3. Discussion

To our knowledge, this is the first case of PML developing 40 years after chemoradiation therapy for HL despite appropriate monitoring. Current National Comprehensive Cancer Network (NCCN) guidelines recommend annual complete blood counts for HL patients in remission [5]. While our patient's total white blood cell and lymphocyte counts were normal, CD4+ counts were not checked until he presented to our clinical attention. Although late onset PML can occur in the setting of relapsed HL, our patient had no signs of recurrent malignancy or other opportunistic disease. We hypothesize that the combination of CD4+ lymphopenia with underlying B-cell dysregulation from prior HL increased his risk of PML, but it remains unclear why he did not develop other opportunistic infections.

Treatment of PML involves addressing the underlying mechanism for impaired cellular immunity. Because therapeutic options are limited in cases of CD4+ lymphophenia, clinical trials are currently underway.

4. Conclusion

Impaired cellular immunity can persist for decades following diagnosis and treatment of HL. However, current NCCN guidelines for disease surveillance may overlook CD4+ lymphopenia in HL survivors. Larger cohort studies are needed to determine which patients remain at risk for opportunistic CNS infections despite current standards of care, which may allow for new surveillance and treatment guidelines.

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Multiple recurrences require long-term follow-up in patients diagnosed with spindle cell oncocytoma of the sella turcica

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ABSTRACT

Spindle cell oncocytoma (SCO) of the sella turcica is characterized as a WHO grade I non-endocrine neoplasm of the sella turcica. Histologically, these tumors contain spindled and variably oncocytic cellular processes. Although SCOs lack immunoreactivity for neuroendocrine markers and pituitary hormones, they are clinically indistinguishable from non-functioning pituitary adenomas. In contrast to the initially described benign clinical course, several reports have subsequently illustrated cases with multiple recurrences with or without histological features of anaplasia in the form of nuclear pleomorphism, frequent mitoses, high Ki-67 index, and/or necrosis. With a follow-up of 14 years, we report a case of SCO with multiple recurrences along with an exhaustive clinico-pathological review of all 41 cases of SCO reported in the literature, of which recurrence has been described in 11 cases. Collectively, this report highlights the importance of long-term follow-up and the possible need for adjuvant radiotherapy in patients diagnosed with a sellar SCO and provides a comprehensive review of this rare nonadenomatous sellar tumor.

1. Introduction

Tumors of the sella turcica are primarily composed of pituitary adenomas. However, less than 10% of these tumors are nonadenomatous, which comprise several other histological diagnoses [1]. Since the initial description in 2002 [2] and subsequent classification in the 2007 WHO handbook [1], spindle cell oncocytoma (SCO) of the sella turcica has been characterized as WHO grade I non-endocrine neoplasm of the sella turcica. Histologically, these tumors contain spindled and oncocytic cellular processes, which mimic meningiomas. Although SCOs lack immunoreactivity for neuroendocrine markers and pituitary hormones, they are clinically

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indistinguishable from non-functioning pituitary adenomas. In contrast to the initially described benign clinical course, several reports have subsequently illustrated cases with multiple recurrences with or without histological features of anaplasia in the form of nuclear pleomorphism, frequent mitoses, high Ki-67 index, and/or necrosis [3–11]. We report a case of SCO with multiple recurrences along with an exhaustive clinico-pathological review of all 41 cases of SCO reported in the literature, of which recurrence has been described in 11 cases. Collectively, this report highlights the importance of long-term follow-up and the possible need for adjuvant therapy in patients diagnosed with sellar SCO and provides a comprehensive review of this rare nonadenomatous sellar tumor.

2. Clinical summary

We present the case of a 47-year-old male who was initially diagnosed with a right suprasellar meningioma following a subtotal resection in 2002 (Fig. 1a–c). Tumor recurrence resulted in a biopsy followed by radiotherapy for the residual tumor in 2004 (Fig. 1d–g). The patient was clinically stable until 2012 when he noticed his vision to worsen but with no consequences to his daily activities. In 2015, at the age 60, he presented with complete temporal loss in both visual field quadrants of his left eye, secondary to tumor compression of the optic chiasm. Cranial nerve testing showed a sluggish pupillary reflex in the left eye with consensual response. No other cranial nerve abnormalities were observed at this time. Fundoscopy showed normal optic discs. Symptoms of hypothyroidism, decreased libido, or erectile dysfunction were not present.

Given the visual changes in his left eye, an endoscopic transnasal transsphenoidal resection was performed for the large recurrent/residual anterior skull base and suprasellar tumor. Pre-operative magnetic resonance imaging (MRI) demonstrated a 2.6 × 3.0 × 2.4 cm lesion in the suprasellar region, which was relatively isointense to gray matter and heterogeneously enhancing on both T1 and T2-weighted images (Fig. 3). The tumor was debulked anterior to posterior. Laterally, the tumor was firmly attached to the right and left carotid arteries, making it impossible to retract on the tumor without risking damage to the carotid arteries. The patient tolerated the procedure well with no major complications. His post-operative course was complicated by meningitis and increased intracranial pressure requiring a lumbar drain. Post-operative MRI demonstrated residual tumor within the sella associated with suprasellar extension measuring 2.9 × 2.5 × 1.9 cm (Fig. 2). At last follow-up in March 2016, the patient had no new symptoms with a stable residual tumor.

