All that glitters is not gold: Rosai-Dorfman as a single cerebellar necrotic lesion

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DESCRIPTION

A 46-year-old man with a family history of renal cancer (father and grandfather), presented with a 5-day complaint of headache, nausea and photophobia. Neurological exam was intact. CT revealed a single mass in the right cerebellar hemisphere compressing the fourth ventricle and brainstem, without hydrocephalus. MRI showed heterogeneously contrast enhancement with surrounding oedema and central areas of necrosis (figure 1).

CT chest, abdomen and pelvis were unrevealing for suspicious systemic lesion. Preoperative assessment was unremarkable with no metabolic or haematological abnormalities. The patient was electively taken to the operating room, under general anaesthesia placed in prone position, with the head slightly flexed, on a Mayfield clamp. A suboccipital craniotomy extended to the foramen magnum was performed; the tumour was exposed and found to have a soft consistency and to be extremely vascularised and haemorrhagic. Frozen section indicated primary central nervous system (CNS) lymphoma, and gross total resection was achieved.

Postoperative MRI revealed no residual lesion, haemorrhages or other complications (figure 2).

Figure 1 (A) Non-contrast CT showing spherical mass in the right cerebellar hemisphere with surrounding oedema and mass effect on the fourth ventricle. (A–C) Axial, coronal and sagittal cuts of a T1 sequence MRI with contrast revealing a 2.8 cm spherical thick-walled heterogeneously enhancing mass in the medial right cerebellar hemisphere with surrounding vasogenic oedema and central areas of internal non-enhancement/necrosis, causing mass effect on the fourth ventricle.

Figure 2 (A–D) Two axial, a coronal and a sagittal cut of a T1 sequence MRI with contrast postoperative, confirming gross total resection of the lesion.

Final pathology report astonished with a diagnosis of primary CNS Rosai-Dorfman disease (figure 3). In addition, the patient developed postoperative aseptic meningitis, which is extremely unusual for this disease, and was treated with corticosteroids. Since the disease is potentially curable with complete resection alone, the patient will keep follow-up without adjuvant treatment.^{1–3}

Figure 3 (A) H&E stain shows a mixture of histiocytes and mature lymphocytes (40×). (B) Rare histiocytes have lymphocytes traversing their cytoplasm which is known as emperipolesis (100×). (C) The histiocytes are positive for S-100 by immunohistochemical stain (100×). The small, non-staining nuclei are lymphocytes traversing the cytoplasm. CD1a was negative. (D) The histiocytes are also positive for CD68 (20×).

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This is a very rare condition to present primarily in the CNS. Few cases have been reported in the literature and even fewer cases presenting as a single cerebellar lesion.^{1–3} Furthermore, all of them presented with usual imaging presentation for the disease, which is a homogeneous enhancing solid lesion on MRI, with dural relations, similar to a meningioma.^{1–3}

This case had an extremely rare presentation of the disease, with a solitary lesion and atypical imaging. It showcases that

some clues can lead to completely different conclusions, thus ‘all that glitters is not gold’.

Contributors GJA was responsible for study concept and design. GJA and ENB-I contributed to acquisition of data. All the authors were responsible for the analysis and interpretation of data. All authors contributed to drafting of the manuscript. RAH and ES contributed to critical revision of the manuscript for important intellectual content. All the authors were responsible for administrative, technical and material support. RAH and ES contributed to study supervision.

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Learning points

- ▶ Although single contrast-enhancing cerebellar mass with central necrosis should suscite suspicious for metastasis, one should never shut their eyes to other possibilities.
- ▶ Due to its lymphocytic nature, Rosai-Dorfman can be confused with lymphoma or other lymphoid diseases, and freezing should not be taken for valid in deciding extent of resection or adjuvant treatment when suspicious is made.
- ▶ Once diagnosis is made, gross total resection is usually curative, and no further treatment is necessary.

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