Hong Kong J Radiol. 2022;25:235-40 | https://doi.org/10.12809/hkjr2217563

BRIEF COMMUNICATION

British Society of Neuroradiologists Grand Round — Paediatric Neuroradiology: Brief Communication

TM Chiu[1], LHQ Chin[2]

[1]Department of Radiology, Hong Kong Children Hospital, Hong Kong
[2]Department of Radiology, Queen Mary Hospital, Hong Kong

INTRODUCTION

The British Society of Neuroradiologists (BSNR) Grand Rounds are webinars held biweekly (every 2nd and 4th Wednesday of the month) that feature interesting and educational neuroradiology case presentations from various institutions in the United Kingdom and around the globe. The Grand Rounds are open to all, and registration is free of charge (https://bsnr.org.uk/grandround/). The BSNR regularly post updates of these sessions and other related educational content via social media (https://twitter.com/thebsnr).

The Hong Kong College of Radiologists (HKCR) Paediatric Training Network is composed of all accredited paediatric radiology training centres in Hong Kong. In collaboration with the HKCR Paediatric Training Network, two teams of interventional neuroradiologists and paediatric neuroradiologists were selected to present BSNR Grand Rounds. The first of these collaborative BSNR Grand Rounds on neuro-intervention was held on 22 September 2021; the second on paediatric neuroradiology was held on 26 January 2022.

A total of four cases were presented as part of the BSNR Grand Round on paediatric neuroradiology. Two cases covered paediatric stroke: focal cerebral arteriopathy of childhood presented by Dr Philip Lee (Tuen Mun Hospital, Hong Kong); and multisystem smooth muscle dysfunction syndrome presented by Dr Claudia Cheung (Hong Kong Children Hospital, Hong Kong). In this report, we provide a summary and discussion of the other two cases on brain tumour–like mimics.

PRESENTATION EXPERIENCE

Cerebral Phaeohyphomycosis — Presented by Dr Leanne Chin

This case highlighted an intriguing and educational diagnostic challenge provided by an unusual paediatric cerebral tumour–like mimic in a 6-year-old boy with recent history of headache, and left upper limb seizure

Correspondence: *Dr LHQ Chin, Department of Radiology, Queen Mary Hospital, Hong Kong*
Email: chin.leanne@gmail.com

Submitted: 10 Apr 2022; Accepted: 30 Jun 2022

Contributors: Both authors designed the study, acquired the data, analysed the data, drafted the manuscript, and critically revised the manuscript for important intellectual content. Both authors had full access to the data, contributed to the study, approved the final version for publication, and take responsibility for its accuracy and integrity.

Conflicts of Interest: Both authors have disclosed no conflicts of interest.

Funding/Support: This study received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors.

Data Availability: All data generated or analysed during the present study are available from the corresponding author on reasonable request.

Acknowledgement: We thank Dr Chris KC Wong (Hong Kong Children Hospital, Hong Kong), Dr Elaine YL Kan (Hong Kong Children Hospital, Hong Kong), Dr Shubhabrata Biswas (The Walton Centre, Liverpool), and Prof Winnie CW Chu (Prince of Wales Hospital, CUHK, Hong Kong) for leading the virtual BSNR Grand Round; and the Hong Kong College of Radiologists (HKCR) and Dr YC Wong (Warden of HKCR) for the coordination and collaborative hosting with the British Society of Neuroradiology (BSNR).

