Suprasellar Hemangioblastoma During Pregnancy: Case Report

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Abstract

Background
Hemangioblastoma is a benign tumor that typically occurs in the posterior fossa, but may be found in anywhere along the neuraxis. Its growth behavior may be difficult to predict and pregnancy has been discussed as a possible growth promoting factor.
Case Description
We present the case of a rare suprasellar hemangioblastoma diagnosed during pregnancy. Following the progressive worsening of symptoms, the baby was delivered by caesarean section. The patient was treated with stereotactic radiotherapy, resulting in a marked improvement in symptoms and impressive reduction in tumor size.
Conclusions
High vascularity and solid growth with a tendency for surgical complications appear typical of suprasellar hemangioblastoma, as indicated by this and several other reported cases. The importance of non-surgical treatment strategies, including radiation therapy, is emphasized by this case. Pregnancy is one of the factors that has been investigated as potentially growth promoting. We speculate that post-partum hormonal changes may have contributed to tumour shrinkage in our case.

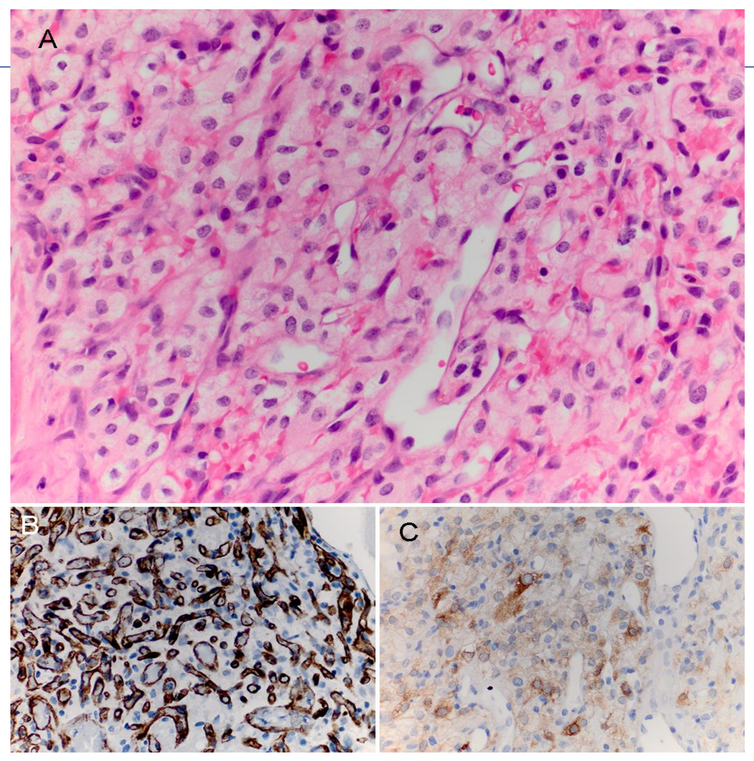
Background

Hemangioblastoma is a benign tumor that typically occurs in the posterior fossa, but may be found in anywhere along the neuraxis, including the parasellar and suprasellar region. Suprasellar hemangioblastoma is a rare entity that represents a unique management challenge. Pregnancy is a possible promoter of hemangioblastoma growth and has been investigated in recent years. We present the case of a 30-year-old pregnant woman with a hemangioblastoma of the suprasellar region managed with stereotactic radiation therapy following delivery.

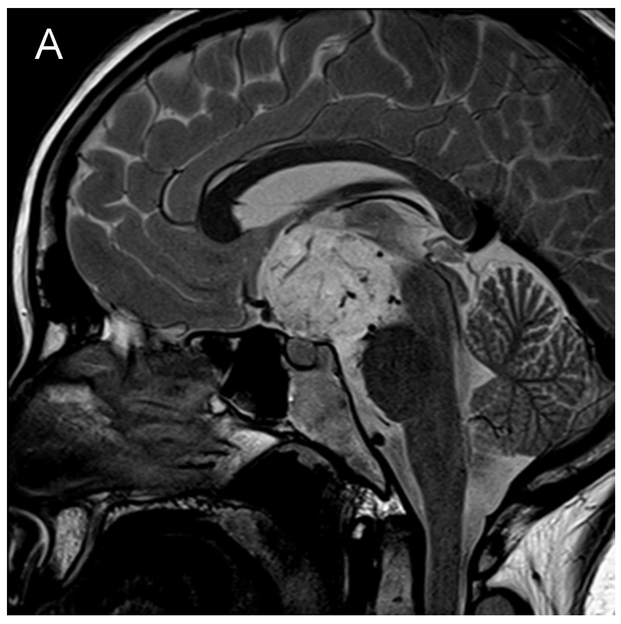
Case Description

A 30-year-old female presented at 35-weeks gestation with a one-month history of gradually progressive bitemporal hemianopsia, headache, impulsiveness, and short-term memory decline. Magnetic resonance imaging (MRI) showed a solid, 3.3 x 3.0 x 3.5 cm lesion in the suprasellar region with post-gadolinium T1 weighted imaging demonstrating avid enhancement of the mass.

