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
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. TMT is generic designations for the occasional teratomatous neoplasm that contains 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. 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. 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
Within the brain, teratomas

Teratoma with malignant transformation: Diverse

Intracranial (right parietal lobe)

Intracranial

2013: Sweiss

Cerebral falx

New evidence for the origin of intracranial germ cell

2010: Agrawal

Intramedullary

2009:

Intracranial

2012: Zhao

1998: Sawamura

[6‑8]

Intracranial (8) and intraspinal (6)

2015: DasGupta

Suprasellar

Reported by

2009: Bare

ed. Lyon:

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

<table>
<thead>
<tr>
<th>Number of cases</th>
<th>Location</th>
<th>Reported by</th>
</tr>
</thead>
<tbody>
<tr>
<td>34</td>
<td>Intracranial</td>
<td>1998: Sawamura et al.</td>
</tr>
<tr>
<td>2</td>
<td>Intracranial (right parietal lobe)</td>
<td>2015: DasGupta et al.</td>
</tr>
<tr>
<td>14</td>
<td>Intracranial (8) and intraspinal (6)</td>
<td>2010: Agrawal et al.</td>
</tr>
<tr>
<td>3</td>
<td>Intracranial</td>
<td>2009: Bare et al.</td>
</tr>
<tr>
<td>1</td>
<td>Suprasellar</td>
<td>2013: Sweiss et al.</td>
</tr>
<tr>
<td>2</td>
<td>Cerebral falk</td>
<td>2012: Zhao et al.</td>
</tr>
<tr>
<td>1</td>
<td>Intramedullary spinal</td>
<td>2009: Borlot et al.</td>
</tr>
</tbody>
</table>

undifferentiated one. The differential diagnosis of glosarcoma 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. 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, choriod carcinomas, 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.

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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Conflicts of interest

There are no conflicts of interest.

References