite tail could be identified intraoperatively. Gross total resection of the lesion was obtained (Fig. 2). Histologic evaluation revealed tumor cells with soap-bubble appearance with optically clear cytoplasmic droplets (Fig. 3). The nuclei were distorted and scalloped by the droplets, resembling lipoblasts. Mitotic figures, including atypical forms, were frequent. Immunohistochemical stains showed the tumor cells to be positive for vimentin but negative for glial fibrillary acidic protein, S-100 protein, and epithelial membrane antigen. Diagnosis of high-grade pleomorphic liposarcoma was given. The patient then underwent radiation therapy with a total dose of 59.4 Gy. At his most recent follow-up appointment, approximately 7 months after surgery, there was no sign of recurrent disease.

Discussion

This case represents a primary dural pleomorphic liposarcoma, based on histological features consistent with this diagnosis. The dural origin is supported by both preoperative and postoperative evaluations that revealed no evidence of this lesion originating from another source. While the head and neck literature contains several reports of liposarcomas involving the skull and scalp [5–12], intracranial involvement has been limited primarily to intracranial metastases [13–17], with very few reports of primary intracranial liposarcomas [2–4]. This case represents to our knowledge the first case of a primary intracranial pleomorphic liposarcoma.

Extradural primary liposarcoma of the skull has previously been reported by Srivastava et al. [10] in the frontal

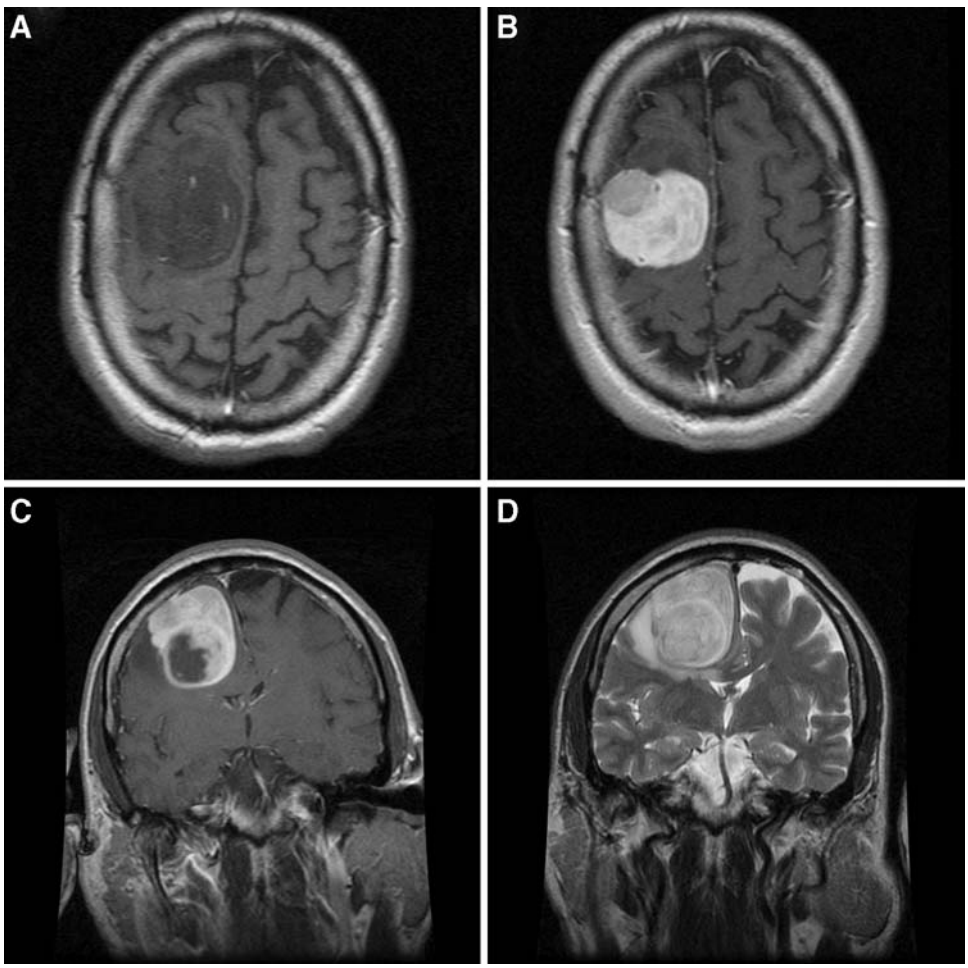

Fig. 1 Axial T1-weighted magnetic resonance (MR) images without (**a**) and with (**b**) gadolinium enhancement, coronal T1-weighted MR image with gadolinium enhancement (**c**), and coronal T2-weighted MR image (**d**) demonstrating a right-sided enhancing mass measuring approximately 5.3 × 4.6 × 4.3 cm, with surrounding edema