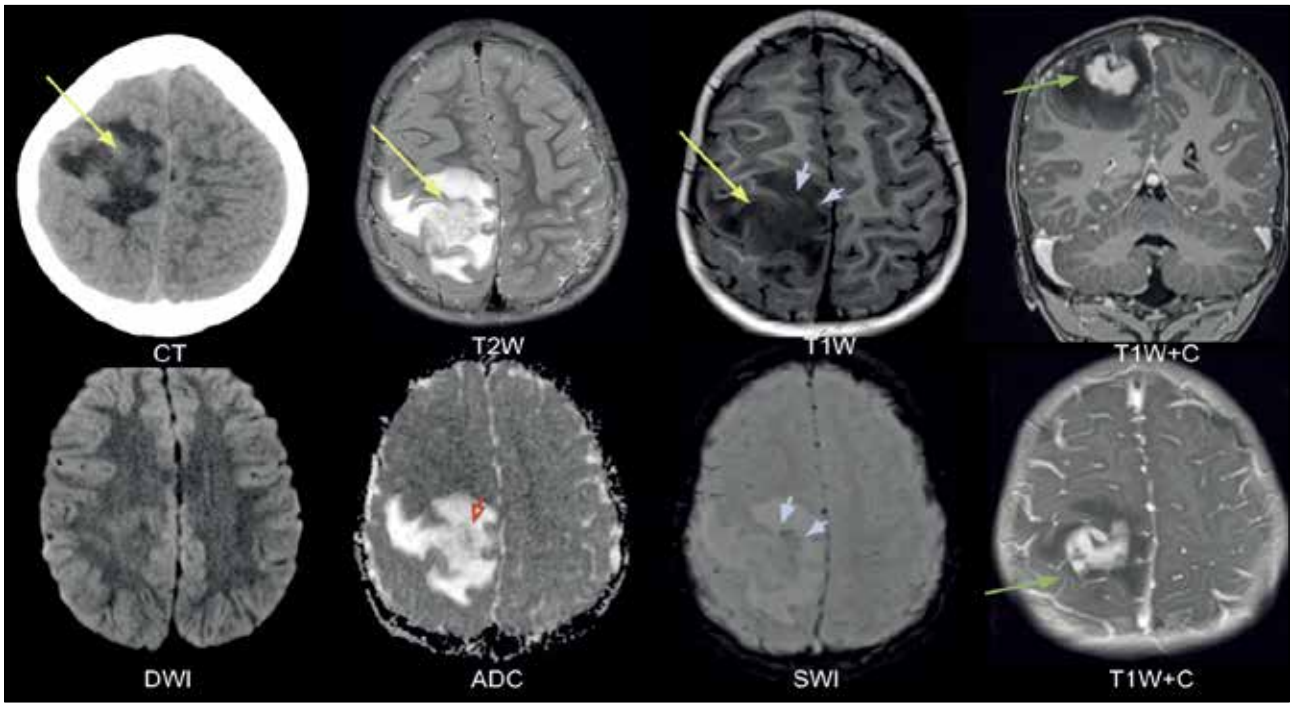

Figure 1. Non-contrast computed tomography (CT) and contrast-enhanced magnetic resonance imaging examinations of the brain. A solitary irregular intra-axial mass is seen at the right parietal lobe which is hyperdense on CT, isointense to grey matter on T1- and T2-weighted images (yellow arrows). Faint peripheral rim of susceptibility artefact is seen which is correspondingly T1-weighted hyperintense (short blue arrows). Focal central areas of restricted diffusion are seen as evident by low apparent diffusion coefficient value (open red arrow). Marked vivid contrast enhancement is demonstrated (green arrows).

for 1 day. Computed tomography and magnetic resonance imaging (MRI) examinations of the brain revealed a solitary intra-axial enhancing T2-weighted hypointense mass in the right upper parietal lobe (Figure 1). The mass exhibited a restricted diffusion pattern, with a susceptibility rim artefact and perilesional oedema. Subsequent MR spectroscopy of the lesion revealed an elevated Cho:NAA ratio and abnormal lipid-lactate peaks, whereas MR perfusion revealed no perfusion (Figure 2). The working diagnosis at the time was high-grade glioma, necessitating an emergency craniotomy. After a successful complete excision, histology revealed an unexpected diagnosis of cerebral phaeohyphomycosis.

On MRI, aggressive cerebral fungal infections can closely resemble high-grade brain tumours in terms of contrast enhancement, restricted diffusion, and vasogenic oedema.[1] On retrospective radiological review, however, thorough analysis of the lesion morphology reveals two characteristic diagnostic features[2]: finger-like extensions of fungal hyphae and paramagnetic elements of melanin production (Figure 3). Although abnormal choline and lipid-lactate metabolite peaks are frequently associated with high-grade malignancies, the absence of lesion perfusion is sometimes considered to be a paradoxical finding. Therefore, reliance on MR spectroscopy and perfusion imaging may not be useful in every case to differentiate between malignancy from aggressive fungal infection.[1,3,4]

An important take-away message from this case is that the occurrence of an isolated cerebral phaeohyphomycosis in an otherwise immunocompetent patient should prompt an active search for underlying CARD9 deficiency, as was confirmed in this patient via genetic testing. The case report by Lai et al[5] provides further detail on this patient and outcome.

