



Hyperkinetic transient ischemic attacks preceding deep ganglionic infarction in a patient with a treated parasellar chondrosarcoma

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ARTICLE INFO

Article history:

Received 13 June 2017

Accepted 23 October 2017

Keywords:

Chondrosarcoma

Radiation-induced atherosclerosis

Hyperkinetic transient ischemic attack (TIA)

ABSTRACT

A 44-year-old right-handed female with a past medical history of parasellar chondrosarcoma status post-surgical debulking and proton beam therapy (70 Gy) three years prior to presentation experienced several hours of brief, repetitive episodes of transient hemiballism and dystonia; this was followed by abrupt onset of fixed hemiparesis and dysarthria weeks later, ipsilateral to her prior hyperkinetic movements. She was found to have total occlusion of the right middle cerebral artery with focal stenosis of the proximal right A-1 segment of the anterior cerebral artery adjacent to the remnants of the chondrosarcoma. These focal areas of narrowing were attributed to accelerated atherosclerotic disease, an adverse effect of the radiotherapy used to treat her chondrosarcoma. As treatments improve and mean survival increases for intracranial malignancy, radiation-induced atherosclerotic disease with protean manifestations such as those presented in this case may be encountered more frequently.

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1. Case description

A 44 year-old right-handed female with a history of parasellar chondrosarcoma, treated three years prior with surgical debulking, and a combination of proton and photon radiotherapy at an outside institution (70 Gy total), developed abrupt onset of recurrent “jumping, twisting” and “flinging” movements of the left arm and leg lasting 20–30 s. Motor function and coordination were intact between recurrent episodes. She was treated with 1 mg of lorazepam and the movements resolved. An emergent CT head demonstrated stability of the chondrosarcoma remnants and no evidence of infarction. Limited sequence MRI (without diffusion-weighted sequences) performed the following day showed no change from previous post-treatment imaging. Seventeen days later, the patient presented with left-sided hemiparesis, left facial droop, and dysarthria. She was noted to have mild left hemineglect and a flattened affect.

MRI brain demonstrated a well-demarcated acute infarction in the right basal ganglia with evidence of slow flow in the right hemisphere and absence of the right middle cerebral artery (MCA) flow void on T2-weighted images (Fig. 1A–C). The patient was transferred to our institution where conventional angiography was performed which demonstrated occlusion of the right MCA at its origin with robust leptomeningeal collateral supply from the right anterior cerebral artery (ACA) and posterior cerebral artery. Additionally there was focal stenosis of the proximal A-1 segment of the right ACA (Fig. 1D and E). The areas of stenosis and occlusion were adjacent to the remnants of the chondrosarcoma and were suggestive of chronic focal atherosclerosis. The patient was started on aspirin and atorvastatin and was able to return to work one year later.

2. Discussion

Chondrosarcomas are rare intracranial neoplasms that result in morbidity through local compression or invasion. They typically

respond well to surgical debulking and high dose (typically proton) radiotherapy and have low five year mortality [1].

Radiotherapy may contribute to vasculopathy of medium and large vessels resulting in symptomatic cerebrovascular disease years after exposure. Animal models suggest that acute radiation-induced endothelial injury followed by chronic inflammation, or injury to the vasa vasorum may result in accelerated atherosclerosis leading to focal narrowing of the vessel lumen [2,3]. The patient's MRI demonstrated the “Ivy sign”, a radiographic sign initially described in patients with Moyamoya, which refers to diffuse leptomeningeal enhancement on post-contrast MRI and/or hyperintensity on FLAIR imaging, suggesting robust leptomeningeal collateralization, in our case due to chronic hypoperfusion of the MCA territories. These findings are in line with a chronic stenosis leading to occlusion [4]. Alternatively, cerebral infarction secondary to direct vascular compression from an intracranial tumor is exceedingly rare and often associated with rapid tumor expansion or hemorrhage, neither of which were noted in this case [2,5].

In our patient, the hyperkinetic movements and subsequent stroke are presumed to be secondary to occlusion of the right MCA with hypoperfusion to deep ganglionic structures [6,7]. Her initial presentation to medical attention, in retrospect, was a hyperkinetic TIA. It is unclear whether initiation of an antiplatelet agent at the time of initial presentation would have influenced her outcome. Currently, no trials adequately assess medical treatment options in primary or secondary stroke prevention in patients receiving head/neck radiation [2,3]. We recommend a high index of suspicion for radiation-induced vasculopathy in patients with transient paroxysmal neurological phenomenon and consideration for traditional vascular risk factor modification and intervention. As treatments improve and mean survival increases for intracranial malignancy, radiation-induced atherosclerotic disease with protean manifestations such as hyperkinetic transient ischemic attacks may be encountered more frequently.

