An Extragonadal Germinative Tumor Presenting as Hemorrhage within the Medulla Oblongata: A Case Report

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1. Background

Germ cell tumors (GCTs) develop from primordial germ cells of the human embryo. They are typically classified histologically into two categories: seminomas and non-seminomatous germ cell tumors (NSGCT).[1,2]

Testicular yolk sac tumors (YSTs) are a rare subtype of malignant NSGCT which are usually accompanied by elevated serum alpha-fetoprotein (AFP) and can be found in a pure form or mixed with other germ cell tumors.[3,4]

2. Case Introduction

A previously healthy 19-year-old patient presented to the neurological emergency department with sudden onset of dysarthria, dysphagia, and hypoesthesia with paresthesia on the right side of the face and left side of the body. Family history revealed his father had a testicular tumor which was surgically treated with orchiectomy.

Laboratory examinations revealed elevated C-reactive protein (95 mg/L), alpha-fetoprotein (AFP = 13308 kU/L) and HCG (30940 U/L) levels.

3. Imaging

A non-contrast computed tomography (CT) scan of the head showed an intramedullary haematoma on the right side of the medulla oblongata.

Contrast-enhanced magnetic resonance imaging (MRI) of the head was done and it revealed a cavernomatous lesion in the posterior medulla oblongata which was hypointense on T2-weighted and isontense on T1-weighted images, had no signs of restricted diffusion on DWI or post-contrast enhancement, had signal loss on SWI, and was surrounded by vasogenic edema. It was later concluded that the lesion was a metastasis.

An x-ray and CT scan of the thorax were performed which revealed metastases in the right upper and lower lobes of the lung.

Contrast-enhanced CT scan of the abdomen showed a large heterogeneous retroperitoneal tumor with central necrosis and signs of neovascularization, invasion and suspected occlusion of left renal vein and invasion of a spinal vein. A subcapsular 12 mm large liver metastasis was also seen on the border of segments II and IVa.

A scrotal ultrasound was performed which showed bilateral microlithiasis and no signs of a primary tumor.

4. Management

To further characterize the lesion a fine needle aspiration biopsy of the retroperitoneal lesion was performed which revealed histological characteristics of a yolk sac tumor.

Stereoatactic radiotherapy of the brain metastasis was performed with 54 Gy in 5 fractions during the first hospital stay.
The patient was treated with 6 courses of chemotherapy.

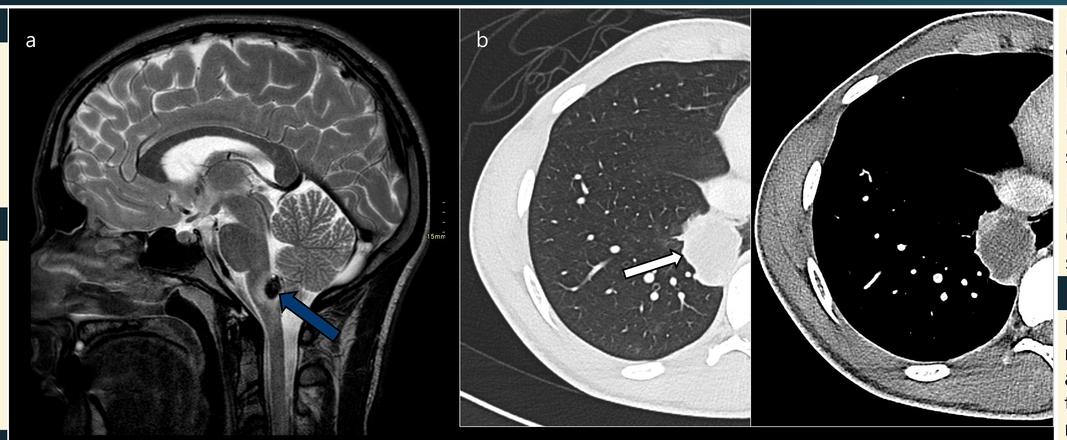

Figure 1. (a-b)

a. Contrast-enhanced MRI of the brain (T2 weighted, sagittal view) showing a hypointense cavernomatous lesion in the posterior medulla oblongata (blue arrow), sorrounded by vasogenic edema - a metastasis
b. Contrast-enhanced CT scan of the thorax (axial view, venous phase, lung window (left) and soft tissue windows (right)) demonstrating a metastasis (white arrow) in the right lower lobe

