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CASE REPORT

Atypical Teratoid / Rhabdoid Tumour in the Sella Turcica of a Female Adult

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ABSTRACT

Atypical teratoid / rhabdoid tumour is a rare and highly malignant cancer that mainly occurs in children younger than the age of 5 years. Only four cases of atypical teratoid / rhabdoid tumour in the sella turcica of an adult have been described in the English language literature. This report is of an atypical teratoid / rhabdoid tumour occurring in the sella turcica of a female adult.

Key Words: *Magnetic resonance imaging; Teratoid tumor, atypical*

中文摘要

一名成年女性蝶鞍內的非典型畸胎瘤樣／橫紋肌樣瘤

鄒起華、羅尚銘、王馨藝、劉百倫、陳智明、鄧國穎、張毓靈

非典型畸胎瘤樣／橫紋肌樣瘤是一種罕見的高度惡性腫瘤，主要好發於5歲以下兒童。英文文獻中只報導過4名蝶鞍內非典型畸胎瘤樣／橫紋肌樣瘤的成年病例。本文報告一名成年女性蝶鞍內的非典型畸胎瘤樣／橫紋肌樣瘤。

INTRODUCTION

Atypical teratoid / rhabdoid tumour was first described by Beckwith and Palmer in 1978.[1] Atypical teratoid / rhabdoid tumour is most commonly diagnosed in the posterior fossa of children younger than the age of 5 years, but it may occur rarely in any site of the central nervous system in adults.[2] Histopathologically, atypical teratoid / rhabdoid tumour is defined by the presence of rhabdoid cells and populations of primitive neuroectodermal, epithelial, and mesenchymal cells.[3] Diagnosis can be confidently made by demonstrating lack of nuclear integrase interactor 1 (INI1) protein expression by immunohistochemical methods.[4]

To the authors' knowledge, only four cases of atypical teratoid / rhabdoid tumour have been reported in the English language literature.[5] This report is of a 43-year-old woman with atypical teratoid / rhabdoid tumour in the sella turcica. The magnetic resonance imaging (MRI) and histopathological findings of this patient are described.

CASE REPORT

A 43-year-old woman presented in 2011 with headache

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and double vision for 10 days. Computed tomography (CT) showed a hyperdense tumour at the sellar region, extending to the suprasellar region and compressing the optic chiasma. There was no calcification noted in the tumour. MRI was immediately arranged, which showed a mass hyperintense on T2-weighted imaging (Figure 1) and isointense-to-hypointense on T1-weighted imaging (Figure 2) at the sellar region. The mass extended to the suprasellar region. On post-contrast MRI, the mass showed strong contrast enhancement (Figure 3). Invasion to the left cavernous sinus was also noted. As pituitary adenoma is the most common tumour located at the sellar region, the preoperative diagnosis was pituitary adenoma.

The intrasellar tumour component was removed via the trans-sphenoidal route. Histopathological study revealed a dull dark brown–coloured tissue macroscopically and tumour cells showing frequent mitotic activity (Figure 4) and pleomorphic nuclei with rhabdoid-like pinkish globular cytoplasmic inclusions microscopically (Figure 5). Mitotic figures and apoptotic bodies were readily identified. On immunohistochemical staining, these tumour cells were positive for cytokeratin, epithelial membrane antigen, and SALL4. The INI1 nuclear signal was lost in the tumour cells with positive internal control (Figure 6). Staining for adrenocorticotropic hormone, prolactin, and growth hormone were all negative. The diagnosis of atypical teratoid / rhabdoid tumour was made based on morphology and immunohistochemical staining features. The patient was treated with post-surgical radical radiotherapy. However, after 2 weeks, the patient developed paraplegia, and MRI of the spine showed multiple tumour disseminations to the spinal cord.

