CASE REPORT

Multifocal desmoplastic noninfantile astrocytoma

Astrocytome non infantile desmoplastique multifocal

K. Santhosha, C. Kesavadas, V.V. Radhakrishnan, M. Abraham, A.K. Gupta

Department of Imaging Sciences and Interventional Radiology, Sree Chitra Tirunal Institute for Medical Sciences and Technology, Trivandrum 695011, India
Department of Pathology, Sree Chitra Tirunal Institute for Medical Sciences and Technology, Trivandrum 695011, India
Department of Neurosurgery, Sree Chitra Tirunal Institute for Medical Sciences and Technology, Trivandrum 695011, India

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Summary
This is a report of a case of multifocal desmoplastic astrocytoma in an 11-year-old child in which we describe the MRI findings and discuss the possible mechanism of its development. The MRI appearances in our case support the view that the tumor is primarily of leptomeningeal or superficial cortical origin, with cystic formation secondary to entrapment of cerebrospinal fluid. The question of whether or not the lesions are metastases or metachronous lesions is also discussed. Desmoplastic astrocytoma at a noninfantile age is extremely rare: only four cases have been reported in the literature so far. Even more unusual is the presence of this lesion in multiple locations at the initial presentation.

Introduction

Desmoplastic infantile astrocytoma (DIA), a tumor originally described in infants, can occur in older children and young adults and is then referred to as the noninfantile variant [1]. Desmoplastic infantile ganglioglioma (DIG) and DIA are...
similar-looking rare tumors that involve the superficial cerebral cortex and leptomeninges, with variable attachment to the dura and are considered grade I in the World Health Organization (WHO) classification system [2]. In spite of this grading, craniospinal metastases have been reported with DIG/DIA in the literature [3]. This is a report of the findings in a series of MRI scans of a rare case of desmoplastic noninfantile astrocytoma presenting with multiple craniospinal lesions.

**Case report**

A 11-year-old boy was admitted to our hospital in January 2007 complaining of gradually progressing stiffness in both legs, neck pain and visual blackouts, lasting one month. Neurological examination was unremarkable except for wasting of the thenar and hypothenar muscles. MRI showed dilated ventricles, a prominent right middle frontal sulcus and a well-defined cystic lesion in the left anterior frontal cortex (Fig. 1). Cystic areas were hyperintense in comparison to cerebrospinal fluid (CSF) on T2-weighted (T2W) imaging and were not suppressed by fluid-attenuated inversion-recovery (FLAIR) MRI. Solid regions were hypointense on T1-weighted (T1W) imaging and hyperintense on T2W. A small intramural nodule at the periphery of the cyst appeared hyperintense compared with gray matter on T2W and FLAIR sequences, and isointense on T1W sequences, probably representing calcification. Multiple meningeal-based, small cystic lesions were seen in the infratentorial fossa. A contrast study revealed a thickened tentorium, however, cysts do not show any enhancement at this time. MRI examination of the spinal cord showed cystic and solid lesions in the cervicothoracic spinal cord. The cystic areas appeared hyperintense on T2W and isointense on T1W, whereas the solid areas were mildly hyperintense on T2W and isointense on T1W, with mild patchy enhancement (Fig. 1g and h). The patient underwent ventriculoperitoneal shunting for the hydrocephalus, after which his symptoms improved. CSF studies, including PCR for tuberculosis, India ink for Cryptococcus and malignant cytology, were all negative. As the patient was neurologically stable, he was managed conservatively and advised to undergo regular follow-up.

