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

Trigeminal Schwannomaya Benzeyen Dambıl Şekilli Primitif Nöroektodermal Tümör: Olgu Sunumu ve Literatürün Gözden Geçirilmesi

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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 yetişkinde 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 alabilmek için subtemporal, pteriyonal ve retrosigmoid yaklaşımların bir kombinasyonu kullanılarak cerrahi yapıldı. İki hafta sonra hasta nörolojik olarak kötüye gitti. Tümör kanaması ve beyin kökü kompresyonuyla komaya girdi ve fiziksel durumu iyi olmaması adjuvan radyasyon tedavisi ve kemoterapiyle daha ileri tedavi verilmesini önledi. PNET'ye doğru tanı konması önemlidir çünkü takibi 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. Neurological examination on admission revealed trigeminal nerve and left abducens nerve palsy, left hearing loss and impaired tandem gait. Her brain computerized tomography showed a dumbbell-shaped tumor across the foramen ovale that extended from the left middle fossa, to the cerebello-pontine angle posteriorly and infratemporal fossa inferiorly (Figure 1A,B). The foramen was enlarged and eroded by the tumor. Coronal T1-weighted magnetic resonance imaging (MRI) disclosed a large dumbbell-shaped isointense lesion, affecting the left cavernous sinus and compressing the pons, with heterogeneous enhancement after gadolinium administration. The infratemporal portion showed 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

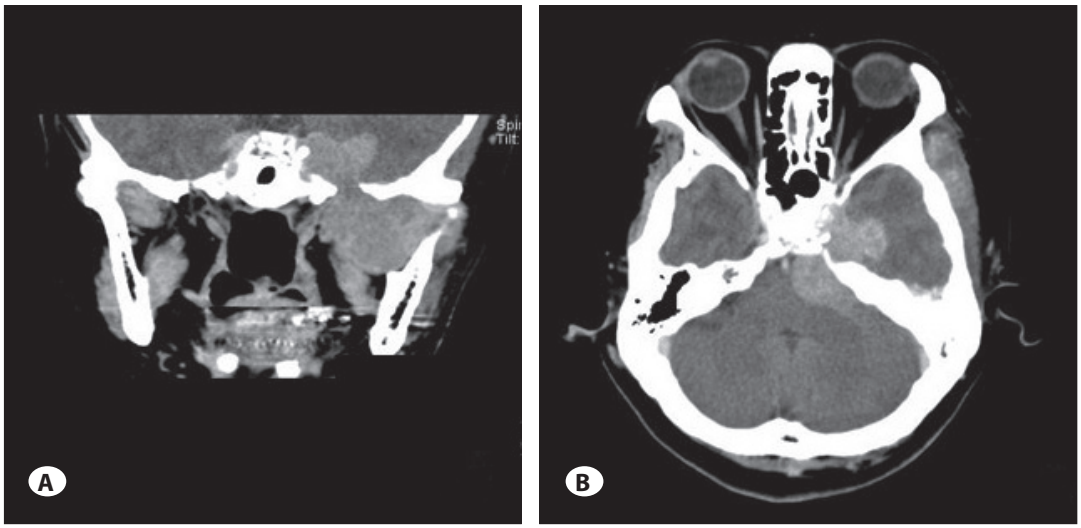

Figure 1: Post-contrast brain CT showed a extra-axial dumbbell-shaped tumor across foramen ovale. Notice the left foramen ovale was much larger than the right side due to tumor erosion **(A)**. The tumor extended to posterior fossa with compression of brain stem. Erosion of anterior portion of petrous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

