Cerebral Tumor with Rhabdoid Features

19

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Contents

Abstract

Rhabdoid tumors have been reported to arise from various locations such as the central nervous system, soft tissues, liver, retroperitoneum, pelvis, and skin, in addition to the kidneys. These tumors with rhabdoid cell components have been reported to be aggressive lesions. Atypical teratoid/rhabdoid tumors (AT/RT) are the most common cerebral tumors with rhabdoid features. Recently, new disease categories, such as rhabdoid meningioma and rhabdoid glioblastoma, have been reported. These tumors are difficult to distinguish radiologically. Histological and immunohistological examinations are useful for making an accurate diagnosis. Recent investigations have revealed that the lack of INI-1 gene expression is the most characteristic feature in AT/RT and malignant rhabdoid tumor. We recently reported a frontal brain tumor with rhabdoid features that could not be categorized in any of the current classifications, and referred to this tumor as a cerebral tumor with extensive rhabdoid features. We present several types of cerebral tumors with rhabdoid features and discuss the differences among them.

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Introduction

Malignant rhabdoid tumor (MRT) was first described as a subtype of Wilms' tumor of the kidney that presented with rhabdomyosarcomatoid features (Beckwith and Palmer 1978).

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However, ultrastructural studies revealed that these tumor cells had not differentiated into rhabdomyoblasts (Fung et al. 1981). Rhabdoid tumors have been reported to arise from various locations: the central nervous system (CNS), soft tissues, liver, retroperitoneum, pelvis, and skin, in addition to the kidneys (Parham et al. 1994). In the CNS, such lesions are commonly called atypical teratoid/rhabdoid tumors (AT/RT); however, some authors differentiate between AT/RT and MRT (Tekkök and Sav 2005). Recently, several cerebral tumors with rhabdoid features other than those of AT/RT or MRT have been reported, such as rhabdoid meningioma and rhabdoid glioblastoma (Perry et al. 1998; Wyatt-Ashmead et al. 2001). These tumors contain various populations of rhabdoid cells, which have eccentrically placed nuclei containing vesicular chromatin, prominent eosinophilic nucleoli, and abundant cytoplasm with eosinophilic globular cytoplasmic inclusions. Genetic studies have revealed that the deletion of chromosome 22q and the inactivation of the INI1/hSNF5 tumor suppressor gene on chromosome 22q11.2 were characteristic features in AT/RT and MRT (Makuria et al. 2008; Biswas et al. 2009; Tekkök and Sav 2005). As described above, new tumor entities with rhabdoid features have been reported, and a new tumor that cannot be categorized using the current classifications has been found. In this chapter, we describe several types of cerebral tumors with rhabdoid features and report our case with a lesion that we have referred as a "cerebral tumor with extensive rhabdoid features", and discuss the differences among them.

Atypical Teratoid Rhabdoid Tumor

Atypical teratoid rhabdoid tumor (AT/RT) is a rare, highly malignant tumor that accounts for 1.3% of all primary pediatric tumors of the CNS (Rickert and Paulus 2001). AT/RT is defined as typically containing rhabdoid cells, often with primitive neuroectodermal cells and with differentiation along mesenchymal, epithelial lines (Rorke et al. 1996). It usually occurs in children under the age of 5 years, although several adult cases have also been reported (Mirone et al. 2009; Raisanen et al. 2005). AT/RT usually occurs in the posterior fossa. Dang et al. (2003) reported that 52% of these tumors were located in the posterior fossa and 39% were supratentorial. AT/RT tend to occur off-midline and to have calcification and cyst formation (Biswas et al. 2009; Lee et al. 2009). The solid component of the tumors had a homogeneous iso-intensity on T1 and T2-weighted images in one series, whereas, another study showed that more than half of the tumors had mixed intensities on T1 and T2-weighted images. Moderate to strong enhancement was found in most cases (Lee et al. 2009; Warmuth-Metz et al. 2008). A band-like rim of the central cyst or necrotic area was strongly enhanced in some cases. Leptomeningeal seeding was often detected (Lee et al. 2009). The optimal treatment has not been elucidated. Because of the young age of the patients, high-dose radiation therapy is not suitable. Various chemotherapeutic regimens have been reported (Biswas et al. 2009). Rorke et al. (1996) reported that the median time-to-progression and overall survival were 4.5 and 6 months, respectively. A recent study showed that the chemotherapeutic response and radiation therapeutic response were 58 and 38%, respectively, and that the 2-year progression-free and overall survival rates were 53 and 70% respectively (Chi et al. 2009).

