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Case Report

Hemorrhagic Presentation of Isolated CNS Neuroblastoma Recurrence: Case Report

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Abstract

Central nervous system (CNS) metastases are rarely seen in neuroblastoma (NB) and a hemorrhagic presentation is even more uncommon. We describe a case of neuroblastoma CNS relapse with haemorrhagic presentation in a 5 year old girl with a previous diagnosis of stage IV NB at age 2. At recurrence she presented with neurological deterioration and a left dilated pupil. Computed tomography (CT) of the brain showed a left temporal lesion, and she underwent a unilateral decompressive craniectomy and excision of the lesion that was reported as neuroblastoma metastasis. Stage IV NB, N-myc gene amplification and elevated serum LDH are proposed as risk factors for CNS metastases. In spite of the small sample sizes in the literature due to its low incidence, awareness of these "red flags" could lead to earlier diagnosis and treatment.

Keywords: *CNS Neuroblastoma; Recurrence; Haemorrhagic Presentation*

Introduction

Neuroblastoma (NB) is the most common extracranial solid tumor in children in their first year, representing 7% of all neoplasm [1]. Metastases are present in up to 70% of patients at the time of diagnosis, predominantly to liver, lymph nodes and bone morrow and bone [2].

Despite NB frequently spread to the skull and orbits, CNS metastases are rare [3] and the recurrence rate in patients that have undergone treatment is between 1.7% and 11.7% [4]. Some authors suggest that the dura acts as a barrier preventing direct invasion [5]. Hemorrhagic presentation has an even lower incidence [6].

Case Report

The female patient initially presented as a 2 year old, with fever, fatigue, weight loss, diarrhea and abdominal distension. Ultrasonography revealed a vascularised mass in the right kidney, further characterized with CT, showed a retroperitoneal mass extending to spinal canal from T10-T12.

Histological findings on bone marrow aspirate informed infiltrating NB, which was confirmed on biopsy of the retroperitoneal lesion. This was consistent with Metaiodobenzylguanidine (MIBG) scintigraphy findings. NMYc amplification was negative. She was diagnosed with a stage IV NB and commenced the COG NB protocol, and an enlarged right adrenal gland was noted on CT and surgically removed. Subsequent chemotherapy with Cyclophosphamide and Topotecan was followed by an autologous bone-marrow transplant, abdominal

36 months after treatment she was admitted with neurological deterioration (GCS 8) and a dilated left pupil. CT brain showed a left temporal lesion (Figure 1). A unilateral decompressive craniectomy and excision of the lesion was performed (Figure 2).

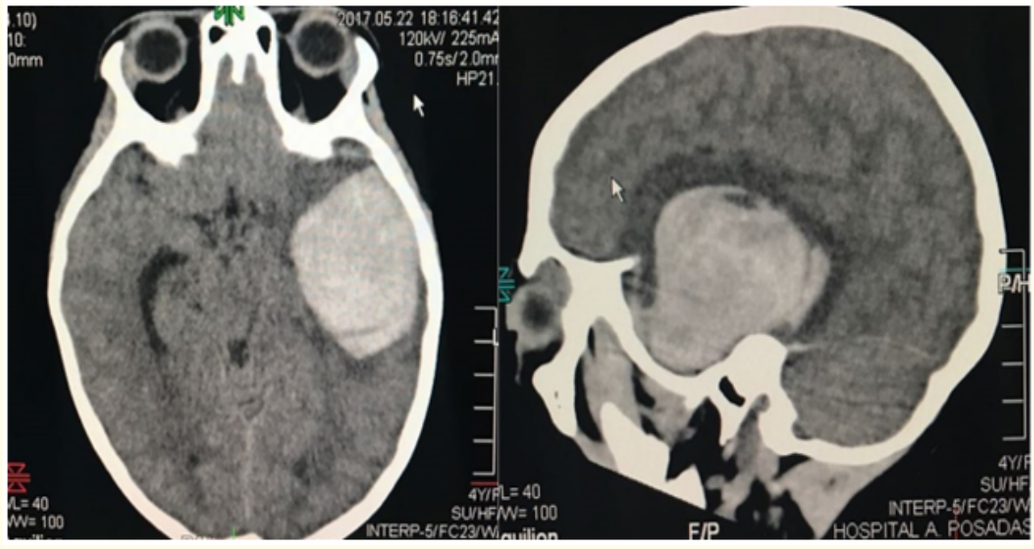

Figure 1: *Non-contrast Brain Computed Tomography (CT) shows a hyperdense lesion located at the left temporal lobe with a diameter 6 x 6 x 4,5 cm that produced mass-effect, midline shift, obliteration of the ambient and crural cistern, and signs of contralateral hydrocephalus.*