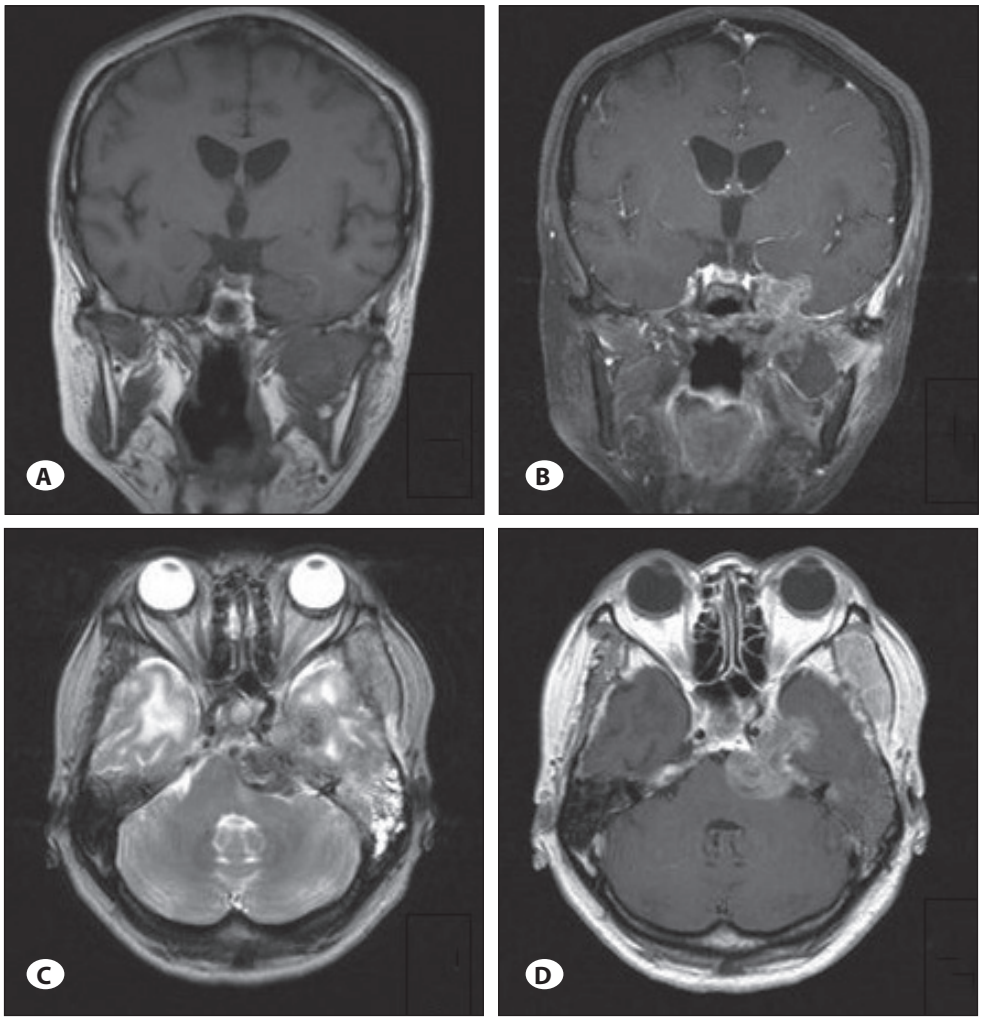

Figure 2: T1W image of MRI showed dumbbell-shaped tumor across the foramen ovale **(A)**. Heterogenous enhancement was demonstrated after injection of gadolinium **(B)**. T2W image showed low signal intensity of the tumor **(C)**. The tumor extended to cerebello-pontine angle. The pons was compressed 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, synaptophsin, 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

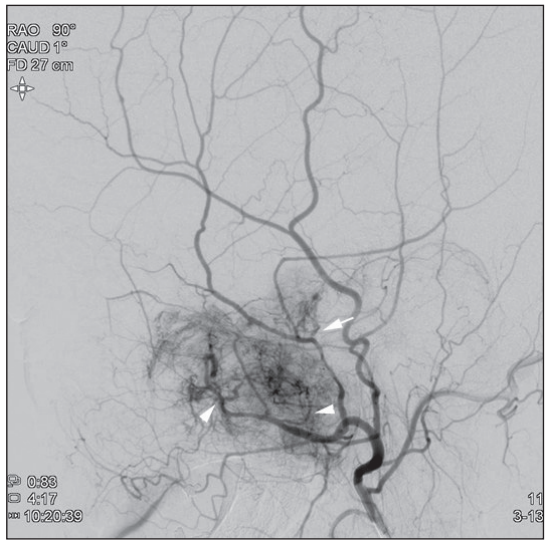

Figure 3: The tumor was supplied mainly by the left internal maxillary artery.