Eight months later, the patient presented again with two episodes of left-sided focal seizures. Examination revealed bilateral optic atrophy, a left upper-motor-neuron type of facial palsy and left hypoglossal nerve palsy. Muscle tone was increased in the left upper limb and both lower limbs, resulting in exaggerated reflexes. There were neither sensory abnormalities nor cerebellar signs. Routine laboratory investigations and CSF studies were normal. Repeat MRI revealed large heterogeneous solid and cystic lesions in the right frontal lobe, with mass effects and a midline shift (Fig. 2). The mass was located at the same site as the earlier sulcal prominence, suggesting that the dilated sulcus probably represented an early lesion that grew over time into a large tumor. The cyst in the left frontal cortex showed a mild increase in size compared with the earlier MRI examination.
Figure 2  Second MRI taken after eight months: axial T2- (a), T1- (b) and FLAIR- (c) weighted images show a large, multilocular cystic mass with peripheral meningeal-based solid portions and a mass effect on the adjacent ventricle. Axial sections at a higher plane show the left frontal cystic lesion. Solid areas appear hyperintense on T2- (d) and T1- (e) weighted images, and are probably calcifications. The volume of the lesion appears to have grown compared with the previous MRI. There is no diffusion restriction on 1000-b-weighted images (f) or on apparent diffusion coefficient (ADC) mapping (g). Small cystic lesions similar to those in the previous MRI are evident in the posterior fossa on T2-weighted axial imaging (h).

Figure 3  Second MRI after eight months: a GRE T2* axial image (a) shows a hypointense nodule on the medial side of the cyst suggestive of calcification. Contrast-enhanced T1-weighted images show enhancement of the tentorium (b), and of the peripheral solid areas close to the meningeal spaces (c and d). MR spectroscopy (TE: 135 ms) shows an elevated choline peak, a reduced NAA peak and a lactate peak.

Figure 3  IRM de contrôle réalisée à huit mois : l'image axiale pondérée T2* (a) montre un nodule hypo-intense à la partie médiale du kyste évoquant une calcification. Les images pondérées T1 après injection montrent un rehaussement de la tente (b) et des zones solides périphériques au contact des espaces méningés (c et d). La spectroscopie RM (TE: 135 ms) montre un pic élevé de choline, un pic diminué de NAA et un pic de lactate.
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There was a small hypointense nodule in the left frontal lobe cystic lesion on gradient-recalled-echo (GRE) images suggestive of calcification. The cystic and solid areas showed no diffusion restriction and the lesions in the infratentorial fossa showed no progression. However, the lesions did show peripheral meningeal-based enhancement of the solid component with adjacent dural thickening. Spectroscopy revealed elevated choline with reduced N-acetyl aspartate (NAA) and a small lactate peak (Fig. 3). The spinal cord lesion had increased in size and T1W imaging showed focal hyperintensity within the lesion. The solid areas showed heterogeneous enhancement. The spinal lesions also showed a tendency to be eccentrically placed (Fig. 4).

The patient underwent right pterional craniotomy and left frontal craniotomy; subtotal decompression of the right frontal lesion and cyst decompression of the left frontal lesion were performed. The spinal cord lesion was left untreated, as there was no progression in the patient’s symptoms. Histopathological examination of the excised tissue revealed densely packed tumor cells interlaced with dense collagen. Tumor cells were positive for reticulin and glial fibrillary acidic protein (GFAP). Ganglionic cells were not identified. The histological diagnosis was desmoplastic variant of astrocytoma (Fig. 5). The patient showed significant improvement postoperatively and neither chemotherapy nor radiotherapy was necessary.

Discussion

Desmoplastic infantile astrocytoma is a rare brain neoplasm that was first described by Taratuto et al. [1] DIG/DIA is generally seen in infants aged under 18 months, with a mean age at diagnosis of five to six months [2,3] However, the noninfantile variant of desmoplastic ganglioglioma is increasingly being reported in the literature [4,5], with these patients ranging from 5 to 25 years of age. The imaging findings and the prognosis for both infantile and noninfantile tumors are often similar [5]

Only four cases of DIA involving the noninfantile age group have been reported in the literature [6—9] In two of these cases, the patients had seizures since infancy and the tumor was diagnosed later; hence, strictly speaking, these cases cannot be described as being noninfantile variant [6,8] In the two remaining cases, the tumors were detected in childhood [7,9]

