Image Findings of Atypical Teratoid/Rhabdoid Tumors in Adult

Poster No.: C-0765
Congress: ECR 2013
Type: Educational Exhibit
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Keywords: Neuroradiology brain, Neuroradiology peripheral nerve, Neuroradiology spine, CT, MR, Imaging sequences, Image verification
DOI: 10.1594/ecr2013/C-0765

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Learning objectives

Our purpose is to clarify the image findings of adult AT/RTs. The subjects are 4 patients of adult AT/RT (60, 21, 31, 25 y.o., Male: Female=1: 1). CT, MRI and Angiography were performed for each case and the image findings are retrospectively evaluated.
Background

Atypical teratoid/ rhabdoid tumor (AT/RT) is a rare tumor occurring in less than 3 years old children. At first, this tumor had been reported as the pediatric renal tumor and, afterward, was reported as the pediatric central nervous system tumor, AT/RT which was different from the primitive neuroectodermal tumor (PNET). The pathological features are proliferation of rhabdoid cells and multiple systemic marker positive. Especially, most characteristic pathological feature is the frequent deletion and/or mutation of hSNF5/INI-1 gene in 22\textsuperscript{nd} chromosome (22q11). The prognosis is very poor (mean survival period; approximately 6 months).

AT/RT in more than 3 years old rarely occurs and adult onset AT/RT is especially extremely rare. Therefore, the systematic review on AT/RT has never been performed so far although several cases have been reported. We have experienced five AT/RT cases so far. However, of five cases, four cases are adult onset AT/RT ones in single institute. Our purpose is to clarify the image findings of adult AT/RTs.
Image Analysis

The subjects are four cases diagnosed as adult onset AT/RT in Yamagata University Hospital from 1999 to 2010 (2 men and 2 women; mean age: 34.3 years old; range: 21-60 years old). All cases were precisely pathologically diagnosed by biopsy, operation or autopsy. We retrospectively evaluated primary sites, image findings (CT, MRI and angiography) and survival period. CT findings were classified into high, iso or low density. MRI findings were classified into high, iso, low or mixed intensity in each sequence (including DWI, T1WI, T2WI and T1WI with gadolinium contrast enhancement). In this connection, "mixed intensity" was defined as a heterogeneous mixture of both high and low intensity in the tumor. Angiography findings were evaluated regarding the existent of tumor stain. These image findings were evaluated by an experienced neuroradiologist (10 years of experience). In Japan, if patients had not come to the hospital for five years, we may cancel image films. Therefore, if the image films were cancelled, a neuroradiologist evaluated the image findings by an image interpretation report.

Image Findings

All image findings and clinical features are summarized in Table 1. Tumors occurred in cervical spinal cord, at suprasellar cistern, at pineal region and at jugular foramen. All tumors showed hypointensity on T1WI, mixed intensity on T2WI and contrast enhancement. One of two cases in which angiography was performed showed obvious tumor stain and early venous filling. In only a case, the AT/RT showed high density on CT, very high intensity on diffusion weighted image (DWI) and decreasing apparent diffusion coefficient (ADC).

Case Presentation

We show three cases as follows in which the images are available.

- A case was a 60 years old male. The tumor, so called dumbbell shape, was seen in C5-Th1 level. MRI showed low intensity on T1WI, mixed intensity on T2WI and contrast enhancement (Fig. 1-4).
- A case was a 31 years old female. The tumor was seen in suprasellar region. MRI showed low intensity on T1WI, mixed intensity on T2WI and well contrast enhancement (Fig. 5-7). Tumor stain in angiography was not seen (Fig. 8).
- A case was a 25 years old male. The tumor was seen in left jugular foramen. CT showed slight high density with faint calcification and well contrast enhancement (Fig. 9, 10). MRI showed low intensity on T1WI, mixed intensity on T2WI, high intensity on DWI, low intensity on ADC map and
well contrast enhancement (Fig. 11-14). Angiography in left external carotid artery showed obvious tumor stain and early venous filling in left jugular foramen. Therefore, it was suggested that the tumor was marked hypervascularity (Fig. 15). The follow up MRI after 1 month revealed extremely rapid growth (Fig. 16) and the follow up CT after 7 months revealed metastasis for multiple organs (liver, bone, lung and so on) (Fig. 17).

