CASE REPORT

Primary central nervous system teratoma with sarcomatous transformation in a young girl: Report of a rare case

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ABSTRACT

A 13-year-old girl presented with chief complaints of severe headache and vomiting followed by hemiparesis. Radiological examination suggested a space occupying lesion in the right parietal lobe. Craniotomy and debulking of the tumor mass were done. Histopathological and subsequent immunohistochemical examination showed a tumor composed of fascicle of atypical spindle cells which revealed reactivity to vimentin with interspersed areas of well-differentiated cartilage tissue. Hence, the diagnosis of teratoma with sarcomatous transformation was given. Detailed discussion including review of literature has been made regarding different aspect of the tumor.

Key words: Germ cell tumor, malignancy, teratoma, sarcoma

Introduction

Teratomas constitute a group of nongerminomatous germ cell tumors (GCT) that are composed of an admixture of different tissue types representative of ectoderm, endoderm, and mesoderm. They recapitulate somatic development from the three embryonic germ layers and account for 3% of all childhood tumors with the majority occurring in the sacrococcygeal regions and the gonads.[1] The histologic picture of these tumors is strikingly similar, regardless of location. Intracranial teratomas are rare, comprising approximately 0.5% of all intracranial tumors, but incidence is slightly higher in children.[2] Teratoma with malignant transformation (TMT) is GCT which underwent malignant transformation of a somatic teratomatous component to histology that is identical to a somatic malignancy (e.g., carcinoma or sarcoma).[3] It is an extremely rare occurrence as only few incidents have been reported earlier.

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Case Report

A 13-year-old girl presented to the hospital with chief complaints of severe headache and vomiting for 1 day and gradually developing left-sided hemiparesis for the last 7 days. Following that, she developed left-sided focal seizures and drowsiness. Computed tomography scan of the brain suggested a large space occupying lesion in the right parietal lobe with midline shift [Figure 1]. The patient then underwent craniotomy, which showed a gray-white tumor mass in the right parietal lobe, and debulking of the tumor mass was done. Postoperatively, the patient was managed in the NeuroIntensive Care Unit and extubated on the next day. She was given intravenous antibiotics, dexamethasone, mannitol, and etiracetam. Then, she was shifted to high dependency unit for further management, where she remained stable and was given oral feed and physiotherapy. In the immediate postoperative period, left-sided hemiparesis persisted, but gradually, power and tone of the muscles on the left side of the body improved. On her first follow-up visit at 1-month, muscle power grade on the left side of the body was three.

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Tumor tissue was subjected to histopathological examination which showed tumor composed of densely packed long bundle of spindle cells arranged in fascicles with the presence of an area of well-differentiated cartilage tissue within the spindle cell component [Figures 2 and 3]. Spindle cells showed moderate to marked nuclear atypia along with mitotic activities [Figure 4]. Necrosis and hemorrhages were noted in the background. On immunohistochemical test, the spindle cell component showed reactivity to vimentin [Figure 5], but they were nonreactive to glial fibrillary acidic protein (GFAP), smooth muscle actin (SMA), desmin, S-100, and CD99. Hence, the diagnosis of teratoma with sarcomatous transformation was given. The patient was given etoposide- and cisplatin-based chemotherapy and radiotherapy as the part of further treatment.

Discussion

Intracranial teratomas are rare nongerminomatous GCTs. They occur primarily in children, and congenital examples are well-recognized. Teratomas account for 2% of intracranial tumors in patients younger than 15 years of age. Earlier reported cases of primary central nervous system teratoma have been depicted in Table 1. The World Health Organization classification of intracranial teratoma delineates three histological variants, namely mature, immature, and TMT.[1] TMT is generic designations for the occasional teratomatous neoplasm that contains as an additional malignant component of conventional somatic type. The latter is most often a rhabdomyosarcoma or undifferentiated sarcoma and less commonly a squamous cell carcinoma or enteric-type adenocarcinoma.[4] Yolk sac tumor elements have also been put forward as the progenitors of enteric-type adenocarcinomas arising from intracranial GCTs. Curiosities in this setting include the development of erythroleukemia, leiomyosarcoma, and carcinoid. The pathologist detecting evidence of such "malignant transformation" should state the specific histological form that this takes.[5] On immunohistochemical investigation, the constituent elements of the teratoma can be expected to express those antigens that are appropriate to their native somatic counterparts. In our case, exact differentiation of the sarcomatous component could not be ascertained as it showed only reactivity to vimentin but negative staining observed when SMA, desmin, S-100, CD99, and GFAP antibodies were used. Hence, we assumed the sarcomatous component as