As a result of our experience presenting this case at the BSNR Grand Round, we have gained new knowledge and skills in developing effective and systematic diagnostic pathways while avoiding radiological pitfalls. The differential diagnosis and case summaries of the pathologies were also well appreciated by the audience.

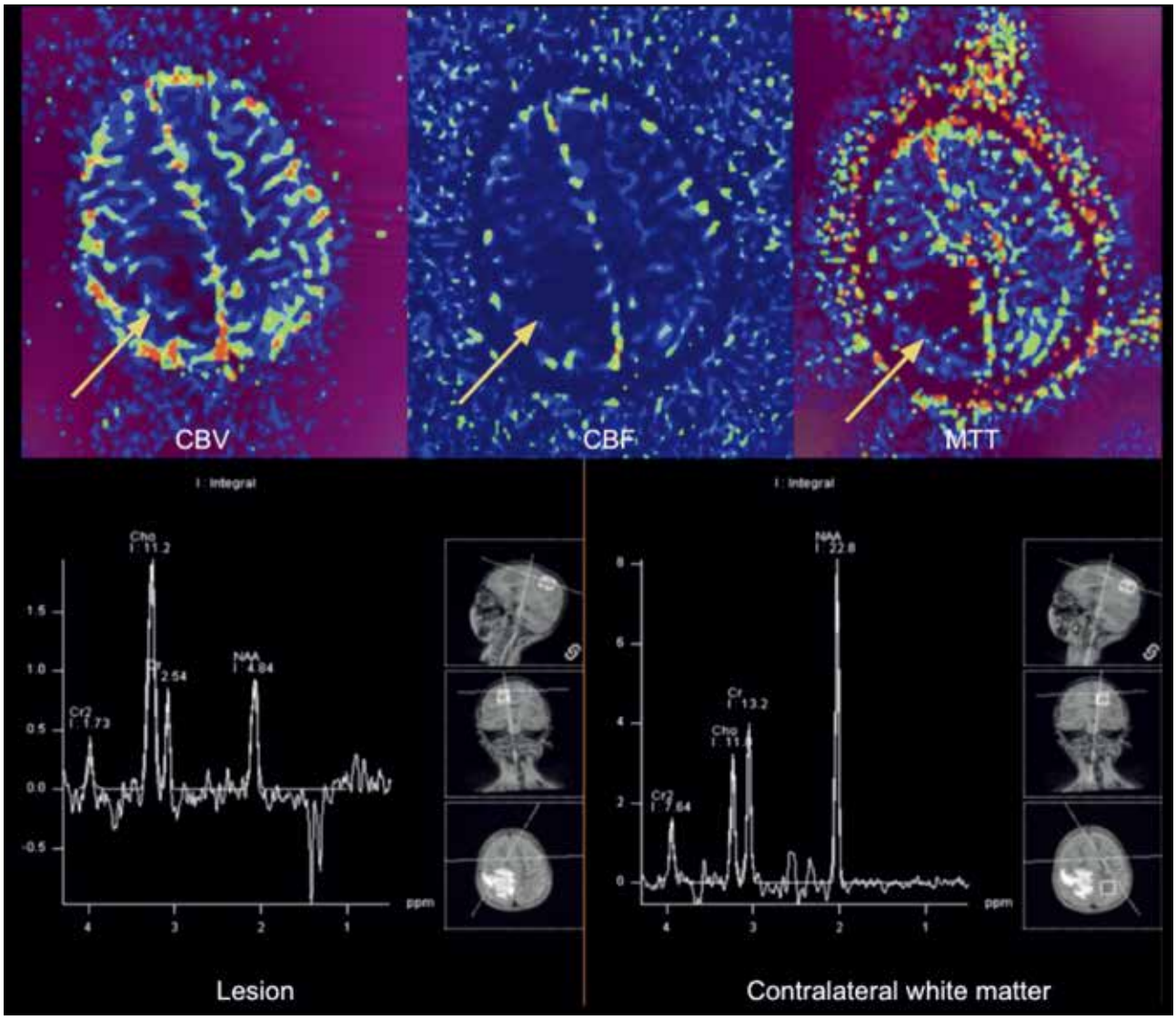

Figure 2. Magnetic resonance (MR) perfusion-weighted imaging (top row) demonstrates complete absence of cerebral blood perfusion within the lesion (arrows). MR spectroscopy (bottom row) acquired at intermediate echo time of 135 ms demonstrates abnormally elevated choline (Cho) peak (at 3.2 ppm), reduced creatine (Cr) peak (at 3.0 ppm), reduced NAA peak (at 2.0 ppm) and abnormal inverted lipid-lactate peak (at 0.9-1.4 ppm). This results in abnormal reduced NAA/Cho ratio of 0.43 and elevated Cho/Cr ratio of 4.48. The normal metabolite profile of uninvolved contralateral white matter is referenced on the bottom.