Histologically, AT/RT have varying proportions of rhabdoid cells. Rhabdoid cells are characterized by abundant eosinophilic cytoplasm, globular inclusion, and an eccentrically placed vesicular nucleus with open chromatin and prominent nucleolus. Rhabdoid cells exist often with undifferentiated PNET component consisting of small cells with a high nuclear/cytoplasmic ratio and oval hyperchromatic nuclei and with differentiation along mesenchymal, epithelial lines (Mohapatra et al. 2010). Immunohistologically, AT/RT is usually positive for epithelial membrane antigen (EMA), smooth muscle actin (SMA), and vimentin and occasionally positive for synaptophysin, cytokeratin, glial fibrillary acidic protein (GFAP) and neurofilament (Mohapatra et al. 2010; Rorke et al. 1996). The MIB-1 index is relatively high, and the mean MIB-1 index was

45% in one series (Mohapatra et al. 2010). Electron microscopic examination revealed whorled bundles of intermediate filaments filling some or most of the perikaryon (Rorke et al. 1996). The characteristic genomic feature seen in AT/RT is the inactivation of the INI-1 gene (Haberler et al. 2006). Monosomy or deletion of chromosome 22 is detected in 75–90% of AT/RT lesions (Bruch et al. 2001; Raisanen et al. 2005).

MRT of the central nervous system was first reported by Briner et al. (1985). The concept of AT/RT was constructed by Rorke et al. in 1996; however, some authors have reported that MRT is not the same disease as AT/RT (Tekkök and Sav 2005). According to Tekkök's review, local recurrence or subarachnoid spread were reported in more than two-thirds of patients after a mean period of 6.9 months, and these patients died after a mean period of 8.9 months after their diagnosis (Tekkök and Sav 2005). MRT is characterized by medium to large round or polygonal cells with prominent eosinophilic cytoplasm and eccentric and round nuclei with prominent nucleoli. MRT is usually immunoreactive for vimentin, EMA and cytokeratin and is often immunopositive for SMA, S-100, and synaptophysin. MRT shows the deletion of region 11.2 of the long arm of chromosome 22 (22q) and the inactivation of the INI1/hSNF5 tumor suppression gene (Tekkök and Sav 2005; Mirone et al. 2009).

Most cases of AT/RT (and primary CNS MRT) occur in children; however, several adult cases have been reported (Mirone et al. 2009; Raisanen et al. 2005). Adult and childhood AT/RTs have been shown to have similar cytologic, architectural, and genetic features (Raisanen et al. 2005). Mirone et al. reviewed 21 cases of cerebral adult AT/RTs (including MRTs) and reported a mean survival time of 38 months. Although leptomeningeal dissemination is also common in adults, adult cases seem to have a relatively good prognosis. This difference may be due to the greater radicality of surgical resection for the tumor and higher-dose radiation therapy, which cannot be tolerated by children. Some cases reportedly exhibited a normal chromosome 22q (Mirone et al. 2009).

Rhabdoid Meningioma

Rhabdoid meningioma was first described in 1998 (Kepes et al. 1998). Rhabdoid meningioma is classified as a WHO grade 3 meningioma and exhibits aggressive progression with a high rate of proliferation (Kim et al. 2007). Two series have been published (Perry et al. 1998; Kim et al. 2007). Perry's study included eight men and seven women with a median age of 50 years (range, 13–73 years) at the time of first surgery and 53 years (range, 13–73 years) at the time that the rhabdoid features became apparent. Kim's study included four men and 11 women with a mean age of 52 years (range, 22–75 years). Rhabdoid meningioma tends to have cystic components, prominent peritumoral edema, and bone involvement (Kim et al. 2007). In Perry's study, 87% of the patients had recurrence, 13% had extracranial metastasis, and 53% died (one patient died from pulmonary embolism on postoperative day 2). The median time until death was 5.8 years after the initial operation and 3.1 years after the first appearance of rhabdoid morphology (Perry et al. 1998). In another study, additional radiation therapy was performed after the operation in all the cases except one case, and only one recurrence (7%) and no extracranial metastases or deaths were found during a follow-up period ranging from 11 to 39 months (Kim et al. 2007).

