Dumbbell-Shaped Primitive Neuroectodermal Tumor Mimicking Trigeminal Schwannoma: A Case Report and Review of Literature

ABSTRACT

Primitive neuroectodermal tumor (PNET) is presumably of neural crest origin, and cases of supratentorial PNET in adult are extremely rare. We reported a case of PNET presenting as a dumbbell-shaped tumor at the middle cranial fossa of skull base mimicking trigeminal schwannoma both clinically and radiologically. The patient underwent surgery on July 13, 2010, using a combination of subtemporal, pterional and retrosigmoid approaches, to achieve decompression and obtain a histological diagnosis. The patient worsened neurologically two weeks after surgery. Her consciousness level became comatose due to tumor bleeding with compression of the brain stem and her precarious physical condition prevented receiving further treatment with adjuvant radiation therapy and chemotherapy. The correct diagnosis of PNET is important because its management differs dramatically from that for trigeminal schwannoma.

KEYWORDS: Primitive neuroectodermal tumor, Trigeminal schwannoma, Dumbbell-shaped tumor

ÖZ

Primitif nöroektodermal tümörünün (PNET) nöral krest kökenli olduğu kabul edilir ve yetişkinlerde supratentoriyal PNET vakaları çok nadirdir. Kafatası kaidesinin orta kraniyel fossasında dambıl şekilli bir tümör olarak ortaya çıkan ve klinik ve radyolojik olarak trigeminal schwannomaya benzeyen bir PNET vakası sunuyoruz. Hastaya 13 Temmuz 2010 tarihinde dekompresyon elde etmek ve histolojik bir tanı almak için subtemporal, pterygoidal ve retrosigmoid yaklaşımların bir kombinasyonu kullanarak cerrahi yapıldı. İki hafta sonra hastanın durumu kötüye gitti ve fiziksel durumu olmaması adjuvan radyasyon tedavisi ve kemoterapiyle daha ileri tedavi verilmesini önledi. PNET’ye doğru tanı konması önemli olan ve trigeminal schwannomadan çok farklıdır.

ANAHTAR SÖZCÜKLER: Primitif nöroektodermal tümör, Trigeminal schwannoma, Dambıl şekilli tümör

INTRODUCTION

Primitive neuroectodermal tumors (PNETs) are small, round, malignant cell tumors, presumably of neural crest origin. Cases of supratentorial PNET mainly originate from the middle cranial fossa and are extremely rare primarily from the middle cranial fossa. We reported a patient with dumbbell-shaped appearance of PNET at the left middle cranial fossa of skull base mimicking trigeminal schwannoma clinically and radiologically. The diagnosis is important because its management differs dramatically from that of trigeminal schwannoma, including surgical excision followed by conventional craniospinal irradiation and chemotherapy for those with CSF dissemination.

CASE REPORT

Mrs. W was a 55-year-old female patient admitted to our hospital with complaints of left facial paresthesia (tingling sensation) and hypoesthesia for half a year. Trigeminal neuralgia was diagnosed at first and treated accordingly. However, her symptoms progressed from maxillary division to the entire territory of left trigeminal nerve rapidly. Diplopia subsequently developed 6 months later. The patient then came to Shin-Kong Hospital’s emergency department with additional complaints of ataxia and left-side deviation gait. The diagnosis was confirmed by a magnetic resonance imaging (MRI) showing a dumbbell-shaped tumor in the pons, extending from the left middle fossa, to the cerebello-pontine angle posteriorly and infratemporal fossa inferiorly (Figure 1A, B). The patient then was referred for surgical decompression and excision of the tumor.
necrosis (Figure 2A,B,D). T2-weighted MRI showed the lesion as non-homogeneously hypointense (Figure 2C).

The patient underwent surgery on 13 July 2010, via a combination of subtemporal, pterional and retrosigmoid approaches, to achieve decompression and obtain a histological diagnosis. The tumor was partially resected. The resected tumor was reddish and fragile. Histological examination of the surgical specimen showed atypical

Figure 1: Post-contrast brain CT showed a extra-axial dumbbell-shaped tumor across foramen ovale. Notice the left foramen ovale was much larger than the right side due to tumor erosion (A). The tumor extended to posterior fossa with compression of brain stem. Erosion of anterior portion of petrous bone was shown (B).

Figure 2: T1W image of MRI showed dumbbell-shaped tumor across the foramen ovale (A). Heterogenous enhancement was demonstrated after injection of gadolinium (B). T2W image showed low signal intensity of the tumor (C). The tumor extended to cerebello-pontine angle. The pons was compressed with enlargement of pre-pontine cistern (D).
round cells containing pleomorphic hyperchromatic round nuclei with occasional mitotic activity (Figure 3). Immunohistochemical examination revealed it was positive for CD99, CD56, NSE, and vimentin but was unreactive to cytokeratin, chromogranin A, synaptoptophysin, and LCA antibodies (Figure 4A-D). The final pathology report gave the diagnosis of PNET.