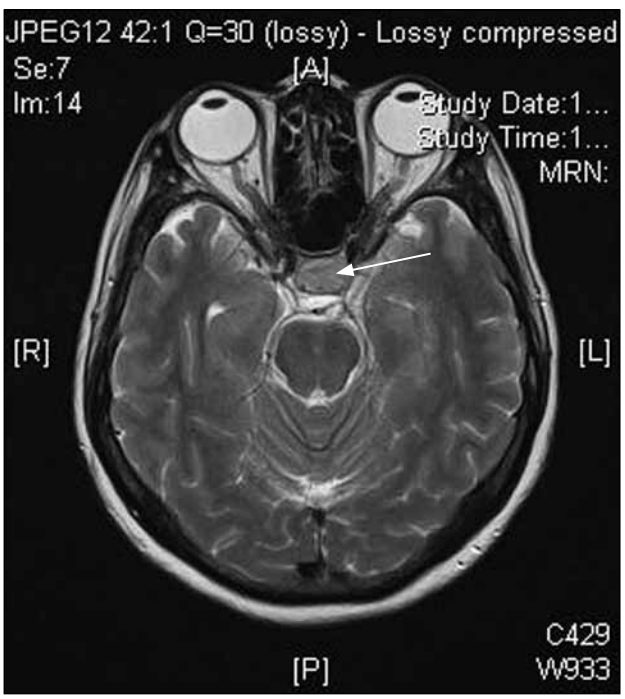

Figure 1. Hyperintense lesion at the sellar region (arrow) on T2-weighted magnetic resonance imaging.