Rhabdoid meningioma is characterized by loosely cohesive sheets of cells with abundant eosinophilic cytoplasm, eccentric nuclei and hyaline, and frequent fibrillar paranuclear inclusion (Perry et al. 1998). The components of conventional meningiomas were present in the majority of tumors, and rhabdoid components often became increasingly prominent with subsequent recurrence (Perry et al. 1998). Electron microscope revealed paranuclear whorls of intermediate filaments, frequently with entrapped lysosomes and mitochondria (Perry et al. 1998). The MIB1 index ranged from 0.8 to 43% (median, 6.3%) (Perry et al. 1998). The tumor cells are immunopositive for vimentin and EMA and are usually

negative for GFAP. The deletion of 22q was identified in 10 of the 14 (71%) cases, and nuclear INI-1 expression was retained in all 16 cases (Perry et al. 2005).

Rhabdoid Glioblastoma

Rhabdoid glioblastoma is a recently reported entity in which an epithelioid glioblastoma is associated with a rhabdoid component (Fung et al. 2004; Kleinschmidt-Demasters et al. 2010; Lath et al. 2003; Wyatt-Ashmead et al. 2001). The typical radiological pattern is a ring-enhanced mass with extensive edema, similar to a conventional glioblastoma. Glioblastoma is one of the most malignant brain tumors. Rhabdoid glioblastoma is highly aggressive with a potential for early recurrence, and patients usually die within several months (Fung et al. 2004; Kleinschmidt-Demasters et al. 2010; Lath et al. 2003; Wyatt-Ashmead et al. 2001). Whether rhabdoid glioblastomas should be treated with surgery followed by aggressive multiagent chemotherapy and cranio-spinal radiation, like other malignant rhabdoid tumors, or treated like typical glioblastomas remains uncertain (Lath et al. 2003). Tumor cells with the rhabdoid phenotype are strong immunostained with vimentin and EMA, and a subpopulation is GFAP immunopositive. The glial component shows prominent mitotic activity and extensive tumor necrosis with GFAP immunopositivity (Fung et al. 2004; Kleinschmidt-Demasters et al. 2010; Lath et al. 2003; Wyatt-Ashmead et al. 2001). The MIB-1 index ranged from 18 to over 30%. Ultrastructurally, the tumor cells contain cytoplasmic whorls of intermediate filaments (Wyatt-Ashmead et al. 2001). Monosomy of 22 and polysomy of 22q were found in two and one cases, respectively, in three examined cases (Kleinschmidt-Demasters et al. 2010; Wyatt-Ashmead et al. 2001). The expression of INI -1 was found in the nucleus of glioblastoma cells without rhabdoid features, but not in a subset of cells with the strongest rhabdoid features (Kleinschmidt-Demasters et al. 2010).

Rhabdoid Melanoma

Rhabdoid melanoma was first described in 1992 (Bittesini et al. 1992). The extent of the rhabdoid changes in the neoplastic cells vary. In a large series of Chang et al., 48% of the samples were composed exclusively of rhabdoid cells, whereas in 52% of them, the rhabdoid cells constituted less than 25% of the tumor (Chang et al. 1994). The presence of rhabdoid features in melanoma is more common in metastatic melanoma than in primary lesions (Magro et al. 2006). Chang et al. reviewed 31 specimens from 29 patients with metastatic melanoma with rhabdoid features. Seventeen patients were men and 12 patients were women, ranging in age from 24 to 78 years. Preexisting cutaneous melanomas had been observed in 25 cases (Chang et al. 1994). Rhabdoid melanoma shows significant histologic, immunohistochemical and ultrastructural heterogeneity (Gavino and Gillies 2008). It is immunopositive for vimentin, but has diverse immunoreactivity patterns for S-100 and frequently loses HMB-45 expression (Magro et al. 2006; Gavino and Gillies 2008). Rhabdoid melanoma is less immunoreactive for S-100 protein than nonrhabdoid melanoma. No difference in staining with HMB-45 has been noted between rhabdoid melanoma and nonrhabdoid melanoma (Chang et al. 1994). The loss of S-100 and /or HMB-45 expression in the cells of rhabdoid melanoma is thought to be a manifestation of the process of dedifferentiation (Gavino and Gillies 2008). Rhabdoid melanoma appears to have a no more aggressive behavior than conventional melanoma (Gavino and Gillies 2008).