Multiple Cerebral Cavernoma — Presented by Dr Milly Chiu

This case provided another illustration of a brain tumour–like mimic, featuring a 4-year-old girl with good past health who was referred to the paediatric oncology team for suspected diffuse intrinsic pontine glioma. She presented with a 2-week history of gaze palsy. Initial computed tomography scan of the brain revealed a solitary lesion centred at the pons, which appeared multiloculated with predominantly hypodense content and a partial hyperdense rim suspicious of blood product or calcification (Figure 4). MRI was performed shortly afterwards, and the lesion appeared predominantly hyperintense on T1- and T2-weighted images with internal dependent hypointense contents. No significant solid enhancing component was seen (Figure 5). The initial imaging differentials considered were brainstem gliomas, possibly complicated with haemorrhage, or lesions that contain calcification, including ganglioglioma, rosette-forming glioneuronal tumour, or pilocytic astrocytoma.

However, on further review of the susceptibility weighted imaging sequence, there were numerous small intra-

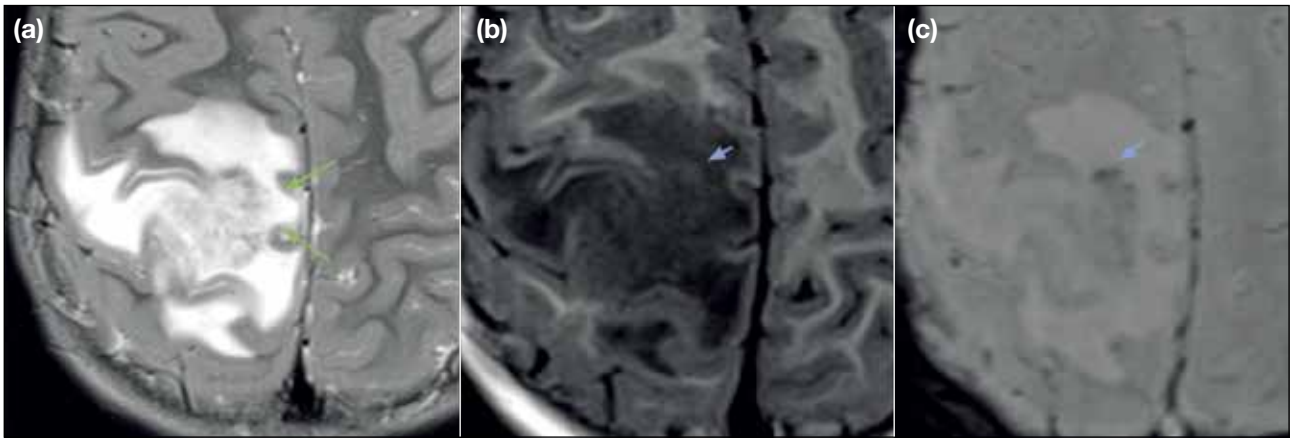

Figure 3. Close-up magnified views of the selected magnetic resonance imaging sequences highlight characteristic features of (a) finger-like extensions (or known as intracavitary projections) as seen as the fluffy borders on T2-weighted images (green arrows), and (b, c) paramagnetic elements (commonly melanin production) as seen as susceptibility artifacts and/or hyperintensities on T1-weighted images (short blue arrows).