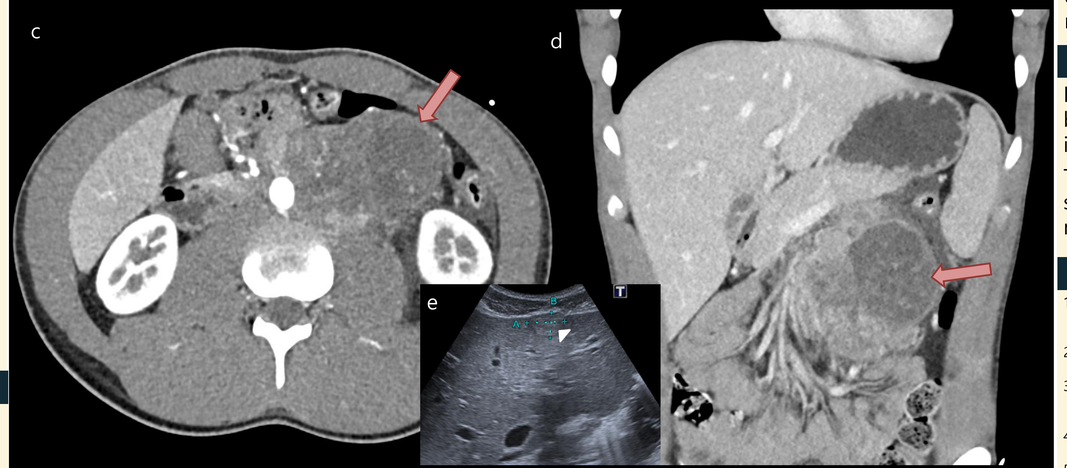

Figure 2. (c-d): Contrast-enhanced CT scan of the abdomen in venous phase axial (c) and coronal (d) views revealing a large heterogenous retroperitoneal tumor (red arrows) with central necrosis, signs of neovascularization, invasion of left renal vein and of a spinal vein, and an abdominal ultrasound (e) showing a subcapsular isoechoic oval-shaped lesion with hypoechoic halo (white arrowhead) - liver metastasis

During the period of chemotherapy serum AFP and β-HCG levels decreased. No serious adverse effects of chemotherapy occurred. The patient's neurological symptoms quickly improved.

Pulmonary metastasectomy by video-assisted thoracoscopic surgery (VATS) was performed later on. The retroperitoneal metastasis was surgically resected.

Follow up studies after surgery and chemotherapy were regularly performed and no new lesions were found in the abdomen, thorax or CNS. One year after the diagnosis the patient was symptom free and the serum tumor markers were within normal range.

5. Discussion

In rare cases of germ cell tumors in males primary testicular lesions are not clinically or radiologically evident yet extragonadal lesions are present and most commonly found in the midline (retroperitoneum, mediastinum, the CNS), and occasionally in the lungs and liver. Such lesions may either represent metastases from a "spontaneously regressed testicular tumor" (also called "burned-out testicular tumor") or primary extragonadal germ cell tumors.[3-7] Patients with burned-out testicular tumors often have non-specific findings at ultrasound such as microcalcification (as was seen in our patient) and parenchymal heterogeneity in one or both testes but the diagnosis can only be confirmed by histological examination which is not routinely performed.[7]

6. Conclusion

In the case presented here no primary lesion was radiographically evident but since no histological examination of testicular tissue was performed it is not possible to determine whether a "burned-out" tumor was present.

The goal of this report is to present a case of a young patient with clinical signs of hemorrhage in the medulla oblongata which was found to be a manifestation of a rare case of an extragonadal germ cell tumor.

7. References

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