case report

Unusual anatomic location of a primary intracranial yolk sac tumor

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Ann Saudi Med 2011; 31(3): 298-300

DOI: 10.4103/0256-4947.76410

A nongerminomatous germ cell tumor occurring in the brain parenchyma is extremely rare. A 2-year-old boy presented with symptoms of abnormal movements in the hand and mouth. MRI scanning revealed a lesion occupying the left temporoparietal region. Craniotomy was performed and the tumor was removed by en bloc resection. Histological examination revealed that the tumor was arranged in a reticular pattern, and Schiller-Duval bodies were evident at the center of the tumor. Immunohistochemical study showed that the tumor cells were positive for alpha-fetoprotein and vimentin, but negative for glial fibrillary acidic protein. The histological diagnosis was pure yolk sac tumor).

Yolk sac tumors can rarely present intracranially. Typically, they are located midline in the pineal region or the suprasellar region, but curiously, this has not been the case in an increasing number of reports. Primary intracranial yolk sac tumor, arising within the substance of the brain, is exceedingly rare but has been well reported. We report this case as an example of this interesting phenomenon, which can be confusing for the pathologist and radiologist.

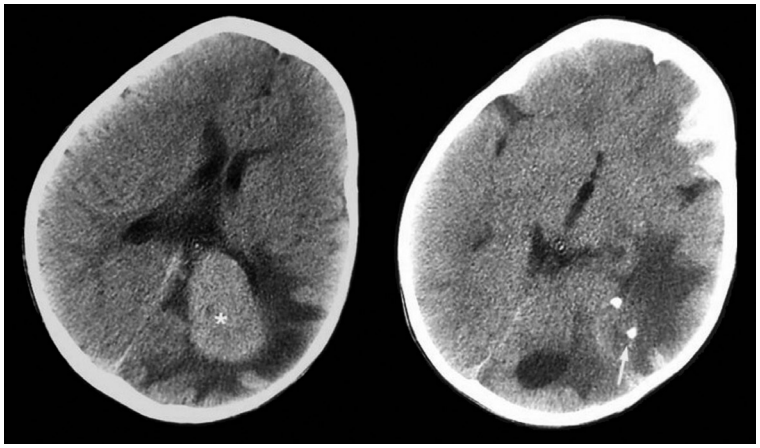

Figure 1. Precontrast transaxial images of the brain demonstrate high-density intra-axial para-midline left temporoparietal mass (star) with two foci of calcifications (arrow); perilesional edema is seen as hypodensity.

CASE

A 2-year-old boy was admitted to King Hussein Medical Center, Jordan, in 2005, with focal epileptic seizure. He complained of abnormal jerky movements in the mouth and the right hand. He had recurrent episodes of vomiting. Physical examination showed papilledema, partial third nerve palsy and a positive Babinski sign, along with fine shaky movements in the right hand and mouth. CT and MRI scans with contrast were performed, which demonstrated a tumor located within the left temporoparietal lobe (**Figures 1, 2**). An elective surgery was scheduled, and the tumor was resected and sent for histopathology. Histopathologic examination showed a tumor with different patterns. Reticular, cystic and solid sheets were all prevalent (**Figure 3**). The cells were relatively large, with a clear cytoplasm and vesicular nuclei. Scattered mitotic activities were noted. At the center of the tumor, Schiller-Duval bodies were evident (**Figure 4**). Periodic acid-Schiff stain showed intracytoplasmic and extracellular eosinophilic globules, which were diastase resistant. Immunohistochemical study is shown in **Figure 5**. The final diagnosis was pure yolk sac tumor. A thorough imaging study of the chest, abdomen, pelvis and testes failed to detect any primary tumor. Hence the diagnosis was changed to pure primary intracranial yolk sac tumor.

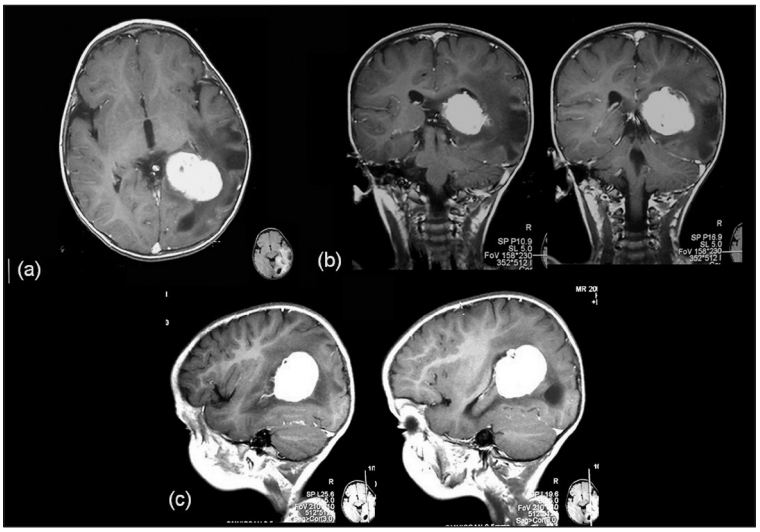

Figure 2. Axial (a), coronal (b) and sagittal postcontrast T1WI MRIs of the brain for the same patient demonstrate homogenous enhancement of the lesion with surrounding vasogenic edema. The lesion appears off-midline and deforming the trigone of the left lateral ventricle.