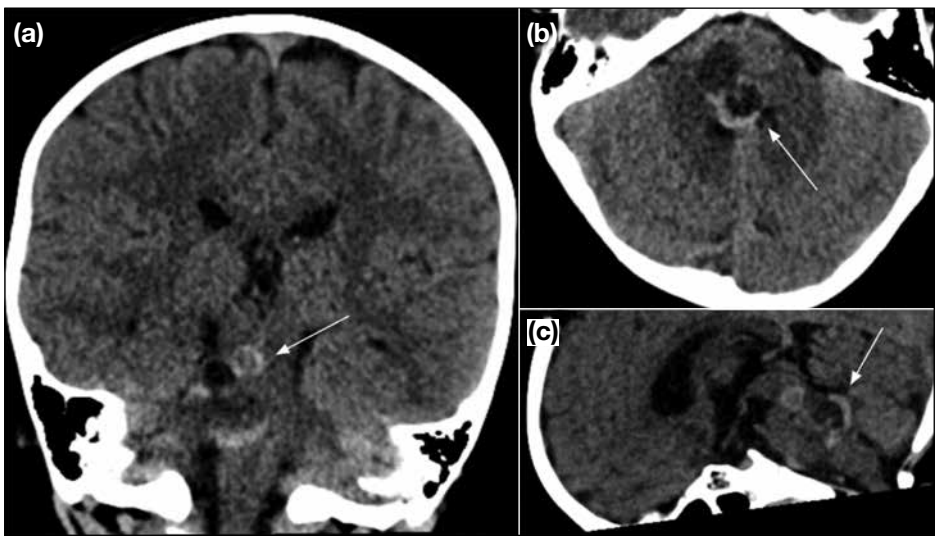

Figure 4. Non-contrast enhanced computed tomography brain with (a) coronal view, (b) axial view and (c) sagittal view showing a multilobulated lesion of predominantly cystic density with a partial hyperdense rim centred at the pontine region causing narrowing of fourth ventricle (arrows).

axial blooming foci in bilateral cerebral hemispheres. These lesions were inconspicuous on all other sequences (Figure 6). The final radiological diagnosis of multiple cavernoma was made, which was concordant with subsequent intraoperative and histopathological findings.

The most classically quoted description of cerebral cavernoma would perhaps be a "popcorn-appearance"—with a multilobulated lesion of heterogeneous signal intensity with a prominent hypointense rim on T2-weighted images, representing multiloculated haemorrhages of different ages.[6] This appearance was not appreciated in this case, such that the possibility of brainstem tumours was cautiously raised. The diagnosis of multiple cerebral cavernoma was made almost certain upon subsequently reviewing the susceptibility weighted images, the less likely differential being haemorrhagic metastases in the absence of a primary malignancy.

Presence of multiple cavernomas raises the concern of familial cerebral cavernoma malformation, and this is highlighted by this case where two disease-causing mutations — *KRIT1* and *CCM2* — were eventually confirmed on genetic studies.[7]