Metastatic Tumor from Carcinoma with Rhabdoid Features

Some carcinomas with rhabdoid features include lung tumors, stomach, jejunum, colon, and so on (Tamboli et al. 2004; Amrikachi et al. 2002). Metastatic cerebral tumors of these primary lesions are different diagnoses of cerebral tumors with rhabdoid features. Tamboli et al. (2004)

reviewed 32 patients with rhabdoid lung tumors who ranged in age from 25 to 82 years. There were 20 men and 12 women. The amount of rhabdoid cells ranged from 10 to 90%. Rhabdoid lung tumors are usually immunopositive for vimentin and epithelial markers. After a mean follow-up period of 8.3 months, 60% of the patients had died, with a mean survival period of 8.1 months (Tamboli et al. 2004). Amrikachi et al. (2002) reviewed 16 cases of gastrointestinal rhabdoid tumors; the patients in this series were 11 men and five women, ranging in age from 52 to 84 years. All the cases were immunopositive for vimentin, and cytokeratin or EMA was positive in 15 of the 16 cases. Electron microscopy showed whorls of intermediate filaments in esophagus, stomach and small intestine tumors (Amrikachi et al. 2002). Seventy five percent of the cases died within 10 months, and 50% died within 3 months (Amrikachi et al. 2002). The prognosis of tumors with rhabdoid features was reported to be poor, independent of their locations (Tamboli et al. 2004; Amrikachi et al. 2002).

Cerebral Tumor with Extensive Rhabdoid Features

We recently reported a 32-year-old pregnant female with a frontal tumor (Ohba et al. 2009). Systemic examinations revealed no other lesions. The tumor was hypercellular and contained a diffuse sheet of various sizes of eosinophilic cells. These cells had a few prominent, eccentrically placed, variously sized, hyperchromatic nuclei. The tumor was immunohistologically reactive for vimentin, EMA, SMA, and BAF47/INI-1 and negative for GFAP, neurofilament protein, S-100, and HMB-45. The MIB-1 index was 4.2%. The deletion of 22q was not detected by the CGH analysis. Chemotherapy and radiotherapy were administered, and the patient is still alive after more than 7 years. Radiological examinations have lowered the possibility of undetected metastatic tumors and rhabdoid meningiomas. Histologically, this tumor had no components containing PNET-like cells, meningiomas, glioblastomas, melanomas, or carcinomas of other origins. Immunohistologically, rhabdoid glioblastoma and AT/RT (MRT) were excluded. The tumor could not be characterized using the current classification. We therefore referred to this tumor as a "cerebral tumor with extensive rhabdoid features".

Discussion

Tumors with rhabdoid features are uncommon; however, rhabdoid cells are found in several types of tumors. In addition to histological examination, immunohistological examination and genetic investigation are often useful to make an accurate diagnosis. In this paper, we described the clinical, radiological, histopathological, and genetic features of several tumors with rhabdoid features and discuss the differences among them.

Radiological Features

It is very difficult to distinguish a tumor with rhabdoid features from other tumors with rhabdoid features using radiology because cerebral tumors with rhabdoid features generally have similar radiological features: cyst formation, homo to heterogeneous images, moderate to strong enhancement, ring enhancement, and remarkable edema. The presence of calcification may help to distinguish AT/RT and rhabdoid meningioma from other lesions. Leptomeningeal dissemination is a key feature associated with AT/RT (Lee et al. 2009; Tekkök and Sav 2005). Rhabdoid meningioma is an extra-axial tumor that is usually attached to the dura mater and sometimes invades bone tissue, unlike the other tumors described here. The presence of other primary lesions is often useful for diagnosing metastatic tumors.

Histological Features

Rhabdoid cells are characterized by abundant eosinophilic cytoplasm, globular inclusion, and an eccentrically placed vesicular nucleus with open chromatin and prominent nucleolus. The proportion of rhabdoid cells varies even among

lesions of the same disease type. In AT/RT, rhabdoid cells often exist with an undifferentiated PNET component and with differentiation along mesenchymal, epithelial lines (Mohapatra et al. 2010). Many tumors with rhabdoid features have an original neoplastic cell component in addition to the rhabdoid cells. Rhabdoid glioblastoma has the classical histological features of a high-grade astrocytoma (Kleinschmidt-Demasters et al. 2010). A conventional meningioma component is present in the majority of rhabdoid meningiomas (Perry et al. 1998). Rhabdoid melanoma usually retains melanocytic attributes (Chang et al. 1994). Other carcinomas with rhabdoid features are usually composed of rhabdoid cells and carcinomatous neoplasm (Tamboli et al. 2004). The lesion in our reported case consisted of only rhabdoid cells, without other variable components.

