

atypical teratoid/rhabdoid 2

가 ,

atypical teratoid/rhabdoid 2

Atypical teratoid/rhabdoid 1987

(1)

가

(1).  
(Primitive neuroectodermal tumor:PNET)

2 × 2.5 cm

가

(Fig. 1A),

가 , 70%

가

1B).

(Fig.

(1 - 2).

가

2

rhabdoid

가

features

, atypical teratoid/

vimentin, S - 100, neuron specific enolase(NSE)

rhabdoid

glial fibrillary acidic protein(GFAP), smooth muscle anti -  
gen(SMA) atypical teratoid/rhabdoid

T1

(1), T2

가

13

(2).

atoid/rhabdoid 2

atypical ter -

2

5

가

3

1

40

3360 g

3

가

2

40

,

3760 g

T1

T2

3 cm

가

, T2

가

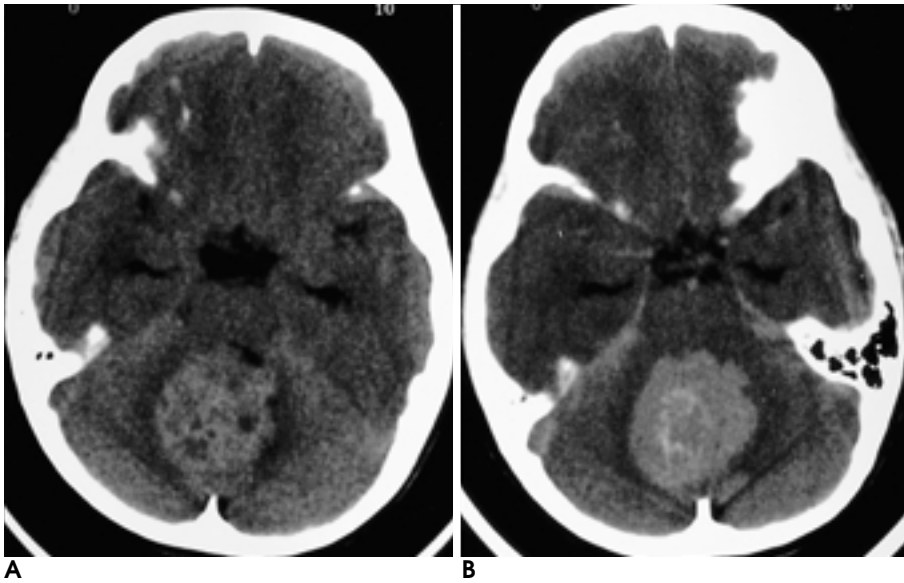
(Fig. 2A, B).

4

4

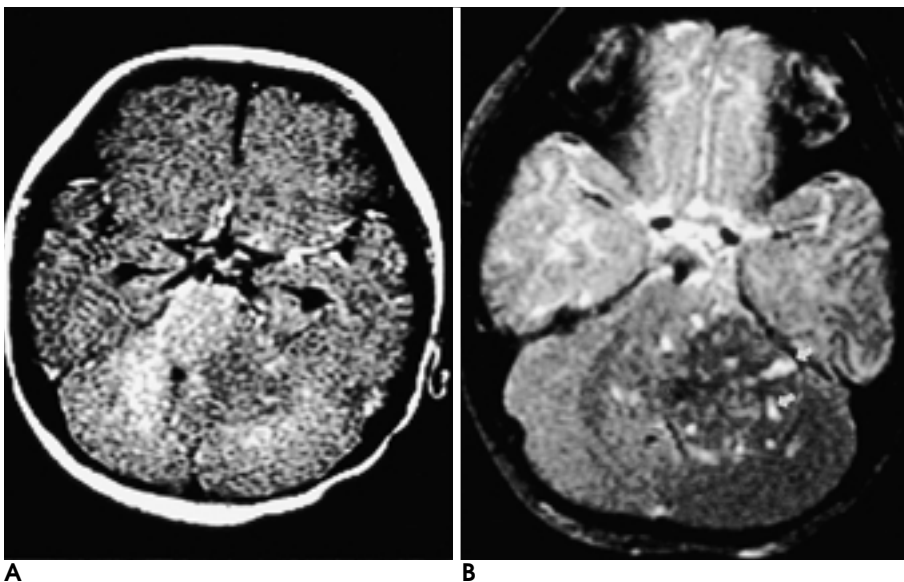
2000 8 3

2001 4 19



**Fig. 1.** A 3-year-old boy with atypical teratoid/rhabdoid tumor in the cerebellar vermis.

**A.** An axial CT scans show a high density mass in the cerebellar vermis and **B.** A patch enhancement with contrast enhancement that is compressing the fourth ventricle and surrounded by hypodense edema.