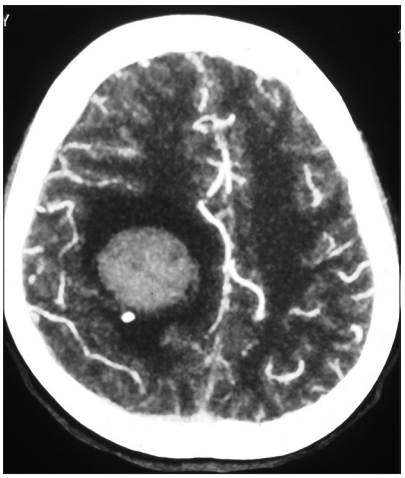

Figure 1: Computed tomography scan of the brain shows a space occupying lesion in the right parietal lobe with peri-space occupying lesion edema

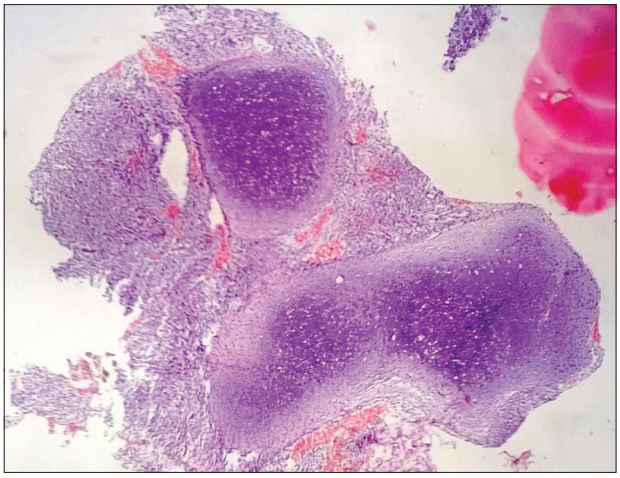

Figure 2: Scanner view showing areas of fascicular arrangement of atypical spindle cell with interspersed area of well-differentiated cartilage tissue

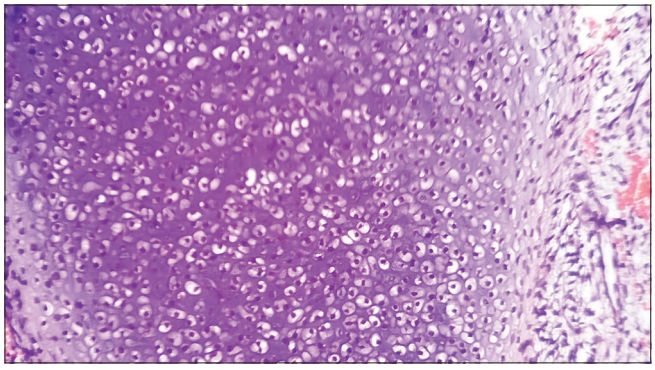

Figure 3: Well-differentiated cartilage tissue (×40)

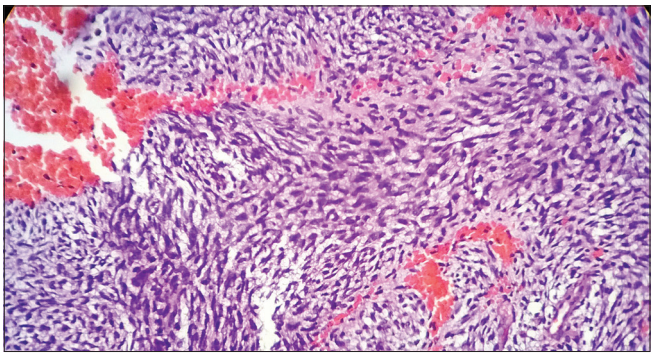

Figure 4: Atypical spindle cells arranged in fascicles and cells have high nucleus-cytoplasm ratio, hyperchromatic nuclei, and scanty cytoplasm. Background shows hemorrhage (×40)