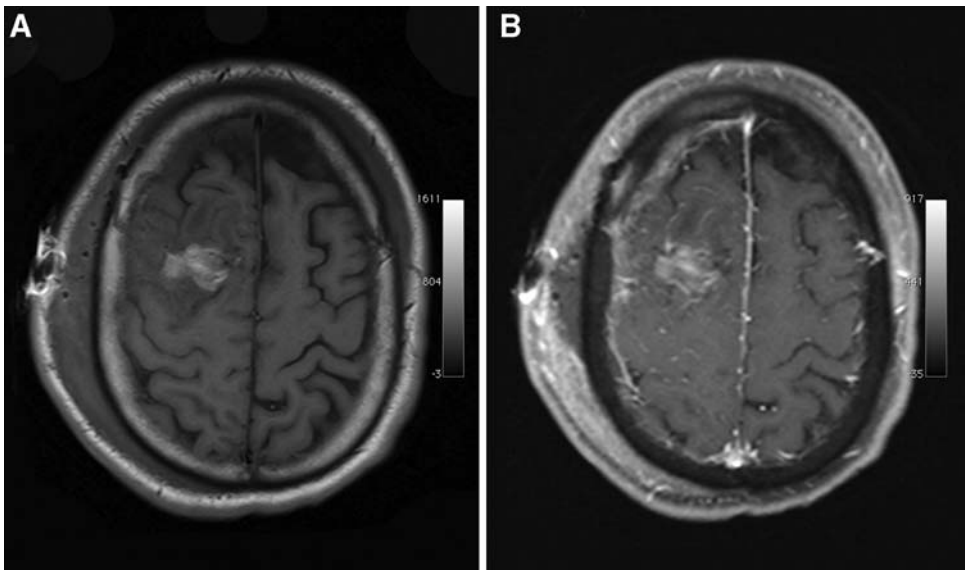

Fig. 2 Postoperative T1-weighted magnetic resonance images obtained before (**a**) and after (**b**) administration of contrast agent

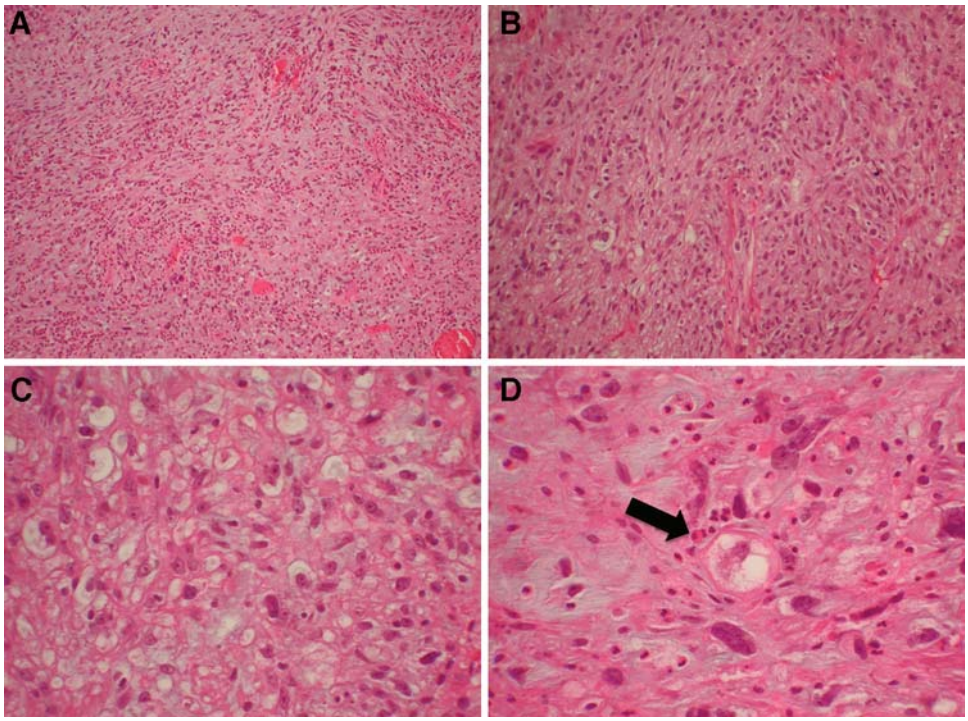

Fig. 3 **a** Hematoxylin and eosin (H&E) section at 4× magnification demonstrating high cellularity. **b** H&E section at 10× magnification showing similar high cellularity. **c** H&E section with 20× magnification showing lipoblasts and diffuse myxoid background. **d** H&E section at 40× magnification indicating lipoblast

bone of a 40-year-old man and by Agarwal et al. [5] in the mastoid of a 4-year-old boy. In 1996, Coatesworth et al. [6] reported a case of liposarcoma in the temporal bone in a patient who had previously received radiation to the parotid gland. In 1994, Stewart et al. [11] added four cases to the 83 cases of head and neck liposarcomas they reviewed in the English literature with only one of those cases originating intracranially. That case was described by O'Malley et al. [12] and involved a 28-year-old man who presented with multiple cranial lesions after radiation for medulloblastoma at 2 years of age. Of note, one of his lesions appeared to be a primary pleomorphic liposarcoma of the scalp but no primary pleomorphic intracranial lesions were noted.