Desmoplastic tumors are often large, slow-growing tumors that preferentially involve the temporal and parietal lobes, but with a tendency towards multilobar involvement. They present as large, superficially located cystic masses, usually arising in the supratentorial compartment. The cystic component can be deeply lying and uni- or multilocular, with solid portions evident in the periphery of the lesion. The solid portion lies adjacent to the meninges and is often attached to the dura and is enhanced along with the thickened meninges. The cyst wall usually is not enhanced with contrast and there is minimal perilesional edema. Tumors with a predominantly solid component or exophytic component have also been described [10,11] Calcifications are noted in 50% of cases and bony abnormalities such as bossing, thinning or deformation may be evident [3] The radiological differential diagnoses include pleomorphic xanthoastrocytoma, primitive neuroectodermal tumors (PNETs), ependymoma and ganglioglioma [3] Histologically, the tumor is mainly composed of neoplastic astrocytes and prominent reticulin-rich, desmoplastic stroma, with a variable extent of neuronal differentiation. Astrocytes are the prominent neuroepithelial component and are prominent in desmoplastic areas. These tumors
are considered grade I, according to the WHO classification system [5].

Most DIG/DIA are associated with a good prognosis after gross surgical excision [12], although several reports have described metastases as well as the aggressive nature of this tumor [13, 14]. To date, a total of six cases of DIG with metastases have been described in the literature. In all of these cases except one, metastases arose after the initial diagnosis; only in the one exception were metastases seen at the time of presentation. Death due to metastases was reported in two of these cases, where the tumors were located in the suprasellar cistern/hypothalamus [13, 14]. However, the occurrence of metastases has not been reported in the noninfantile type of DIA/DIG, making ours the first case report of a noninfantile variant of DIA presenting with metastases/multifocal lesions at the time of diagnosis. Nevertheless, all cases (where the outcome was specified) had a favorable prognosis with no deaths reported, including our case [4, 13]. However, reports of DIG/DIA with metastases and mentioning the long-term follow-up are lacking.

Serial MRI appearances showing transformation of DIG from a solid tumor into a typical cystic lesion have been described [15]. Serial MRI in our case can throw some light on the mechanism of development of this tumor. The lesion located in the right frontal region appeared initially as diffusely thickened meninges over the frontotemporal region, with a prominent middle frontal sulcus. Neither a solid nor cystic mass was identified at this time. Serial imaging showed a large multicystic mass in the frontal lobe with thickened meninges typical of DIG. It has been postulated that the DIA arises from the subpial astrocytes that produces basal lamina in normal brain [16]. The serial MRI appearances in our case support the notion that the tumor is primarily of leptomeningeal or superficial cortical origin, with cystic formation secondary to entrapment of CSF due to some sort of valve-checking mechanism, as described by Taguchi et al. [17].

Multiple lesions in the brain as well as spinal cord could represent either metachronous or metastatic lesions. The right frontal lesion appeared initially as a focally dilated sulcus, which subsequently evolved into a large cystic-solid mass typical of DIA. At this time, other lesions that had been seen on the initial MRI remained either static or showed only mild increases in size. If the lesions had been metastatic, there would have been an increase in the size of the so-called primary lesion over time. However, these lesions remained relatively stable, whereas the right frontal focally dilated 'sulcus' had shown considerable growth. This led us to speculate that the lesions are metachronous.

Serial MR imaging also demonstrated a leptomeningeal or superficial cortical origin of the tumor. Both the lesions that had grown and those that had remained static presented features suggestive of a similar origin. The eccentric location of the spinal tumors may also indicate meningeal origin. It is possible that some of these smaller lesions may also grow over time. In most of the reported cases of metastases in DIA, the prognosis was related to the location of the tumor rather than the presence of metastases. This reaffirms our contention that multifocal lesions may, in actuality, represent metachronous lesions with prognoses that are the same as for the primary tumor. Although no long-term follow-up of DIA with metastases is available, the site of the tumor/metastases and the mass effect associated with the tumor appear to be predictive of the poor prognosis in the cases thus far reported.

**Conclusion**

This was the first reported case of a noninfantile variant of DIA with multiple craniospinal lesions at presentation. Serial MRI features of this tumor indicate a leptomeningeal origin of this lesion. Multiple new lesions appearing over time together with the relatively static older lesions could be indicative of the metachronous nature of this neoplasm. DIA/DIG has a propensity to be multifocal and, hence, these patients should be regularly followed up. Also, the prognosis in these cases depends on the tumor location rather than its histological or radiological appearances. Given the similarity between the infantile and noninfantile forms of DIG/DIA on radiology and in clinical presentation, these tumors should probably be considered a single entity.
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References