Discussion

We have experienced four adult onset AT/RT cases in single institute so far. It is quite a surprising fact. The AT/RT is very rare, However, there is some possibility of occurring AT/RTs in adult cases. Actually, we have experienced a case of AT/RT in a 60 years old case. AT/RTs occur in a variety of sites (spine, suprasellar region, pineal region and jugular foramen). The prognosis was extremely poor (mean survival time: 16.5 months). MRI findings on T1WI and contrast enhancement was non specific. Mixed intensity on T2WI was seen in all cases. On the other hand, in one case, there was high intensity on DWI, decreasing ADC and hypervascularity on angiography. It may be reflect high cellularity in the tumor. In pediatric cases, charasteristic image findings is also high density on CT, calcification, hemorrhage, necrosis, cystic changes, high intensity on DWI and contrast enhancement. It may be specific image findings in adult onset AT/RTs as well.
### Table 1

<table>
<thead>
<tr>
<th>Age</th>
<th>Gender</th>
<th>Site</th>
<th>CT</th>
<th>MRI T1</th>
<th>MRI T2</th>
<th>MRI DWI</th>
<th>MRI CE</th>
<th>Tumor stain</th>
<th>Survival (month)</th>
</tr>
</thead>
<tbody>
<tr>
<td>60</td>
<td>Male</td>
<td>C5-Th1</td>
<td>NE</td>
<td>Low</td>
<td>Mixed</td>
<td>NE</td>
<td>+</td>
<td>NE</td>
<td>18</td>
</tr>
<tr>
<td>31*</td>
<td>Female</td>
<td>Suprasellar</td>
<td>NE</td>
<td>Low</td>
<td>Mixed</td>
<td>NE</td>
<td>+</td>
<td>—</td>
<td>24</td>
</tr>
<tr>
<td>21</td>
<td>Female</td>
<td>Pineal region</td>
<td>NE</td>
<td>Low</td>
<td>Mixed</td>
<td>NE</td>
<td>+</td>
<td>NE</td>
<td>14</td>
</tr>
<tr>
<td>25</td>
<td>Male</td>
<td>Lt. jugular foramen</td>
<td>High</td>
<td>Low</td>
<td>Mixed</td>
<td>High</td>
<td>+</td>
<td>+</td>
<td>10</td>
</tr>
</tbody>
</table>

NE: non evaluated


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Fig. 1: 60 years old male, T2 weighted image

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**Fig. 2:** 60 years old male, T1 weighted image

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Fig. 3: 60 years old male, T1 weighted image with gadolinium-based contrast media

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Fig. 4: 60 years old, male. T2 weighted axial image. The dumbbell shape tumor is seen (white arrow).

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Fig. 5: 31 years old female, T1 weighted image. Suprasellar tumor was seen.

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Fig. 6: 31 years old female, T2 weighted image.

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Fig. 7: 31 years old female, T1 weighted image with gadolinium-based contrast media.

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**Fig. 8:** Angiography (left internal carotid artery)

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Fig. 9: 25 years old, male. Plain CT.

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Fig. 10: 25 years old, male. Contrast enhanced CT.

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Fig. 11: 25 years old, male. T2 weighted image. The tumor is seen in left jugular foramen (white arrow head).

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Fig. 12: 25 years old, male. T1 weighted image.

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Fig. 13: 25 years old, male. Diffusion weighted image and apparent diffusion coefficient. The tumor is showed white arrow head.

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Fig. 14: 25 years old, male. T1 weighted image with gadolinium-based contrast media.

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**Fig. 15:** 25 years old, male. Angiography (left external carotid artery) showed obvious tumor stain and early venous filling.

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**Fig. 16:** 25 years old, male. The follow up CT after 1 month. T1 weighted image with gadolinium-based contrast media.

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Fig. 17: 25 years old, male. The follow up CT after 7 months. Multiple organs metastasis are seen in liver, bone, lung and so on.

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Conclusion

Adult onset AT/RT occurs in various sites and shows a variety of image findings. In spite of adult cases, AT/RT should be taken into consideration if the tumor shows mixed intensity on T2WI, high density on CT, hyperintensity on DWI and strange behavior of the tumor such as markedly rapid growth and metastasis for the other organs.
References


