

Primary Intracranial Hemangiopericytoma Presenting as Hemiparkinsonism

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Structural causes of hemiparkinsonism are rare. Intracranial hemangiopericytoma is a rare tumour, which is highly vascular, with high malignant potential, and on imaging can appear similar to a meningioma.¹ We report a patient who had an intracranial hemangiopericytoma presenting with hemiparkinsonism, which improved after surgical removal of the tumour.

CASE REPORT

A 49-year-old right handed man presented to the movement disorders clinic for assessment of his right sided tremor. He had a five month history of rest tremor in his right hand, micrographia and intermittent rest tremor in his right leg. He did not have any involvement of his left side. His gait was unsteady but he was not dragging his feet. He had a one month history of progressive right sided facial droop. He denied any focal weakness in his arms or legs, and no dysphagia and no changes in his sensation. His past medical history included strabismus with decreased vision in his left eye since birth. Dexamethasone, phenytoin and ranitidine were started after his imaging findings.

On examination, he had mild dysarthria and a slight dysconjugate gaze, likely long standing, and otherwise full extraocular movements. He had a right facial droop sparing the frontalis muscle. He had rigidity and rest tremor of his right arm, and an intermittent rest tremor in his right leg. He had a postural tremor in his right arm and slight kinetic tremor in both arms. He had bradykinesia in his right hand and mild hesitancy in his foot taps on the right. His writing was tremulous and micrographic. His strength, reflexes and sensory exam were normal with flexor plantar responses. His gait was unsteady but not shuffling, and he had a mildly stooped posture.

Prior to being seen in clinic, he had a magnetic resonance image of the brain showing a 5 cm enhancing mass from the left sphenoid wing extending into the left temporal region where it compressed the midbrain (Figure 1). There was midline shift to the right, prominent draining veins in the left frontal regions and enlargement of the left middle meningeal artery. Initially these findings were suggestive of a meningioma. He was seen by neurosurgery and due to the vascularity of this lesion, he underwent pre-operative embolization of the left middle meningeal artery. He had a craniotomy for resection of the lesion. See pre-operative video on-line at www.cjns.org.

Histological examination revealed a vascular, highly cellular tumour (Figure 2). The cells had elongated nuclei and were disposed randomly and in fascicular and storiform patterns. The tumour had branching, dilated, thin-walled vascular profiles with a "staghorn" appearance. The tumour cells showed cytoplasmic staining for vimentin and CD34. They were negative for

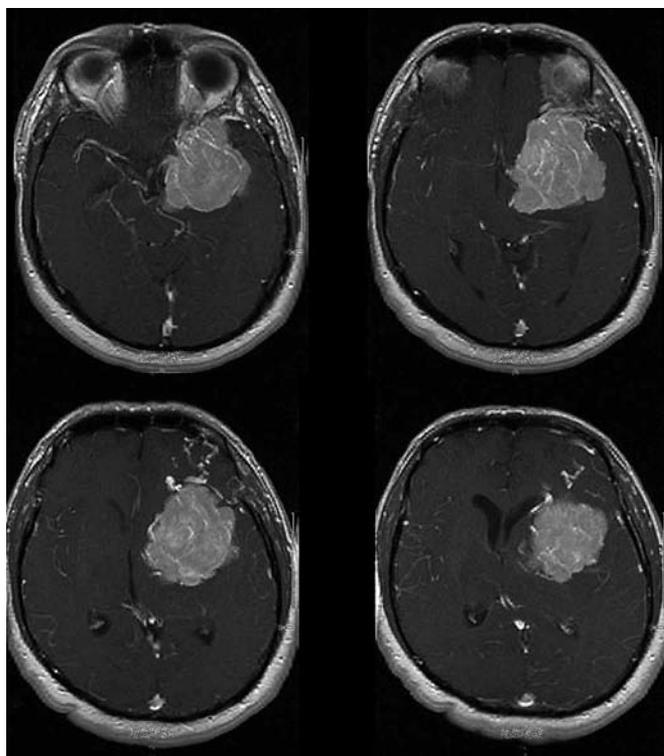


Figure 1: Magnetic resonance imaging of the brain (T1 weighted with gadolinium) showing enhancing extra-axial mass arising from the left sphenoid wing extending to left temporal region and compressing left midbrain.

epithelial membrane antigen. Reticulin staining revealed a delicate pericellular reticulin pattern. The MIB1 proliferation index was variable but reached 20% focally. CD34 immunostaining pattern was patchy in distribution as opposed to being diffuse throughout the tumour. Combined with the high cellularity and staghorn-type vascular pattern, we favoured a diagnosis of hemangiopericytoma.