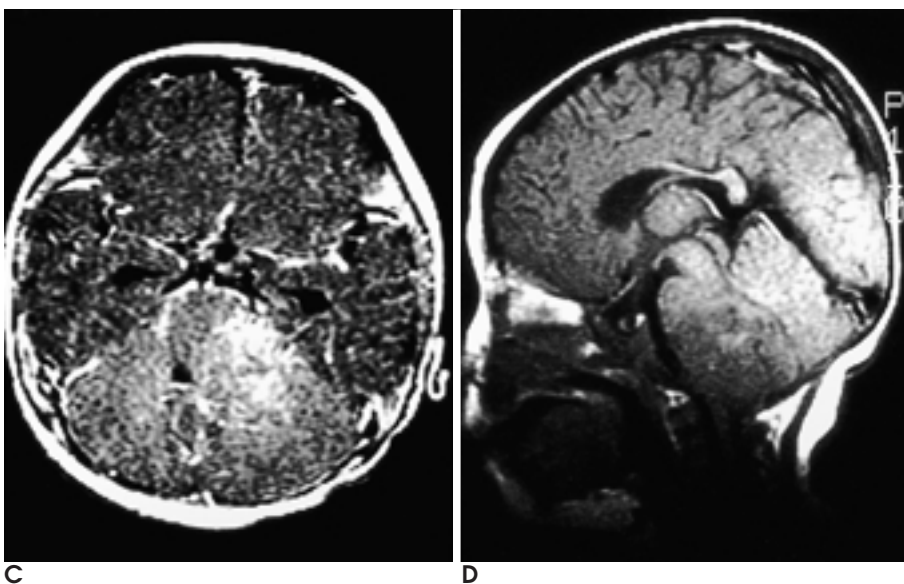
**Fig. 2.** A 5-month-old girl with atypical teratoid/rhabdoid tumor in left cerebellar hemisphere extending to pons.

**A.** An axial T1-weighted image shows a hypointense mass in the left cerebellar hemisphere with a mass effect to displace the fourth ventricle to the right side, resulting in a mild dilatation of temporal horn of lateral ventricle.

**B.** An axial T2-weighted image at lower level shows an isointense to hypointense mass containing multifocal cystic portions(white arrows).

**C.** The mass is inhomogeneously enhanced after gadolinium injection.

**D.** A sagittal T1-weighted image shows a large mass involving left cerebellar hemisphere.



4

(Fig. 2C, D).

vimentin, cytokeratin, epithelial membrane antigen (EMA)  
GFAP atypical teratoid/rhabdoid

4

Atypical teratoid/rhabdoid Beckwith & Palmer (1, 3)가 (Children's Cancer Group) Wilm 가

1984 Parham (3, 4)

42

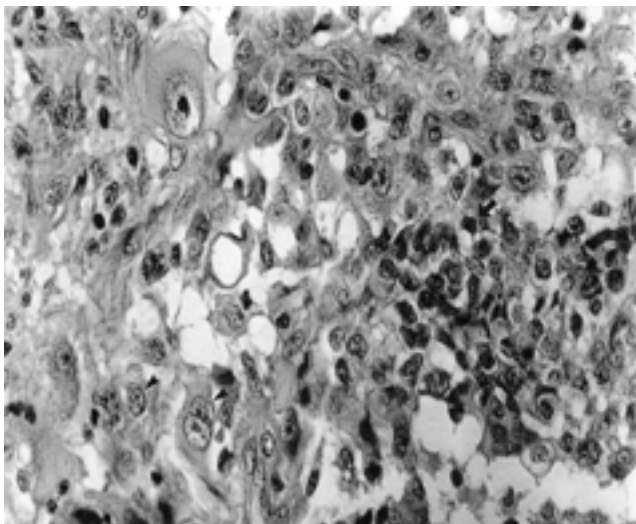
Lefkowitz (3)

Rorke (5, 6)  
atypical teratoid/rhabdoid 가

doid

Atypical teratoid/rhabdoid

가 (60%), (20%), (6%),



**Fig. 3.** There are scattered and small aggregates of larger cells (arrowhead) as well as small cell cluster. The larger cells have prominent nucleoli and abundant eosinophilic cytoplasm, which appears rhabdoid feature. The small cells (arrow) are similar to that seen in medulloblastoma (H & E,  $\times 400$ ).