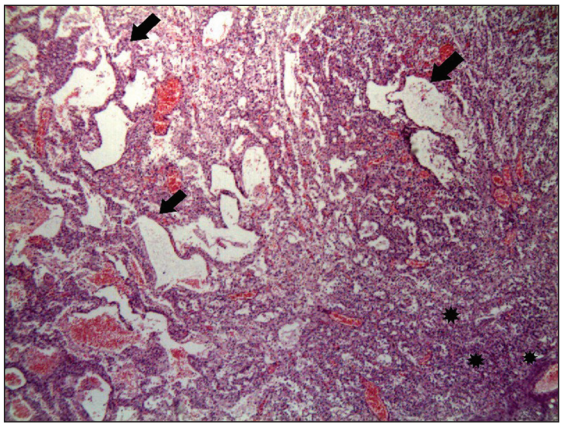

Figure 3. The tumor shows an intervening meshwork of irregular tissue spaces and anastomosing channels. Cystic spaces (arrows) and solid areas are also formed (stars) (hematoxylin and eosin stain, ×40).

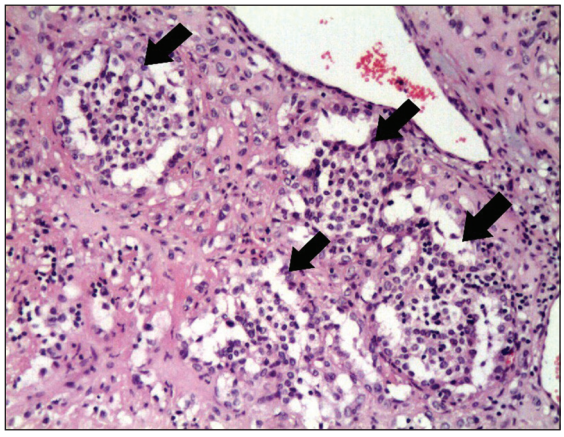

Figure 4. Schiller-Duval bodies: bilayered festoons of cells surrounding a fibrovascular core, reminiscent of primitive glomeruli that are characteristic of yolk sac tumors (arrows) (hematoxylin and eosin stain, ×200).

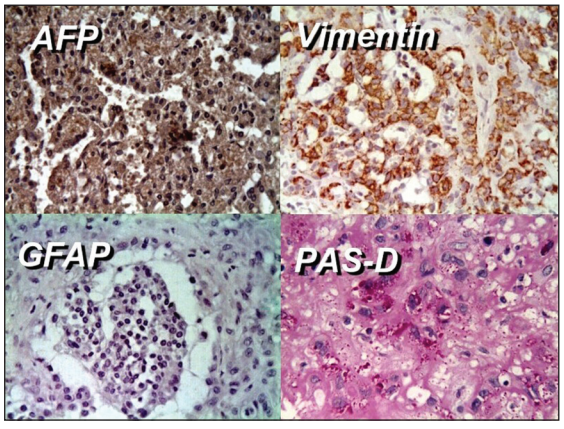

Figure 5. The tumor cells are positive for alpha-fetoprotein and vimentin but negative for glial fibrillary acid protein. Periodic acid Schiff-positive diastase-resistant globules are evident.

DISCUSSION

Intracranial germ cell tumors are rare and constitute less than 2% of all intracranial neoplastic lesions.[1] Among these, pure yolk sac tumor is rare. It typically presents in the pineal region or the suprasellar area.[2] Other unusual sites for presentation have been described, including the cerebellum,[3] fourth ventricle,[4] and frontal lobe.[5,6] To the best of our knowledge, previously only one case has been reported of tumor to arise from the temporoparietal lobe.[7] Primary spinal cord yolk sac tumor has also been reported.[8]

The clinical symptoms of primary intracranial yolk sac tumors depend on their anatomic location. Those of the pineal gland present with increased intracranial pressure. In our case, the patient complained of abnormal mouth and right hand movements and increased intracranial pressure. A test for serum level of alpha-fetoprotein (AFP) was not performed as the diagnosis was not expected.

While the 5-year survival in cases of intracranial germ cell tumor is over 90%, the prognosis in cases of yolk sac tumor is relatively poorer than in cases of germinoma.[9]

It has been shown that there is a strong tendency for spread via cerebrospinal fluid.[8] A combination of surgical resection, chemotherapy and radiotherapy is recommended.[4] In our patient, the post-surgery MRI follow-up, 1 and 3 years later, showed no residual tumor. Because of the parents' wish, neither chemotherapy nor radiotherapy was used, and the patient was tumor free since the time of surgery.

Primary intracranial yolk sac tumor has an interesting pathogenesis. The current proposed theory is related to the aberrant migration of primordial germ cells. As these cells normally migrate to the primitive gonadal folds during embryogenesis, misplacement from midline may occur, giving rise to extragonadal germ cell tumors.[8]

We conclude that pathologists should be aware that primary intracranial yolk sac tumors can present at unusual anatomic sites, and such awareness will aid in the diagnosis, if encountered, in difficult cases, especially when dealing with a frozen section. Another noteworthy observation in our case is the relatively good course of disease with surgical treatment alone.

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