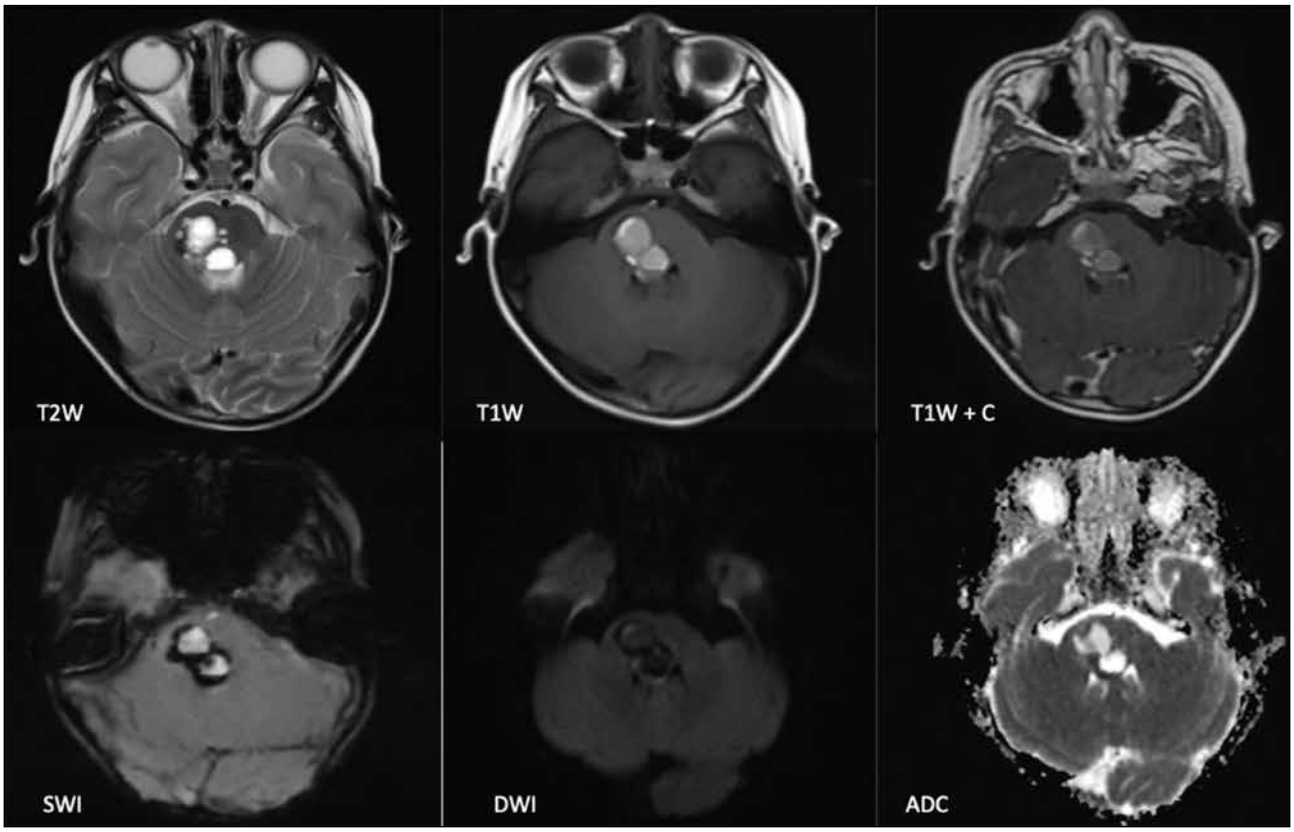

Figure 5. Magnetic resonance imaging brain with sequences as labelled. The pontine lesion showing hyperintense contents on T1- and T2-weighted sequences with a rim of blooming artefact. No significant solid enhancing component or restricted diffusion.

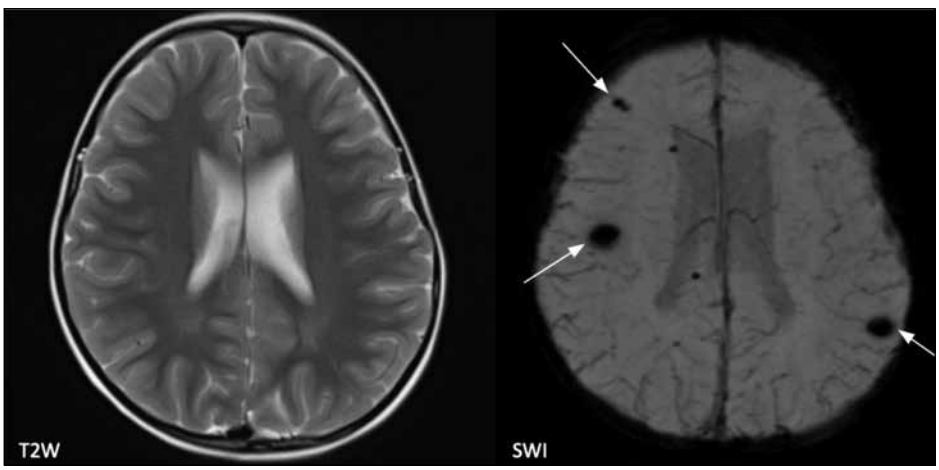

Figure 6. Selected axial images at the same level from T2-weighted and susceptibility-weighted imaging sequences of the magnetic resonance imaging brain showing multiple blooming artefacts (thin white arrows) at bilateral cerebral hemispheres which are only visualised on susceptibility-weighted images but not on T2-weighted images.

It was gratifying to receive a significant positive response from the audience, with the majority of them finding this case to be highly unusual, interesting, and educational to their training and practice.

CONCLUDING REMARKS

It was a rewarding experience to present at the BSNR Grand Round, to have received an overwhelming amount of support from seniors and colleagues and constructive

feedback from the international audience. We hope that the audience appreciated these two fascinating cases and learned that not all disease processes that result in mass formation are indicative of brain tumours. A wide spectrum of pseudotumour-like mimics exists with overlapping clinical and radiological features, but scrutiny of their unique imaging features will aid in the radiological diagnosis.

Similar to the BSNR Grand Rounds, the HKCR Paediatric Training Network is excited to launch regular online teaching rounds and webinars in the near future, with the goal of expanding professional training, learning opportunities, and collaboration between Hong Kong and international radiologists.

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