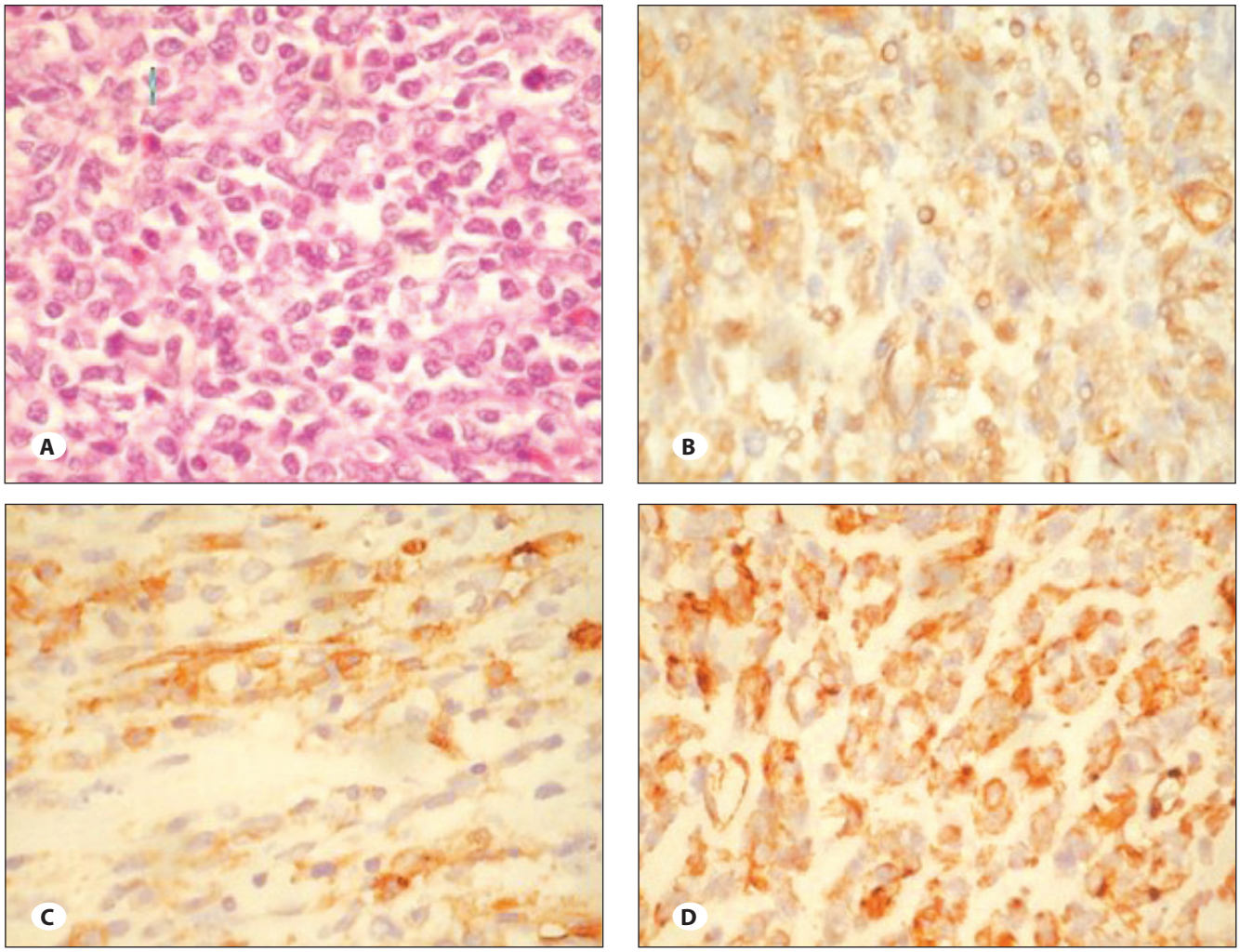

Figure 4: Photomicrograph showing the tumor cells with small, round, deeply basophilic nuclei, rich in chromatin but with little cytoplasm, indicating a highly cellular tumor **(A)**. Immunohistochemical staining revealed positive reaction to CD99 **(B)**, CD56 **(C)** and vimentin **(D)**.

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 primitive 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 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 heterogenicity 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 important factor determining the patient's prognosis is whether aggressive 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 radiotherapy 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 schwanoma. 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.

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