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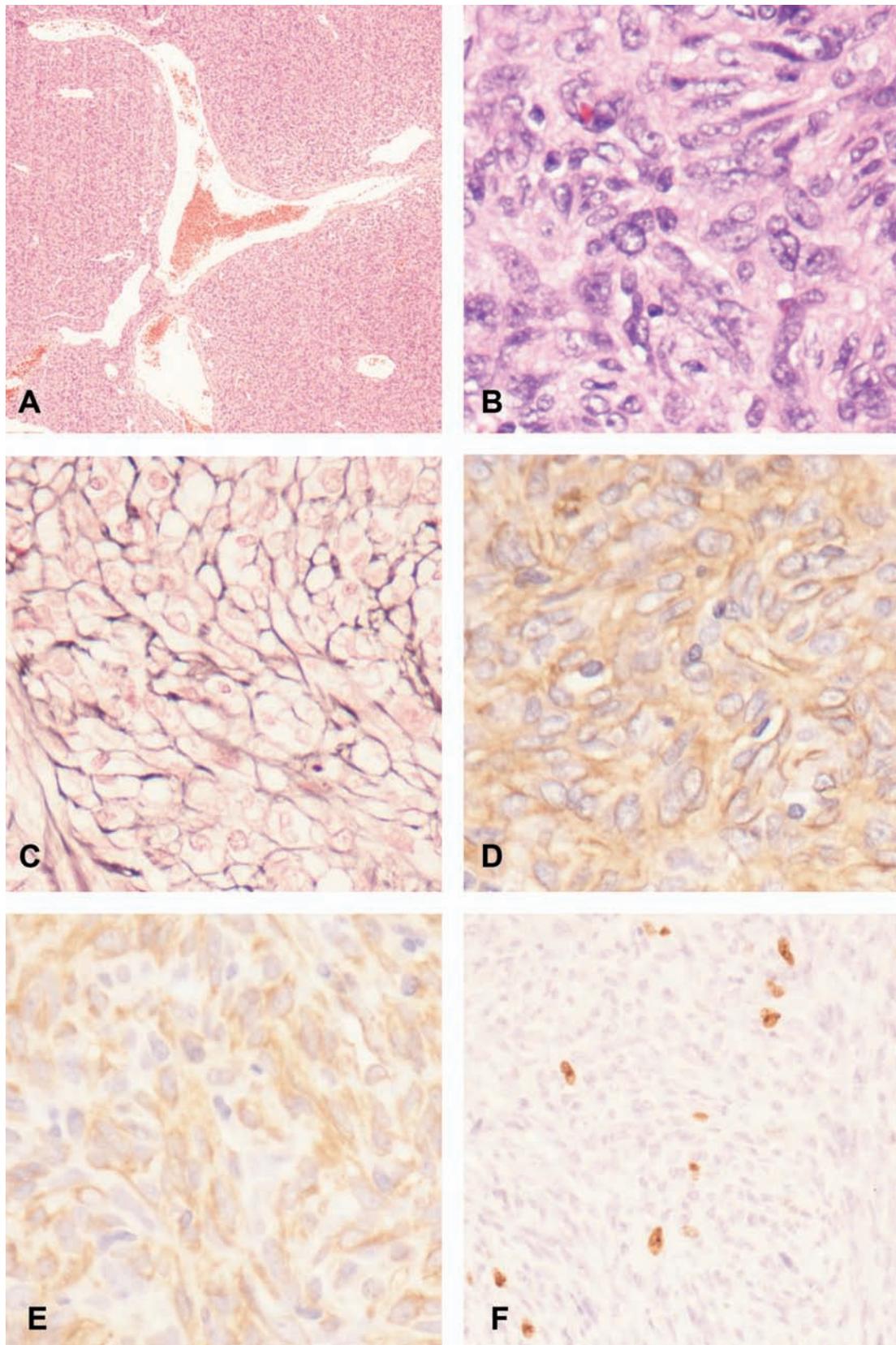


Figure 2: Hemangiopericytoma. A: "Staghorn" vascular profiles (H&E; X100). B: Spindle-shaped nuclei in storiform patterns (H&E; X400). C: Delicate pericellular reticulin network (reticulin stain; X400). D, E: Cytoplasmic immunostaining for CD34 (D), vimentin (E; both X400). F: Moderate MIB1 proliferation index (X400).

