A Case Report of Yolk Sac Tumor in Cerebellar Hemisphere and Review of Literature

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Case report

Keywords: yolk sac tumor, cerebellar hemisphere, alpha-fetoprotein, histopathological examination, imaging

Posted Date: July 8th, 2020

DOI: <https://doi.org/10.21203/rs.3.rs-38769/v1>

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Abstract

Background: Yolk sac tumor also known as endodermal sinus tumor, is a rare intracranial germ cell tumor. We reported a case of yolk sac tumor in cerebellar hemisphere, and reviewed associated literatures. The majority of tumor locations are near the midline. On review of literature, very few case reports of intracranial yolk sac tumor have been published, and there is only one case report has described a yolk sac tumor arising from the cerebellum. Case presentation: A two years old boy admitted to our hospital due to headache and unsteady gait for six days. CT and MRI demonstrated a tumor in the right cerebellar hemisphere, and the blood and cerebrospinal fluid alpha-fetoprotein were found increased. It was diagnosed as yolk sac tumor after operation confirmed by histopathological examination. Postoperative chemotherapy was performed, and the patient suffered no tumor recurrence one year and a half after the surgery. Conclusions: The clinical characteristics and imaging diagnosis of intracranial yolk sac tumor are lack of specificity, the confirmed diagnosis is depending on the combination of elevated alpha-fetoprotein and histopathological examination.

Introduction

Yolk sac tumor also known as endodermal sinus tumor, is a rare intracranial germ cell tumor (14). The incidence of intracranial germ cell tumors is low in all central nervous system tumors, less than 5% in western countries, but may be more common in Southeast Asia (8, 11). The tumor is more common in childhood and adolescence, and has a male predominance (6). The germinomas is most common in germ cell tumors, followed by teratoma, endodermal sinus tumor and choriocarcinoma are rare (10). The majority of locations are near the midline, such as the saddle and pineal region, basal ganglia and the thalamus (24, 4, 12), a few may occur in the lateral ventricles, third ventricle, cerebral hemispheres or brainstem (1, 19, 25). On review of literature, very few case reports of yolk sac tumor have been published. To our knowledge, there is only one case report has described a yolk sac tumor arising from the cerebellum (1).

Methods

We analyzed the clinical characteristics, diagnosis, treatment, CT and MRI imagings, histopathological data of a case of yolk sac tumor in cerebellar hemisphere, and reviewed associated literatures.

Case Presentation

Clinical characteristics

A two years old male admitted to our hospital due to headache and unsteady gait for six days. Six days before admission, he occurred headache, paroxysmal, with unsteady gait. No disturbance of consciousness, no vomiting, no significant visual impairment, no limb movement disorder. Urine and stool tests were normal.

Laboratory tests

Blood alpha-fetoprotein (AFP): 90.4 ng/ml, cerebrospinal fluid AFP: 18.4 ng/ml. Serum human chorionic gonadotropin beta (β -HCG): < 1 mIU/ml, CSF: β -HCG 3.79 mIU/ml.

Imaging

CT scan: a cystic-solid mixed density mass lesion was found in the right cerebellar hemisphere, with perifocal edema and strong solid components enhancement. The midline structure was shift to left. CT-angiogram (CTA) demonstrated the branch of the right anterior inferior cerebellar artery distributed in the mass (Fig. 1).

MRI scan: the signal of the solid components of the tumor on T1WI was isointensity, surrounding cystic hypointensity. Heterogeneous hyperintensity on T2WI, with strong gadolinium contrast enhancement (Fig. 2).

Surgical findings

The tumor was located in the right cerebellar hemisphere and around blue cyst wall. The cyst fluid was yellow, bloody, contained mucus-like material. The tumor was up to the cerebellar tentorium and down to the atlas level, with dimensions of 5 cm * 4 cm * 4 cm. The circumscribed red firm tumor is extremely rich in blood supply. The blood supply came mainly from the ventral posterior inferior cerebellar artery (Fig. 3).

Histopathological examination

Hematoxylin and eosin (H&E) staining showed the glomerular-like structures formed by tiny blood vessels, which covered single layer of columnar cell, protruded into the cavity consisted of flattened cells. Cytoplasmic and extra-cellular eosinophilic globules were visible. Immunohistochemistry: PLAP (+), CD117 part (+), AFP part (+), CK (+), Vim (+), CD99 part (+), EMA (-), NSE (-), Syn (-), CD57 (-), NF (-), GFAP (-), α -Inhibin (-), Ki67 (+) > 70% (Fig. 4).

Follow-up

Two weeks after surgery, MRI showed complete resection of the tumor. Postoperative chemotherapy was performed, and the patient suffered no tumor recurrence one year and a half after the surgery (Fig. 5).

Discussion

The central nervous system is one of several extragonadal sites of presentation of germ cell tumors. Yolk sac tumor is a nongerminomatous germ cell tumor, also known as endodermal sinus tumor (14). Germ cell tumors originated in embryonic germ cells, that were evolved pluripotent stem cells and embryonal carcinoma stem cells, further formed embryonal carcinoma, choriocarcinoma, yolk sac tumor and teratoma (6, 15).