Rates of distant metastasis of sarcomas range from 15 to 35% [18, 19]. Although intracerebral metastases have also been described in multiple reports [13–17], they remain quite rare considering the common hematogenous route of dissemination in sarcomas [16]. Fitzpatrick et al. [14] described the case of a 74-year-old woman who presented

with a hemorrhagic liposarcoma of the brain occurring 2 years after removal of a liposarcoma of the thigh. Kumar and Teasdale [16] described a similar patient in whom intracranial metastatic liposarcoma developed 1 year after surgery for a right lower extremity tumor. Ferguson et al. [13] described a patient in whom cranial meningeal liposarcoma metastasis occurred 23 years after her primary tumor was treated. Utsunomiya et al. [17] presented the case of a 44-year-old patient with a multiply recurrent myxoid liposarcoma with subsequent metastases to the dura and brain.

The first case report of a primary dural liposarcoma was published in 1970 [3]. Kothandaram described a 30-month-old child who was found at autopsy to have a dural liposarcoma. She had initially presented at 4 months of age because of increasing head circumference and had undergone evacuation of a subdural hematoma in the same location. At that time it was stated to be the first known occurrence of a primary intracranial liposarcoma. Histological evaluation did reveal the presence of some pleomorphic cells, but a specific type of liposarcoma was not described at that time. Interestingly, Cinalli et al. [2] also reported sarcomas in the subdural space in two children who had previously undergone subdural hematoma evacuation. One of these children was found to have a liposarcoma, although again the subtype was not described. Sima et al. [4] described a 70-year-old woman with speech and gait disturbance who was found to have a primary liposarcoma closely attached to and infiltrating dura. The pathological analysis differed in that this liposarcoma was considered well-differentiated with round-cell and pleomorphic areas.

The only previously reported case of a primary pleomorphic liposarcoma involving the dura was presented in 2008 and occurred in the spine. Lmejjati et al. [20] described a 35-year-old patient who presented with cauda equina syndrome initially thought to represent recurrent disc herniation or fibrosis. The patient underwent imaging demonstrating involvement of the L4 and L5 vertebral bodies with intradural involvement of the nerve roots and cauda equina. Resection yielded histopathological findings of pleomorphic liposarcoma. Unfortunately, even with aggressive resection and radiotherapy, the patient died within 3 months.

Once diagnosis is made histologically, clinical features, treatment, and prognosis can be further established. Misdiagnosis can lead to inadequate or delayed treatment. In their series of 30 patients with head and neck liposarcomas, Davis et al. [7] found that one third were initially misdiagnosed. The available literature regarding treatment of liposarcomas primarily focuses on truncal and extremity disease. Aggressive resection continues to be the mainstay treatment with differing forms and protocols involving radiation and chemotherapy available. Success of treatment is heavily dependent on site of presentation and type of liposarcoma [1]. Enzinger and Weiss [21] previously reported the 5-year survival rates for the histological subtypes occurring throughout the body. The pleomorphic type has a 5-year survival rate of approximately 20%, whereas the 5-year survival rate of the well-differentiated type is around 90%. These findings are further supported by the findings of Davis et al. [7], who reported that disease-specific survival in patients with head or neck sarcoma was 100% for well-differentiated types but was just 45% in the pleomorphic type. This series also supports the need for aggressive treatment, especially considering the high rate of local recurrence with pleomorphic liposarcoma in the head and neck [7]. Interestingly, size of tumor did not affect prognosis in a study of 76 patients with head and neck liposarcoma by Golledge et al. [8], but again the principal determinant of outcome was histological grade. Because obtaining adequate margins in any tumor in the head and neck region can be unrealistic given the adjacent structures, the use of adjuvant therapies such as radiation and chemotherapy in these patients is important.

Conclusion

This patient presented with the first reported primary pleomorphic liposarcoma originating intracranially. Given the rarity of presentation, the clinical course is difficult to predict. Unfortunately, not only was the primary tumor site unfavorable for aggressive surgical margins, but, as the literature has shown, the pleomorphic type is associated with an aggressive course and high rate of recurrence. In addition to having undergone gross total resection and radiation therapy, our patient will continue to be monitored closely with routine imaging and clinical evaluation.

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