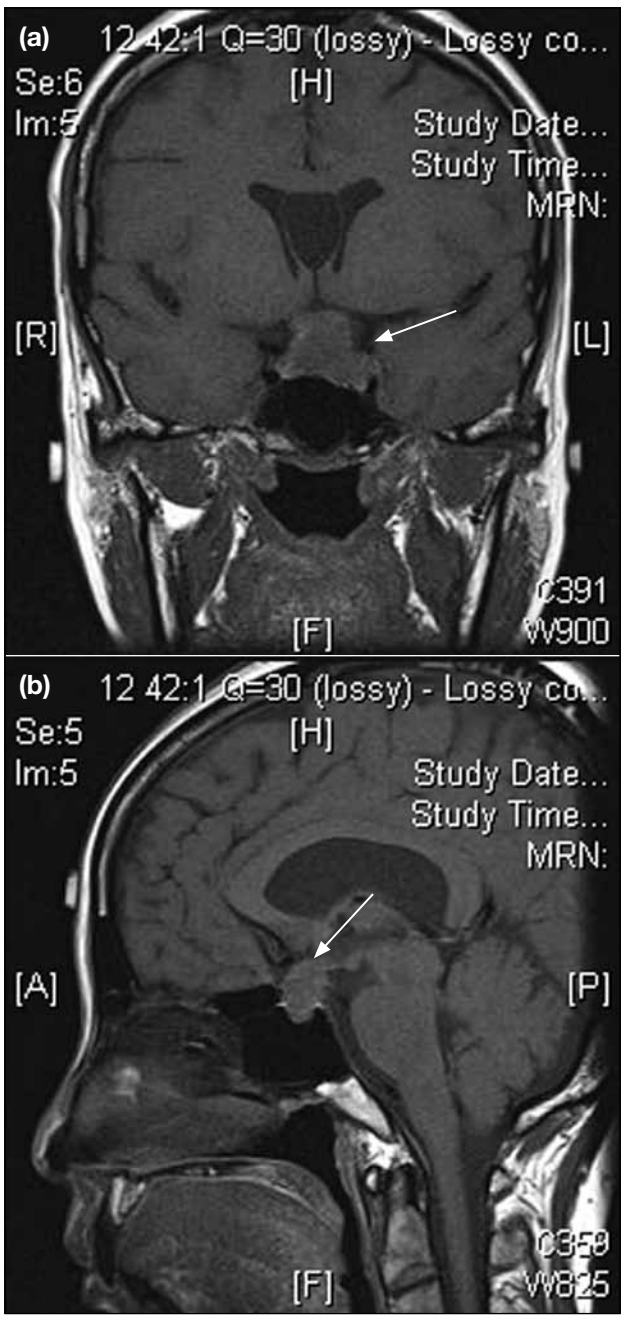

Figure 2. Isointense-to-hypointense lesion at the sellar region extending to the suprasellar region and compressing the optic chiasma (arrows) on T1-weighted magnetic resonance imaging: (a) coronal view and (b) sagittal view.

DISCUSSION

Atypical teratoid / rhabdoid tumour is an aggressive cancer of infancy and childhood; 94% of reported patients are aged 5 years or younger.[6] In 1992, Horn et al[7] were the first authors to recognise atypical teratoid / rhabdoid tumour in an adult. A search of the English language literature shows that only 28 cases were reported in adults between 1992 and 2009.[8] Occurrence of atypical teratoid / rhabdoid tumour in the sellar turcica is rare, with only four cases reported in adults to date.[5,8,9]

Imaging findings are usually non-specific at the time of diagnosis. On CT, the tumour, probably due to a high cellular content, appears hyperdense with a cystic / necrotic component.[10] Calcifications have been reported in some patients. On MRI, the solid component of this tumour shows similar signal intensity characteristics as grey matter on both T1- and T2-weighted images. Multiple cystic or necrotic areas are common. The solid portion of the tumour shows restricted diffusion.

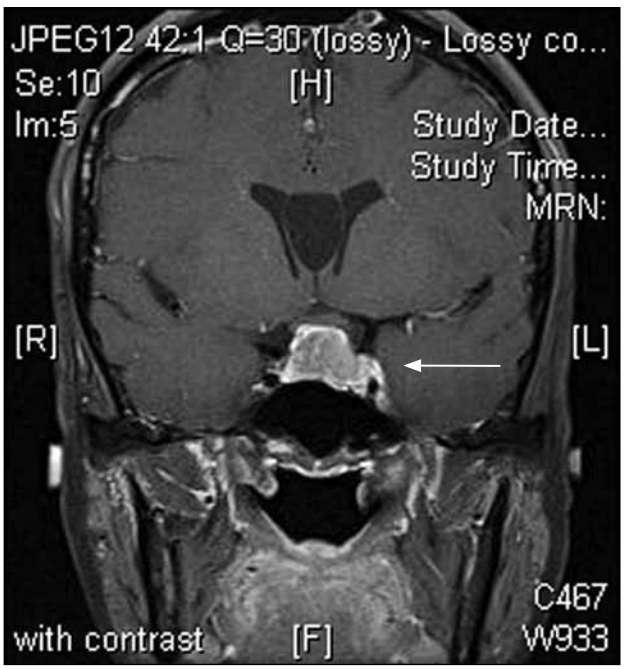

Figure 3. Contrast-enhancing lesion with invasion to the left cavernous sinus (arrow) on magnetic resonance imaging.

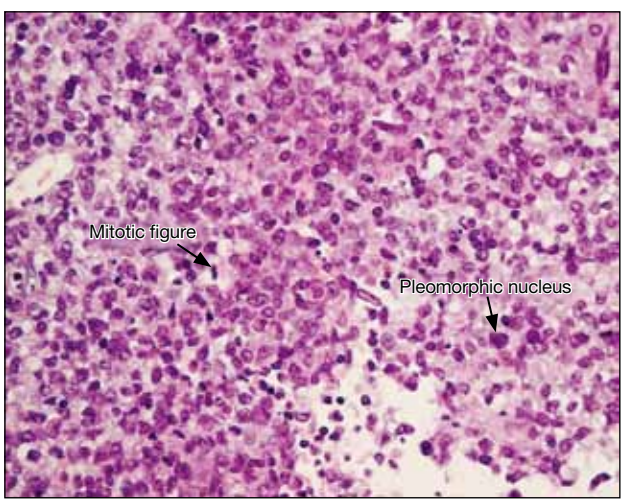

Figure 4. Histopathology shows syncytial sheets of tumour cells with frequent mitotic activity and pleomorphic nuclei (arrows) [H&E; original magnification x 200].