Immunohistochemical Features

Tumors with rhabdoid features are usually immunopositive for vimentin and EMA (Mohapatra et al. 2010; Perry et al. 1998; Rorke et al. 1996). AT/RT (and MRT) are usually positive for EMA, SMA, and vimentin and are occasionally positive for synaptophysin, cytokeratin, GFAP and neurofilaments (Mohapatra et al. 2010; Rorke et al. 1996). Rhabdoid meningioma is immunonegative for GFAP (Perry et al. 1998; Kepes et al. 1998). The glial component of rhabdoid glioblastoma is immunopositive for GFAP (Lath et al. 2003). Rhabdoid melanoma shows diverse patterns of immunoreactivity to S-100 and frequently loses HMB-45 expression. Our case was immunohistologically reactive for vimentin and EMA and negative for GFAP, neurofilament protein, S-100, and HMB-45. The MIB-1 index is usually remarkably high in tumors with rhabdoid features and moderately high in rhabdoid meningioma but was not so high in our case.

Ultrastructure

Ultrastructurally, rhabdoid tumors show three patterns. The most common pattern is that of whorled filamentous bodies with entrapped lipid and organelles. The second pattern is that of intermediate filaments packed in elongated, gently curved bundles. The third form is characterized by a filamentous pattern composed of twisted sheaves of intermediate filaments, resembling tonofilaments of squamous epithelial cells (Gavino and Gillies 2008; Haas et al. 1981).

Chromosome 22 and INI-1 Expression

The loss of INI-1 expression is the most characteristic feature of AT/RT and MRT (renal and extra-renal). Monosomy or deletion of chromosome 22 is detected in 75–90% of AT/RT (Bruch et al. 2001; Raisanen et al. 2005), and all AT/RT cases stain negatively for INI-1 protein (Biswas et al. 2009; Mohapatra et al. 2010). Neoplasms with rhabdoid features that are not associated with the suppression of INI-1 protein or gene include carcinomas, melanomas, meningiomas, and gliomas (Perry et al. 2005). All the cases of rhabdoid meningioma showed the INI-1 expression. The expression of INI-1 was found in the nuclei of glioblastoma cells without rhabdoid features, but not in a subset of cells with the most rhabdoid features (Kleinschmidt-Demasters et al. 2010). INI-1 expression was detected in composite rhabdoid tumors, such as melanoma and carcinoma (Perry et al. 2005), as well as in our case. Perry et al. (2005) suggested that composite rhabdoid tumors were genetically distinct from MRT and AT/RT and retained INI-1 expression.

Treatment

The optimal treatment for AT/RT has not been elucidated. If possible, surgery followed by chemotherapy and radiotherapy are often performed. Because most patients are young, there is reluctance to perform radiation therapy. Surgery with additional radiation therapy is recommended for rhabdoid meningioma. Whether rhabdoid glioblastomas should be treated like other MRTs or treated like typical glioblastomas remains uncertain (Lath et al. 2003). In our case, radiation

therapy and chemotherapy were performed after tumor removal, and these treatments appeared to be effective (Ohba et al. 2009).

Prognosis

Tumors with rhabdoid cell components have been reported to be aggressive lesions. Rhabdoid meningioma is classified as WHO grade 3, whereas conventional meningioma is WHO grade 1. Glioblastoma and malignant melanoma are potentially aggressive. The occurrence of metastasis to the brain from a primary carcinoma represents a severe disease stage. Although the prognosis of AT/RT has been reported to be poor (Rorke et al. 1996), a recent study showed a better outcome than that of previous reports (Chi et al. 2009). Compared with them, our case responded well to chemotherapy and radiation therapy and had a good outcome (Ohba et al. 2009). The MIB1 index of this case was not high, which might explain the good outcome. The factors that define the prognosis remain uncertain, and further investigations are needed to identify these factors.

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