3. Pathological findings

The microscopic findings from the initial 2002 resection (Fig. 3a) identified no evidence of atypical or anaplastic features with a histopathology in keeping with a meningothelial meningioma. Similarly, material obtained from the 2004 biopsy (Fig. 3b) maintained a tumor with a spindly architecture. The meningothelial cells were monotonous and bland. There were no areas of tumor necrosis or increased mitotic activity and calcification or psammoma bodies were not identified.

Given that the tumor entity of spindle cell oncocytoma of the sellar region was not characterized or described in 2002 and 2004 and was not codified in the WHO lexicon until 2007, material obtained from the most 2015 (Fig. 3c, d) resection was evaluated in conjunction with re-evaluation of the previous surgical specimens from 2002 to 2004. Sections from the 2015 resection showed a cellular neoplasm, largely composed of spindled cells that were more polygonal in some areas. The tumor largely showed an interwoven fascicular-type architecture. There was mild to moderate nuclear pleomorphism with some atypia. The mitotic activity was not increased. These features were similarly observed in the previous two specimens.

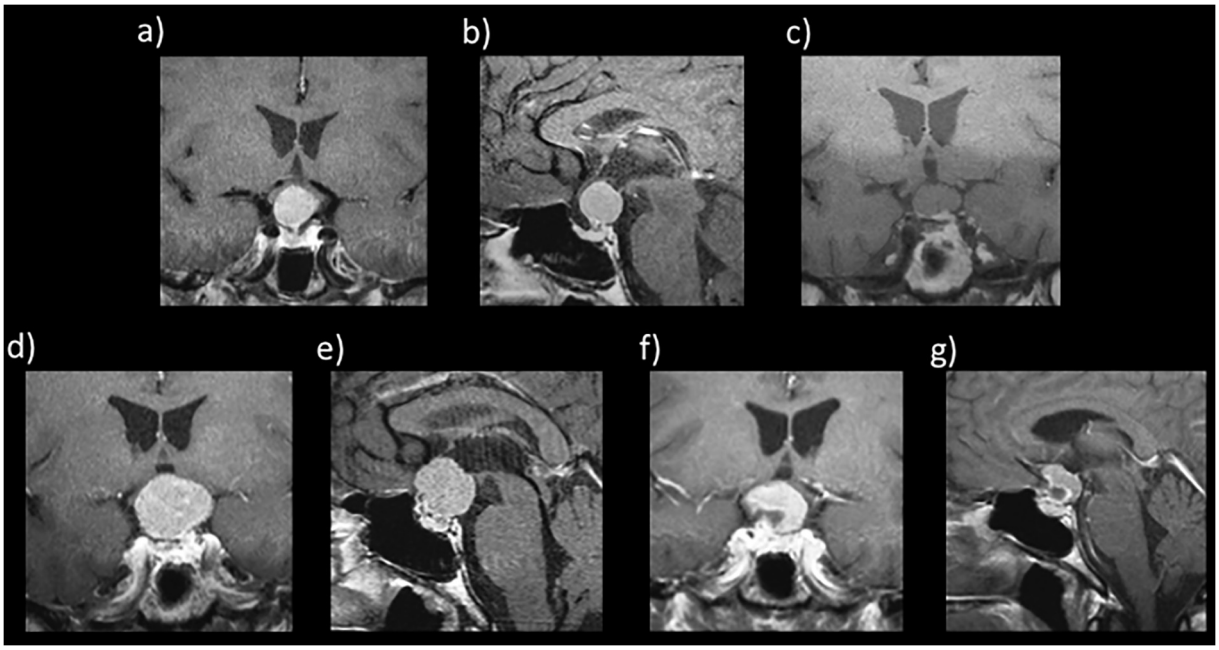

Fig. 1. Pre- and post-operative MRI of primary tumor and first recurrence. Primary tumor pre-operative T1W (a) coronal and (b) sagittal MRI with gadolinium of suprasellar mass (1.73 × 1.49 × 1.65 cm) displacing the optic chiasm with uniform moderate enhancement. (c) Post-operative T1W coronal MRI with gadolinium demonstrating residual suprasellar mass with resection of anterior inferior corner. Continued compression of optic chiasm with no extension into sella. Pre-operative T1W (d) coronal and (e) sagittal MRI with gadolinium showing first tumor recurrence (2.5 × 1.7 × 2.5 cm), homogeneous enhancement, and marked compression of optic chiasm. Post-operative T1W (f) coronal and (g) sagittal MRI with gadolinium showing residual tumor (2.3 × 1.7 × 2.3 cm) and heterogeneous enhancement containing a cystic component.