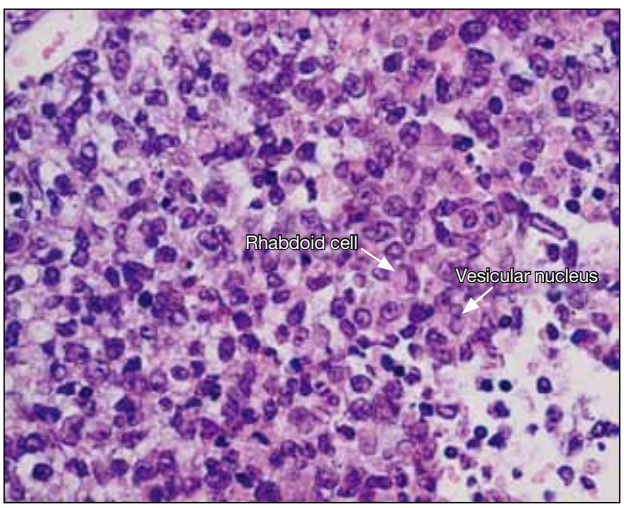

Figure 5. Histopathology shows high-powered view of tumour cells with vesicular nuclei and occasional tumour cells with rhabdoid features (arrows) [H&E; original magnification x 400].

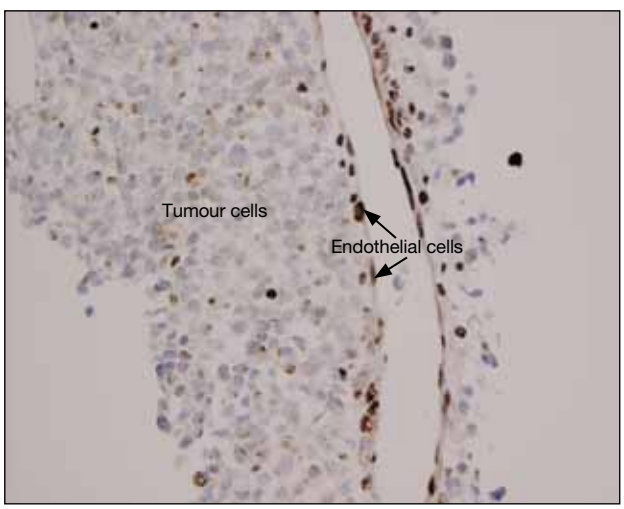

Figure 6. Immunohistochemical staining for nuclear integrase interactor 1 (INI1) shows the tumour cells with loss of nuclei signals with positive internal control in the endothelial cells and inflammatory cells (arrows) [INI1 (clone 25/BAF47, dilution 1:80); original magnification x 200].

At the microscopic level, the rhabdoid cellular component is outstanding, although the coexistence of epithelial-like and mesenchymal areas, as well as the presence of pseudorosettes, could lead to a misdiagnosis. Clinical presentation and imaging studies do not differentiate atypical teratoid / rhabdoid tumour from similar tumours such as rhabdomyosarcomas, primitive neuroectodermal tumours, malignant teratomas, and carcinosarcomas.

Immunohistochemistry is of vital importance for the diagnosis of atypical teratoid / rhabdoid tumour. Loss of nuclear INI1 protein expression by immunohistochemical methods has been proven to be the gold standard in the diagnosis of this tumour.

Survival in adults, based on the literature reports, averages 38.7 months.[11] This survival period is longer than that for children. Recent advances using aggressive therapy might have contributed to better outcomes for adults than for children.

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