Sources of support

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

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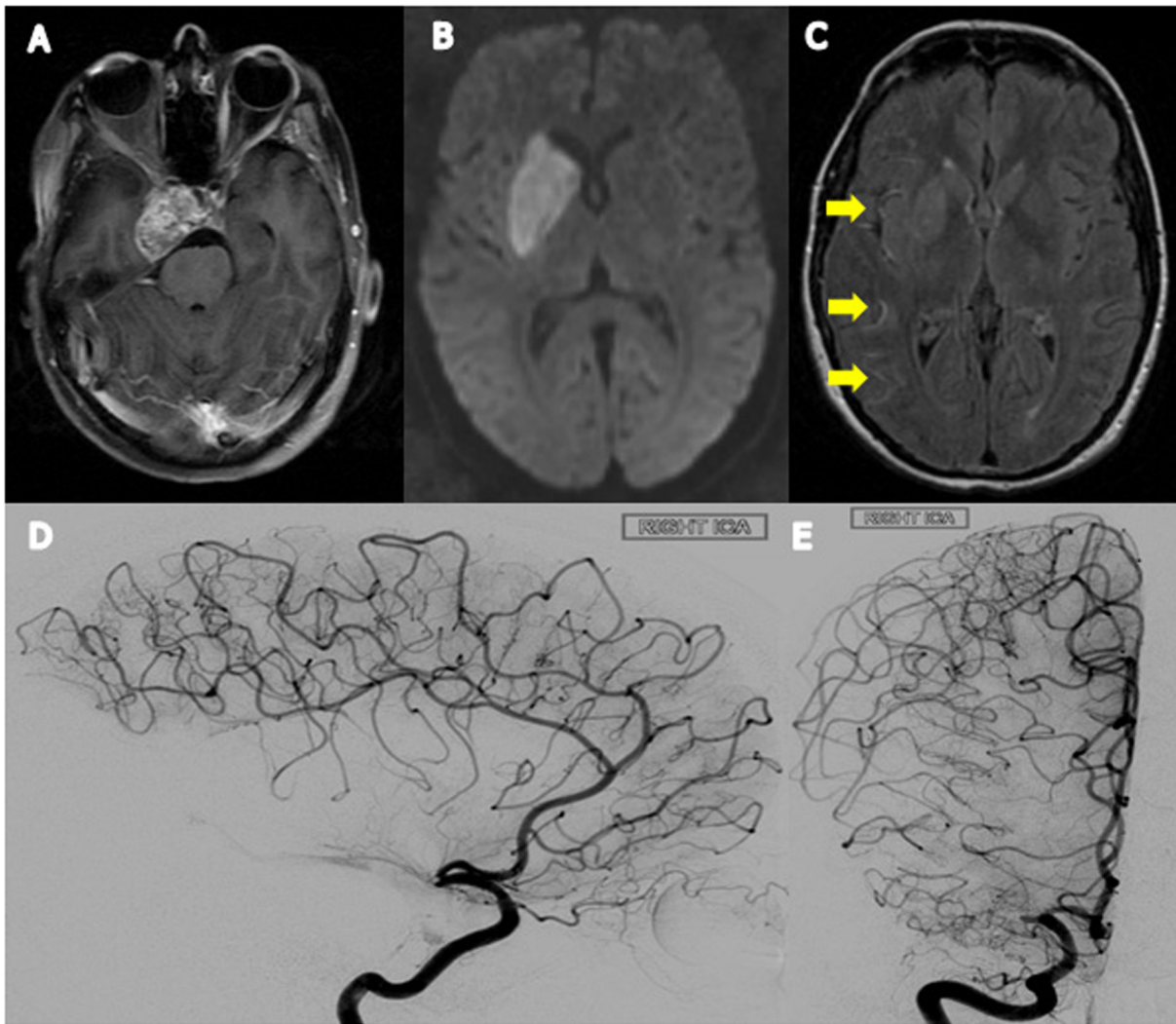


Fig. 1. (A) Axial T1-weighted FLAIR with contrast demonstrating residual chondrosarcoma involving the right skull base; (B) Axial DWI image demonstrating acute infarction of the right basal ganglia and both anterior and posterior limbs of the internal capsule; (C) Axial T2-weighted FLAIR image demonstrating asymmetric increased vascular signal (yellow arrows) over the right MCA distribution secondary to increased leptomeningeal collateralization from chronic hypoperfusion of the right MCA distribution—“Ivy sign”; (D and E) Conventional cerebral angiography of the right internal carotid artery demonstrating occlusion of the proximal right MCA M1 segment approximately 5 mm distal to its origin with robust leptomeningeal collateral arteries over the cerebral convexity.

Conflict of interest

The authors declare no conflicts of interest.

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<https://doi.org/10.1016/j.jocn.2017.10.076>