(6, 7, 9).

Atypical teratoid/rhabdoid

가 .  
가

fibrovascular stroma

papillae gland-like space가

. Atypical teratoid/rhabdoid 가

(dispersed chromatin)

가 (Fig 3).

가 . ,

atypical teratoid/rhabdoid .

vimentin, EMA, SMA  
atypical teratoid/rhabdoid  
(1, 5).

5

가

가 4

가 ,

(8, 9).

24

5

가

, 10%

(8, 9).

Atypical teratoid/rhabdoid

가 . ,

teratoid/rhabdoid

가 , atypical (7,

9).

3 - 5

, atypical teratoid/rhabdoid 2

, 가 ,

(Pediatric Oncology Group)

40 - 60%  
atypical teratoid/rhabdoid  
1  
가 ,  
atypical ter -  
atoid/rhabdoid 가 (4, 6, 7).  
atypical teratoid/rhabdoid  
atypical teratoid/rhabdoid  
, 2 가  
atypical  
teratoid/rhabdoid , atyp -  
ical teratoid/rhabdoid  
가  
atoid/rhabdoid  
(3).

- Ultrastruct Pathol* 1997;21:369-378
2. Rorke LB, Packer R, Biegel J. Central nervous system atypical teratoid/rhabdoid tumors of infancy and childhood. *J Neurooncol* 1995;24:21-28
  3. Hilden JM, Watterson J, Longee DC, et al. Central nervous system atypical teratoid/rhabdoid tumor : response to intensive therapy and review of the literature. *J Neurooncol* 1998;40:265-275
  4. Choon-sik Y, Sylvester C, Venita J. Primary malignant rhabdoid tumor of the brain: CT and MR findings. *Yonsei Med J* 2000;41:8-16
  5. Proust F, Laquerriere A, Costantin B, Rochoux MM, Vannier JP, Freger D. Simultaneous presentation of atypical teratoid/rhabdoid tumor in siblings. *J Neurooncol* 1999;43:63-70
  6. Rorke LB, Packer RJ, Biegel JA. Central nervous system atypical teratoid/rhabdoid tumors of infancy and childhood: definition of an entity. *J Neurosurg* 1996;85:56-65
  7. Burger PC, Yu IT, Tihan T, et al. Atypical teratoid/rhabdoid tumor of the central nervous system : a highly malignant tumor of infancy and childhood frequently mistaken for medulloblastoma. *Am J Surg Pathol* 1998;22(9):1083-1092
  8. Atlas SW. *Magnetic resonance imaging of the brain and spine*. Pennsylvania : Lippincott- Raven, 1996 : 324 - 376
  9. Barkovich AJ. *Pediatric neuroimaging*. Philadelphia: Lippincott Williams & Wilkins, 2000: 453 -456, 501-502

1. Bhattacharjee M, Hicks J, Langford L, et al. Central nervous system atypical teratoid/rhabdoid tumor of infancy and childhood.

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## Imaging Findings of Atypical Teratoid/Rhabdoid Tumor of Infancy & Childhood in CNS: Report of Two Cases<sup>1</sup>

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Atypical teratoid/rhabdoid tumor rarely occurs in the CNS, though is most common in infants under two years of age. It is characterized by unique histologic features, has an extremely aggressive natural course, and is located mainly in the infratentorial region. Radiologically, it is difficult to distinguish from primitive neuroectodermal tumor or medulloblastoma. We report the radiologic findings of two cases of atypical teratoid/rhabdoid tumor.

**Index words :** Brain neoplasm, CT  
Brain neoplasm, MR  
Children, central nervous system  
Infants, newborn, central nervous system

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