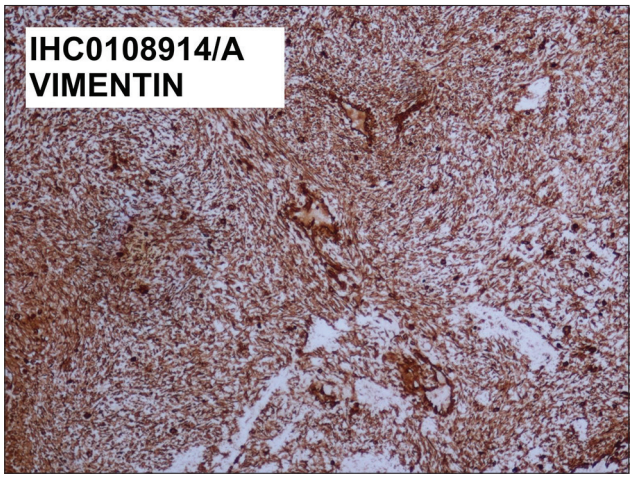

Figure 5: Spindle cells show reactivity to vimentin

Table 1: Earlier reported cases of primary central nervous system teratoma

Number of cases	Location	Reported by
34	Intracranial	1998: Sawamura *et al.*
2	Intracranial (right parietal lobe)	2015: DasGupta *et al.*
14	Intracranial (8) and intraspinal (6)	2010: Agrawal *et al.*
3	Intracranial	2009: Bare *et al.*
1	Suprasellar	2013: Sweiss *et al.*
1	Cerebral falx	2012: Zhao *et al.*
1	Intramedullary spinal	2009: Borlot *et al.*

undifferentiated one. The differential diagnosis of gliosarcoma could be excluded as the spindle cell component did not show immunoreactivity to GFAP. The histogenesis of the intracranial GCTs including teratoma remains poorly understood, but they are thought to arise from ectopic primordial germ cells, which failed to undergo apoptosis and are retained in the midline of the central nervous system. At present, a disturbance in the mechanisms that control germ cell migration appears to be the most probable cause of these tumors. The mechanisms are unknown and may include a mutation in one of the genes involved in anti-apoptotic or pro-survival pathways. Epigenetic changes that lead to aberrant gene expression and consequent protein dysfunction may also be involved. Alternatively, it was suggested that each type of GCT represents the neoplastic correlate of an embryonic stage of development so that the germinoma would derive from the misrouted primordial germ cells while teratomas derive from the embryonic differentiated cells. Another hypothesis implicates totipotential or pluripotential stem cells in the histogenesis of central nervous system (CNS) GCTs.[6-8] Within the brain, teratomas arise in the midline from optic chiasm to the pineal gland. Midline is a location with great potential for misplacement of embryonic tissues. Intracranial teratomas may arise from the pineal gland, quadrigeminal plate, and the wall of the third ventricle, suprasellar region, or cerebellar vermis. However, our case deviates the rule as it occurred in the right parietal lobe. Histological subtype is the single factor most predictive of CNS GCT outcome. Most virulent are yolk sac tumors, embryonal carcinomas, choriocarcinomas, and mixed lesions in which these subtypes are prominently represented while immature teratomas and mixed tumors dominated by teratoma or germinoma and containing high-grade nongerminomatous components in relatively limited amounts appear to occupy an intermediate position in terms of biologic potential. The historically dismal prognosis for patients with these malignant, nongerminomatous tumor subtypes has been improved with vigorous adjuvant chemotherapy strategies that continue to be investigated. While local recurrence and cerebrospinal fluid-borne dissemination are the usual patterns of disease progression, abdominal contamination via ventriculoperitoneal shunts and hematogenous spread (principally to lung and bone) may be encountered.[9]

Conclusion

Central nervous system teratoma is an uncommon tumor, and sarcomatous transformation has made the tumor extremely rare as only few cases have been reported so far.

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Department of Pathology, Chittaranjan National Cancer Institute.

Conflicts of interest

There are no conflicts of interest.

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