Coexistence of Radiation-induced Meningiomas and Shunt Related Pneumocephalus in a Patient with Successfully Treated Medulloblastoma

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The authors encountered a case of simultaneous radiation-induced multiple meningiomas and ventriculoperitoneal (VP) shunt-related pneumocephalus. A 35-year-old man, who had undergone surgery for medulloblastoma 21 years previously and subsequently received high-dose craniospinal irradiation with adjuvant chemotherapy and later underwent a VP shunt because of hydrocephalus, presented with a severe headache and weakness of both lower extremities. Computed tomography showed an air pocket lesion in the left temporal lobe and a large amount of pneumocephalus with a bony defect of the left tegmen tympani. In addition, a 3 cm sized well enhancing mass was noted in the in the right middle cranial fossa and additional small enhancing nodule in the left frontal pole. He was treated by left temporal craniotomy and repair of the bony and dural defects of the left tegmentum to the brain through extradural and intradural approaches, respectively. Afterwards, he underwent right temporal craniotomy and gross total removal of a rapidly growing right middle fossa mass and a left frontal mass. The histological examination was consistent with atypical meningioma, WHO grade II. In conclusion, physicians have to consider the serious long term complications of high dose radiation therapy and VP shunt placement and need to perform the neuroradiologic follow-up after such treatments for several decades.

KEY WORDS: Medulloblastoma · Radiation induced · Meningioma · Ventriculoperitoneal shunt · Pneumocephalus · Tegmen tympani.

Introduction

The induction of secondary neoplasia after conventional high-dose radiation is a well known long-term complication. In particular, radiation-induced meningiomas are at least five times more numerous than gliomas or sarcomas in the world literature. Twenty-six cases of radiation-induced meningiomas in patients previously treated for medulloblastoma (MBL) have been reported.

Delayed and spontaneous (non-traumatic) pneumocephalus after a ventriculoperitoneal (VP) shunt operation are also rare complications; only about forty cases have been reported. Various treatment modalities have been used to address this complication, i.e., shunt externalization or obliteration, shunt revision, direct leakage point closure and observation.

The authors describe a case of successfully treated cerebellar MBL with high-dose radiation that twenty one years later developed multiple atypical meningiomas and VP shunt related pneumocephalus.

Case Report

A 35-year-old man underwent surgical resection of a posterior fossa MBL at the age of 14 years in 1983. He subsequently received high-dose craniospinal irradiation (5700 cGy) followed by adjuvant chemotherapy. Fourteen years later, he underwent VP shunt placement because of delayed hydrocephalus, which had presented with severe headache and slowly aggravating visual disturbance in 1997.

Seven years later, he presented with severe headache and gait disturbance due to weakness of both lower extremities. Brain and temporal bone CT showed a cystic air pocket lesion in the left temporal lobe and a large amount of pneumocephalus associated with the bony defect of the left...
Nodule four months after the removal of the right temporal mass. Both pathologies were turned out to be atypical meningioma, WHO grade II (Fig. 4).

Discussion

High-dose radiation-induced meningiomas

The induction of secondary intracranial neoplasia is a rare complication of radiation therapy (RT) for MBL, and only 26 cases have been reported (Table 1). These cases comprise 13 male patients and 13 female patients. The median age of these patients at RT for MBL, including the present case, was 5 years (range 0.7-15 years), mean age at diagnosis of radiation-induced meningioma was 22 years (range 6.7-41 years). These findings support the previously held opinion that patients who develop radiation-induced meningiomas are younger than those who develop sporadic meningiomas in the fifth and sixth decades of life.29 The median latency period between the commencement of RT for MBL and clinical onset of meningioma was 16 years (range 2.2-27 years). We found no statistically significant correlation between patient age and latency time (P=0.301).

Salvati et al. described following characteristics of the development of high-dose radiation-induced meningioma; (1) children appear particularly sensitive to the development of this tumor; (2) there is a greater female predominance; (3) these tumors present a peak frequency in the third decade of life; and (4) frequently these tumors are atypical subtype and having the tendency of recurrence. With the exception of a female predominance, high-dose radiation-induced meningiomas including our case have these characteristics.

This case fulfils all the criteria required for radiation-induced meningioma, i.e., occurrence within a previously administered irradiation field. The meningioma was not present prior to the first irradiation, and its histology differed from that of the tumor which prompted the original radiotherapy. Moreover, there was a 21-year latent period between the first irradiation and the appearance of the radiation-induced tumor, and the

![Fig. 1. Brain computed tomography (CT) showing a cystic air pocket lesion in the left temporal lobe (A) and a large amount of pneumocephalus (B). Temporal bone CT demonstrates this left tegmentum tympanum bony defect which caused the pneumocephalus in this case (C).](image1)

![Fig. 2. Gross photographs depicting direct repair of the bony defect by packing with muscle fragments through an extradural approach (A, B) and of the left tegmentum tympanum dural defect with surgical glue (Tisseal) through an intradural approach (C).](image2)
Table 1. Literature review of radiation-induced meningiomas after high-dose radiotherapy for medulloblastoma

<table>
<thead>
<tr>
<th>No</th>
<th>Authors &amp; Year</th>
<th>Sex</th>
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Fig. 3. Brain magnetic resonance (MR) images show a 3 cm-sized well enhanced round mass in the right middle cranial fossa (A), Follow up brain MR images show an increased size of the right middle cranial fossa mass (B) and the left frontal convexity mass (C).

easily, we believe that surgical removal is the treatment of choice in most cases. However, for small-sized atypical meningiomas, Gamma knife radiosurgery may be an alternative treatment modality.<sup>8</sup>

**Ventriculoperitoneal shunt related tension pneumocephalus**

Most cases of pneumocephalus are caused by fractures of the skull base, sinus and mastoid air cells, tumors of the skull base, infectious conditions of the skull base or the congenital defects of the skull base.<sup>33</sup> Pneumocephalus is thought to be caused by two mechanisms, both involving low intracranial pressure that "sucks" air in through a bony or dural defect. The first involves a vertical posture, which creates a pressure gradient within the cerebrospinal fluid (CSF) system, whereas the second involves a ball valve effect with air entering through a fistula, which is then tamponaded with brain tissue<sup>14,27</sup>.

The low intracranial pressure caused by a VP shunt and leakage point caused by a bony or dural defect might play causative role in VP shunt related pneumocephalus. Therefore, localization and repair of the leakage point are indispensable for treating VP shunt related pneumocephalus<sup>6,31</sup>.

Based on these arguments, we summarize the treatment protocol proposal for VP shunt related pneumocephalus in Fig. 5. High resolution and thin section CT findings and repairing the leakage point are more important than externalizing shunt devices, titrating CSF drainage, or removing of them. It is noticeable that the leakage points could be easily produced by bony erosion due to a chronically increased intracranial pressure and a thinned CSF barrier, which could be broken easily by sneeze or light exercise in the absence of major trauma or infectious condition<sup>14,27</sup>.
Conclusion

Both high-dose radiation-induced meningioma after MBL treatment and VP shunt related pneumocephalus are rare delayed complications. The localization and direct surgical repair of leakage points is a treatment of choice for shunt related pneumocephalus, surgical removal of radiation-induced secondary meningioma is also a treatment of choice. Neurosurgeons must take into consideration the serious potential long term complications of high dose radiation therapy and VP shunt placement, and need to undertake neuroradiologic surveillance after these treatments for several decades.

References