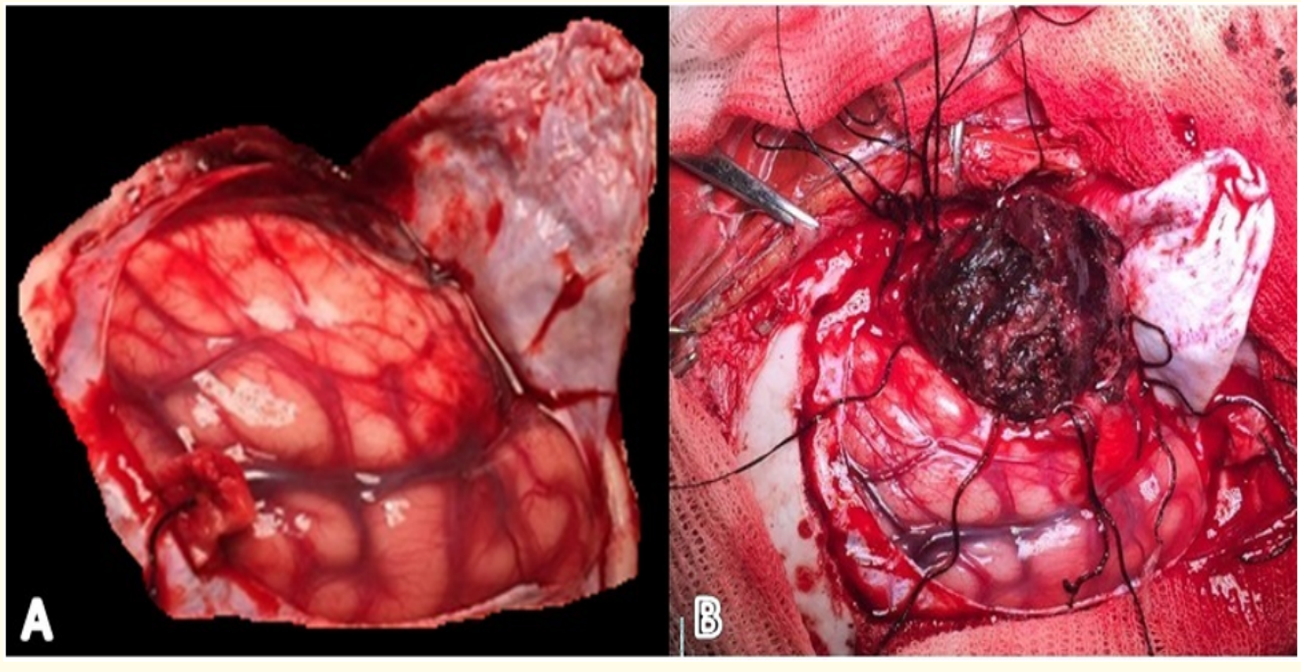

Figure 2: *A: Intraoperative image showing a herniated left temporal lobe with signs of cerebral edema. No signs of dural compromise. B: Intraoperative image showing the dissected hemorrhagic tumor lesion.*

Postoperative she had a mild right hemiparesis and reversal of anisocoria. Postoperative CT brain showed total resection (Figure 3A and 3B). Formal histopathology confirmed: metastatic neuroblastoma. The parents were informed of the diagnosis and poor prognosis, and decided not to continue with the treatment suggested by the Oncology department. The patient passed away six months later.