The patient was admitted to hospital for investigation, and within days, her vision deteriorated to only light perception in the right eye and counting fingers in the left. Because of her rapidly progressive symptoms, the baby was delivered by Caesarean prior to neurosurgical intervention. The patient underwent bifrontal craniotomy and bilateral orbital osteotomy for attempted resection of the lesion. Because of excessive bleeding from the tumor, resection was abandoned and the procedure was limited to a biopsy, which demonstrated hemangioblastoma. The patient was subsequently treated with stereotactic radiotherapy (SRT) for residual tumor to a total dose of 5400 cGy, given in 1.8 Gy fractions over six weeks.



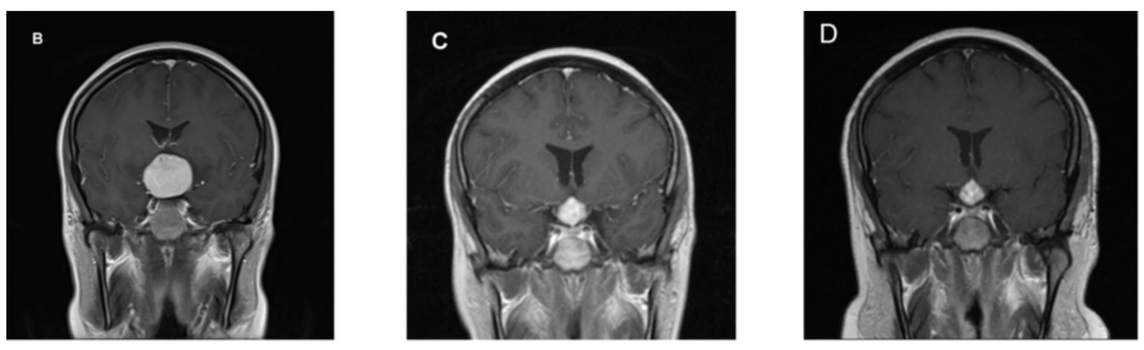
Biopsy of tumor demonstrating hemangioblastoma.
A) Stromal cells with vacuolated cytoplasm and dense capillary network (H&E, 40x). B) CD34 immunohistochemistry highlights endodethlial cells and capillary network (CD34 immunohistochemistry with hematoxylin counterstain, 10x). C) Stromal cells are immunohistochemically positive for inhibin (Inhibin immunohistochemistry with hematoxylin counterstain, 40x).



A) Sagittal T2 weighted image shows a solid, 3.3 x 3.0 x 3.5 cm lesion in the suprasellar region with multiple flow voids.

Outcome

Six months after her initial presentation, and three months following the completion of SRT, the patient's vision had returned to normal. At presentation, mini-mental status examination revealed significant deficits in memory and concentration; however, at five months post-SRT, detailed neuropsychological assessments revealed only very subtle deficits, suggesting considerable improvements in cognition. Five years after the conclusion of her course of radiotherapy, the patient's scan showed a residual suprasellar lesion measured at 1.6 x 1.5 x 1.4 cm.



B) Post-gadolinium coronal T1 weighted image demonstrates avid enhancement of the mass. C) Post-treatment (3 months) post-gadolinium coronal T1 image shows significantly reduced tumor size to 2.1 x 1.9 x 2.1 cm. D) Post-gadolinium coronal T1 weighted image 5 years after treatment shows a residual tumor measuring 1.6 x 1.5 x 1.4 cm.

Discussion

Suprasellar hemangioblastomas are exceedingly rare. No reported cases arose in pregnancy. Most demonstrate solid, often large, highly vascularized tumors that show prominent flow voids on MRI. Hemorrhage can complicate resection, limiting surgical intervention. Because of the limited data available in the literature, it is difficult to ascertain the rate of success following radiotherapy (RT) for hemangioblastomas in the suprasellar region. Three cases identified in the literature describe suprasellar hemangioblastomas in which total resection was not performed, and RT resulted in good outcomes for all three cases. In addition to the present case, these emphasize the utility of RT in the management of suprasellar hemangioblastomas.

Conclusions

High vascularity and solid growth with a tendency for surgical complications appear typical of suprasellar hemangioblastoma, as indicated by this and several other reported cases. The importance of non-surgical treatment strategies, including radiation therapy, is emphasized by this case. Pregnancy is one of the factors that has been investigated as potentially growth promoting. We speculate that post-partum hormonal changes may have contributed to tumour shrinkage in our case.

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