Yolk sac tumor was the product of pluripotent stem cell differentiation in the extraembryonic structure, and characterized by abnormal development of mesoderm in the embryo and endoderm in the yolk sac. The tumor texture was firm, with hemorrhage and necrosis, and could be local infiltration, or spread along the subarachnoid space, and also along the VP shunt tube planted into the peritoneal cavity (16, 18). Yolk sac tumor cells are undifferentiated epithelial cells of the primitive endodermal sinus, arranged in irregular labyrinthine-like adenoid structures, often formed micro-cystic or cribriform. The glomerular-like structures formed by tiny blood vessels, which covered single layer of cubic cell, can be seen protruded into the cavity consisted of flat cells. Schiller-Duval bodies, formed by tubulopapillary sinusoidal structures with central vascular core and cuboidal to columnar epithelial-like cell lining, as well as cytoplasmic and extra-cellular eosinophilic globules are diagnostic features (6, 16).

The majority of locations are near the midline, such as the saddle and pineal region, basal ganglia and the thalamus (24, 4, 12), a few may occur in the lateral ventricles, third ventricle, cerebral hemispheres or brainstem (1, 19, 25). The imaging diagnosis of intracranial germ cell tumors in typical regions are more clear, but in rare site are easily misdiagnosed. The appearance of yolk sac tumours demonstrated a variety of imaging features, usually with heterogeneous enhancement (20, 23, 3). In this case the tumor located in the right cerebellar hemisphere, occurred in a two years old boy. It was difficult to distinguish with cerebellum glioma, hemangioblastoma and medulloblastoma. The diagnosis was depending on the combination of elevated AFP and histopathological examination (9, 21).

Tumor markers in intracranial germ cell tumors could be detected in a lot of kinds, including AFP, β -HCG, placenta alkaline phosphatase (PLAP), carcinoembryonic antigen (CEA) and so on. These positive markers indicated the possibility of intracranial germ cell tumors. The efficacy of the treatment of germ cell tumors could be evaluated by the changes of these tumor markers. If AFP and β -HCG elevated or only AFP elevated should be considered nongerminomatous malignant germ cell tumors (9, 21).

Large differences are existed in the prognosis of germ cell tumors. The prognosis of intracranial germ cell tumors are associated with pathology. The prognosis of germinomas and mature teratoma are the best. Pure germinomas could be cured, most literatures reported more than 90% of 5 years survival rate, and more than 80% of ten years survival rate. The prognosis of immature and malignant teratoma and germ cell tumors were poorer. Embryonic carcinoma, choriocarcinoma, yolk sac tumor and mixed germ cell tumor had highest malignant degree and the worst prognosis (17). Matsutani et al. reported the 3 years survival rate of germinomas was 70%, but much lower of nongerminomatous germ cell tumors (10).

There are various reports of current treatment options of intracranial germ cell tumors (7). Germinomas have long been known to be highly curable with both radiotherapy and chemotherapy (2, 5). Nongerminomatous germ cell tumors are benefited from a combination of surgical resection, chemotherapy and radiotherapy (13, 22). However, specific treatment of nongerminomatous germ cell tumors is much less clear. Further multicenter studies are needed to help improve treatment approaches and prognosis.

Conclusion

The clinical characteristics and imaging diagnosis of intracranial yolk sac tumor are lack of specificity, the confirmed diagnosis is depending on the combination of elevated AFP and histopathological examination. There are various reports of current treatment options and prognosis, which need further study.

Declarations

Ethics approval and consent to participate

The study protocol was approved by our institution's Committee on Clinical Research Ethics. Informed consent was obtained from the legal guardians.

Consent for publication

All authors have contributed to, read and approved the final manuscript for publication.

Data Sharing

The additional unpublished image data is available upon request from the corresponding author.

Conflict of interest statement

There are no conflict of interest exists for any of the authors.

Funding

The project was funded by General project of Chongqing Science and Technology Commission (cstc2019jscx-msxmX0143).

Author Contributions Statement

Lusheng Li and Yuting Zhang collected data and wrote the paper. Ling He revised the paper. All authors approved the final version of the paper.

Acknowledgements

The authors would like to thank Dr. Jin Zhu (Department of Pathology, Children's Hospital of Chongqing Medical University), for his contributions to this work.

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Figures

Figure 1

A: Axial CT demonstrate a cystic-solid mixed density mass in the right cerebellar hemisphere, with perifocal edema. B: Axial CT with contrast demonstrate strong solid components enhancement. C: CTA demonstrate the blood supply of the right anterior inferior cerebellar artery.

Figure 2

A: Axial T1-weighted MRI showing isointensity of the solid components of the tumor, surrounding cystic hypointensity; B: Axial T2-weighted MRI showing heterogeneous hyperintensity of the solid components of the tumor, surrounding cystic hyperintensity; C,D: Contrast-enhanced axial and sagittal T1-weighted MRI showing strong solid components enhancement.

Figure 3

Surgical findings: the circumscribed red firm tumor is extremely rich in blood supply.

Figure 4

A: HE * 200, showing the glomerular-like structures formed by tiny blood vessels, which covered single layer of columnar cell, protruded into the cavity consisted of flattened cells; B: HE * 400, showing cytoplasmic and extra-cellular eosinophilic globules; C: The tumor cells were immunoreactive for AFP * 200.

Figure 5

A.B: Contrast-enhanced axial and sagittal T1-weighted MRI showing complete resection of the original tumor, two weeks after surgery; C.D: Contrast-enhanced axial and sagittal T1-weighted MRI showing no tumor recurrence one year and a half after the surgery.