Post-operatively, he developed a transient left third nerve palsy, mild right finger extensor weakness with complete resolution of his rest tremor and bradykinesia. Due to the malignant nature of this tumour, the patient received adjuvant radiation therapy. At 1.5 years of follow-up, he has not had any recurrence of his tumour nor return of his bradykinesia but did have an intermittent postural and kinetic tremor. See post-operative video on-line at www.cjns.org.

DISCUSSION

Intracranial hemangiopericytomas account for less than 1% of brain tumours.¹ It has been postulated that they are derived from meningeal pericytes and can metastasize. There is a high rate of misdiagnosis based on clinical and radiological studies, as they mimic meningiomas.¹ Achieving a definitive diagnosis is essential because hemangiopericytomas are more aggressive and often require adjuvant radiotherapy.¹

Hemiparkinsonism as a result of a brain tumour is rare. In a study of 907 patients with supratentorial tumours sparing the basal ganglia, only 0.3% presented with parkinsonism.² Post-operatively, many had improvement of their parkinsonism, suggesting that the parkinsonism was not merely an incidental occurrence, but directly related to the presence of the tumour.

The pathophysiological substrate of hemiparkinsonism resulting from structural lesions is thought to be due to dysfunction of the nigrostriatal circuitry, likely caused by mass effect, impaired perfusion, and alterations in dopamine metabolism.²⁻⁴ Many of the brain tumours that cause parkinsonism are frontal or temporal in location, resulting in posterior and medial distortion of the nigrostriatal axis.¹ Mass effect from the lesion or edema can result in compression of non-anastomosing end arterioles in the basal ganglia,⁵ resulting in hypoperfusion in the striatal-pallidal region.⁶

This hypoperfusion, without necessarily causing infarct, could cause reversible impairment in an area with high metabolism. One patient with craniopharyngioma presenting with parkinsonism, had decreased dopamine and its metabolites in the caudate and putamen, as well as decreased dopamine receptors in the caudate; the tumour likely compromised both the

presynaptic dopaminergic nigrostriatal neurons and postsynaptic dopamine receptors.³ Another patient had a falxine meningioma resulting in decrease in regional glucose metabolism in the striatum and normal presynaptic dopamine transmission on positron emission tomography scan.⁴ In our patient, there was dramatic improvement of his hemiparkinsonism post-operatively. This suggests that the mechanism underlying his hemiparkinsonism was reversible and was likely attributable to mechanical distortion and compression resulting in impaired metabolism within the nigrostriatal pathway.

Structural lesions can mimic Parkinson's disease. Imaging is recommended in patients with atypical features including pyramidal signs or focal weakness, postural instability without rigidity, impaired visual function, significant headache, seizures, poor responsiveness to levodopa and younger patients. The atypical feature in our patient was an upper motor neuron type of facial droop. It is important to perform a detailed clinical examination even in cases where the diagnosis seems obvious or is made "as the patient walks into the office" as treatable causes of parkinsonism do occur.

REFERENCES

1. Wu W, Shi JX, Cheng HL, et al. Hemangiopericytomas in the central nervous system. *J Clin Neurosci*. 2009;16(4):519-23.
2. Krauss JK, Paduch T, Munding F, Seeger W. Parkinsonism and rest tremor secondary to supratentorial tumours sparing the basal ganglia. *Acta Neurochir (Wien)*. 1995;133(1-2):22-9.
3. Garcia de Yébenes J, Gervas JJ, Iglesias J, Mena MA, Martín del Río R, Somoza E. Biochemical findings in a case of parkinsonism secondary to brain tumour. *Ann Neurol*. 1982;11(3):313-6.
4. Miyagi Y, Morioka T, Otsuka M, Fukui M. Striatal glucose metabolism and [¹⁸F]fluorodopa uptake in a patient with tumour-induced hemiparkinsonism. *Neurosurgery*. 1993;32(5): 838-41.
5. Ling MJ, Aggarwal A, Morris JG. Dopa-responsive parkinsonism secondary to right temporal lobe haemorrhage. *Mov Disord*. 2002;17(2):402-4.
6. Salvati M, Frati A, Ferrari P, Verrelli C, Artizzu S, Letizia C. Parkinsonian syndrome in a patient with a pterional meningioma: case report and review of the literature. *Clin Neurol Neurosurg*. 2000;102(4):243-5.