The patient deteriorated neurologically two weeks after surgery. Her consciousness became comatose due to tumor bleeding with compression of the brainstem and her precarious physical condition ruled out the possibility of adjuvant radiation therapy and chemotherapy. The patient died of brainstem failure two months after operation.

**DISCUSSION**

Medulloblastoma and supratentorial PNETs are rarely found in adults. Adult patients with PNETs generally present with symptoms and signs referable to a posterior fossa lesion, and the disease is usually limited to the posterior fossa at the time

![Figure 3: The tumor was supplied mainly by the left internal maxillary artery.](image)

![Figure 4: Photomicrograph showing the tumor cells with small, round, deeply basophilic nuclei, rich in chromatin but with little cytoplasm, indicating a highly cellular tumor (A). Immunohistochemical staining revealed positive reaction to CD99 (B), CD56 (C) and vimentin (D).](images)
of initial staging (4). PNETs have been considered embryonal
tumors composed of undifferentiated neuroepithelial cells
with a capacity of differentiation into different cellular lines,
such as astrocytic, ependymal, melanotic and muscular. They
are thought to arise from a neoplastic transformation of
primary neuroepithelial cells and can therefore be present in
virtually any part of the nervous system (1). The reported
patient is remarkable because trigeminal neuralgia is an
uncommon initial clinical presentation of primary extra-axial
PNET and that the middle cranial fossa is possible but rare
initial presentation site of PNET. It exhibited the neuroimaging
features of dumbbell-shaped tumor at the middle cranial
fossa mimicking trigeminal schwannoma clinically and
radiologically.

There was a case report about a dumbbell-shaped appearance
of spinal PNET (3). However, to our knowledge, this is the first
report case of primary dumbbell-shaped PNET located at the
middle cranial fossa and infratemporal fossa with intradural
and extradural components.

Peterson, K reviewed 54 adults with PNET and reported that
the disease in most patients was limited to the posterior fossa
at the time of diagnosis (4). For patients with supratentorial
PNETs, they are usually large, bulky, heterogeneous masses
with “cystic” (necrotic) areas, calcification, and very little
edema (5). This case showed heterogeneity and necrosis in
the largest infratemporal component of the tumor, probably
due to the tumor’s rapid growth.

PNET mimicking cranial schwannoma is a rare disease entity
and proper differential diagnosis between the two diseases
is crucial as their treatment modalities are vastly different.
Tripathy, LN reported a case first treated as an acoustic neuroma
with radiosurgery instead of craniospinal radiotherapy. One
year later, the patient developed craniospinal dissemination
(7).

Adjuvant combined radiochemotherapy is clearly essential
for the treatment of peripheral PNETs, but the most impor-
tant factor determining the patient’s prognosis is whether ag-
gressive surgical intervention is possible or not. Smee, R.I. et
al. reported that the outcome for adults is the same as that of
pediatric patients treated with craniospinal radiotherapy and
chemotherapy. The authors also suggested that radiothera-
py could control local diseases in which complete resection
could not be achieved (6).

It has been recently reported that the mean time to cranial
recurrence of adult patients with cerebellar medulloblastoma/
PNET was 19 months; to distant metastases was 18 months.
The relapse rate was up to of 21% to 49%. The high possibility
of delayed cranial relapse and metastasis mandates close
follow-up of all patients with this disease, with prompt
evaluation of new symptoms (2,4).

**CONCLUSION**

PNET can present initially as atypical trigeminal neuralgia,
a condition more commonly associated with trigeminal
schwannoma. Differential diagnosis and pathological proof
is warranted before initiation of treatment in any patient
presenting with atypical trigeminal neuralgia related to
dumbbell-shaped tumor located at the middle cranial fossa,
as the treatment of PNET and trigeminal schwannoma vastly
differ from one another.

**REFERENCES**

1. De Tommasi A, De Tommasi C, Occhiogrosso G, Cimmino A,
Parisi M, Sanguedolce F, Ciappetta P: Primary intramedullary
primitive neuroectodermal tumor (PNET)--case report and
2. Ertas G, Ucer AR, Altundag MB, Durmus S, Calikoglu T,
Ozbagi K, Abanuz H, Altundag K, Demirkasimoglu A:
Medulloblastoma/primitive neuroectodermal tumor in
adults: Prognostic factors and treatment results: A single-
3. Hrabalek L, Kalita O, Svebisova H, Ehrmann J Jr, Hajduch M,
Trojanec R, Kala M: Dumbbell-shaped peripheral primitive
neuroectodermal tumor of the spine - case report and review
4. Peterson K, Walker RW: Medulloblastoma/primitive neuroec-
5. Robles HA, Smirniotopoulos JG, Figueroa RE: Understanding
the radiology of intracranial primitive neuroectodermal
tumors from a pathological perspective: A review. Semin
Ultrasound CT MR 13(3): 170-181, 1992
6. Smee RI, Williams JR: Medulloblastomas - primitive neuroec-
todermal tumours in the adult population. J Med Imaging
7. Tripathy LN, Forster DM, Timperley WR: Adult primitive
neuroectodermal tumour. A case report and review of the