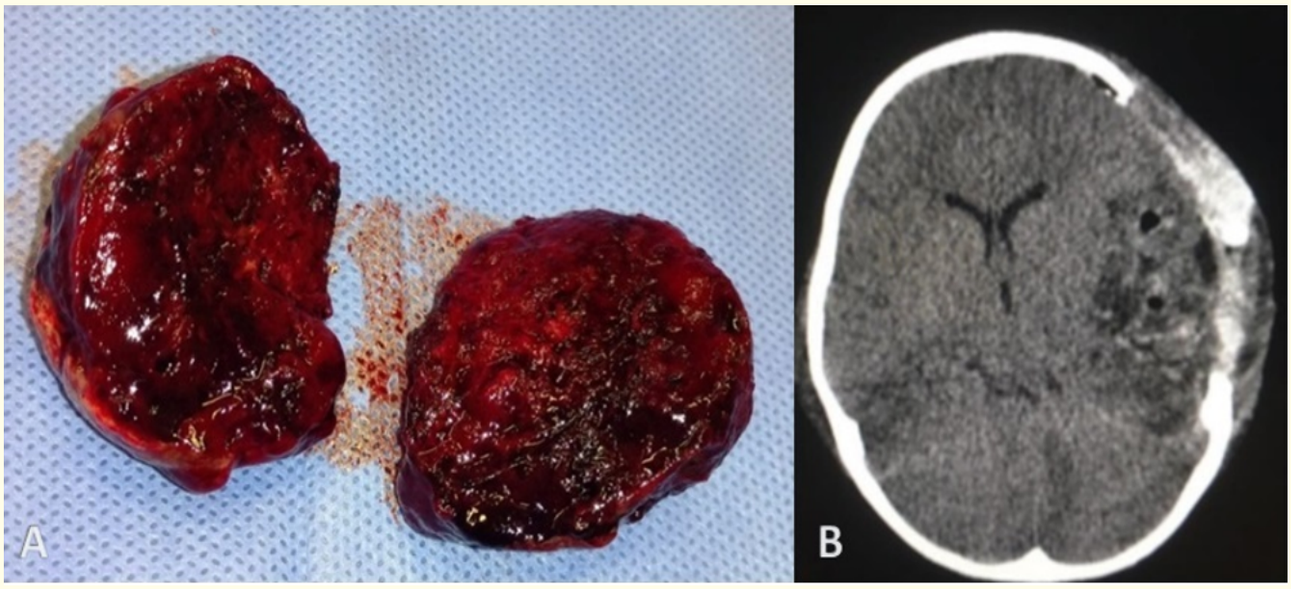

Figure 3: *A: Postoperative photography showing a resected tumor with hemorrhagic appearance. B: Postoperative brain CT scan, where we can observe a satisfactory tumor resection and a decompressive craniectomy.*

Discussion

Although the incidence of CNS metastases in patients with NB is very low, it is increasingly recognized as a stage IV complication [7]. This could be the result of the advances in treatment that improve patient survival, exposing them to a greater risk.

These lesions are associated with poor prognosis and late diagnosis [7]. MIBG scanning has proven not to be a reliable predictor of CNS disease, CT may be more sensitive in detecting bone metastases, with MRI a better method to detect CNS recurrence [8]. The MIBG scan of our patient was reported as non-pathological at the time of CNS disease diagnosis.

The median interval relapse from the diagnosis of NB to CNS metastases is between 5 and 32 months [9]. Our patient recurrences was diagnosed 36 months after the initial diagnosis.

N-myc gene amplification is considered a high risk factor for CNS metastases [10]. Our patient tested negative.

Elevated serum LDH over 1500 U/mL is a related finding to CNS metastases [9]. Our case showed LDH of 6.084 U/mL at diagnosis, and 857 UI/mL at relapse.

Matthay., *et al.* [10] reported that stage IV NB showed a risk of CNS metastases.

In our case we recognize two “red flags” as high risk factors: stage IV NB and elevated LDH values at the diagnosis.

We suggest that contrast-enhanced MRI should be included in follow up for NB patients with significant risk factors of CNS involvement, to enable early diagnosis and treatment. However, we acknowledge that a larger sample of patients would be needed to strengthen this recommendation.

Conclusion

Neuroblastoma CNS metastases is a rare entity and haemorrhagic presentation is even more unusual, associated with poor prognosis and late diagnosis.

Stage IV NB diagnosis, N-myc gene amplification and serum LDH over 1500 U/ml are proposed as high risks factors of CNS metastases.

In spite of the small sample sizes in the literature due to its low incidence, awareness of these "red flags" could lead to an early diagnosis and treatment.

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