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Cerebellar Mutism Following Posterior Fossa Tumor Resection in Children

Çocukluk Çağı Arka Çukur Tümörleri Cerrahisi Sonrası Serebellar Mutizm

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ABSTRACT

AIM: Cerebellar mutism is a documented complication of posterior fossa surgery in pediatric ages. Risk factors such as the type of tumor, size, and location of tumor, hyrdrocephalus, postoperative cerebellar swelling for cerebellar mutism were investigated in this study.

MATERIAL and METHODS: A consecutive series of 32 children with a cerebellar tumor were operated on at the Haseki Educational and Research Hospital, Department of Neurosurgery, between 1990 and 2005. Their speech and neuroradiological studies were systematically analysed both preoperatively and postoperatively

RESULTS: Cerebellar mutism developed in ten children (32%) in the early postoperative period. The type of tumor, midline localization, and vermian incision were significant single independent risk factors. In addition, an interdependency of possible risk factors (tumor>5 cm, medulloblastoma) was found. The latency for the development of mutism ranged from 0 to 90 days (mean 15.6 d). The speech returned to normal in eight patients. All cases were accompanied by cerebellar ataxia.

CONCLUSION: Mutism after posterior fossa tumor resection is also associated with ataxia. Cerebellar mutism usually has a self-limiting course and a favorable prognosis.

KEY WORDS: Astrocytoma, Children, Medulloblastoma, Mutism, Posterior fossa tumor

ÖZ

AMAÇ: Serebellar mutism çocukluk çağında arka çukur tümörleri cerrahisinden sonra gelişebilen bir komplikasyon olarak dökümente edilmiştir. Bu çalışmada, tümör çapı, lokalizasyonu, hidrosefalus ve operasyon sonrası beyincik şişmesi gibi serebellar mutism için risk faktörleri araştırıldı

YÖNTEM ve GEREÇ:1990 ile 2005 yılları arasında Haseki Eğitim ve Araştırma Hastanesi Beyin Cerrahisi Kliniğinde ardı sıra opere edilen 32 arka çukur tümörlü çocuklardan operasyon öncesi ve sonrası konuşma özellikleri nöroradyolojik bulguları sistematik olarak not edildi.

BULGULAR: Toplam 10 (%32) çocuk erken operasyon sonrası dönemde serebellar mutizm geliştiği gözlendi. Tümör tipi, orta hat lokalizasyonu ve vermian insizyon risk faktörleri idi.% 64 olgu ile medulloblastoma en sık ikinci sırada % 36 ile astrositoma tespit edildi. Gerçekten de 5 cm den daha büyük medulloblastomalarda daha belirgindi. Mutism gelişimi operasyon sonrası ilk gün ile 90 gün arasında (ortalama 15.6)) değişiyordu. Konuşmaları 8 hastada geri döndü ve tamamında cerebellar ataksi eşlik ediyordu.

SONUÇ: Mutizm sıklıkla serebellar ataksi ile beraberdir ve kendi kendine iyileşebilen prognozu iyi olan bir klinik tablodur.

ANAHTAR SÖZCÜKLER: Arka çukur tümörü, Astrositoma, Çocuk, medulloblastoma, Mutizm

INTRODUCTION

Postoperative or iatrogenic cerebellar mutism (CM), defined as a condition of complete absence of speech that is not associated with other aphasic symptomatology or alteration of consciousness may occur in various neurological conditions such as hemorrhage, infection, degenerative disease, neoplastic disease of the cerebellum (9,12,24,34) . The resection of posterior fossa tumors in the pediatric ages is also associated with postoperative mutism, and it has been well characterised with multiple case reports and reviews (1,3,5-7,13,15-22, 24, 27,28-31, 34,35).

The core features of this predominantly pediatric syndrome are;

1- it occurs after resection of a cerebellar mass lesion

2- there is delayed onset of mutism after a brief interval of 1-4 days of relatively normal speech postoperative surgery.

3- there is delayed onset mutism that lasts from 1 day to 3 months, followed by a severe absence of speech, which recovers fully in between 1 and 3 months

4- frequent association with other neurologic manifestations, such as long tract signs and neurobehavioral abnormalities (32,33).

CM after posterior fossa tumor resection is always associated with midline tumors, and medulloblastoma predominates as the underlying pathology.

In retrospective studies various risk factors have been suggested such as tumor size, length of the vermian incision at surgery, and postoperative complicatipons such as edema within the pontine tegmentum or brachium pontis, hydrocephalus, and meningitis (9,12,24,34).

In this study we examined whether the type, size, and location of the tumor, the type of cerebellar incision, preoperative and postopeprative hydrocephalus, postoperative infection, and edema are related to CM.

MATERIAL and METHODS

A consecutive series of 32 children with a cerebellar tumors were operated at the Haseki Educational and Research Hospital, Department of Neurosurgery, between 1990 and 2005. 32 children (19 boys and 13 girls, age range 3-13 years) with normal psychomotor development and age-appropriate speech were admitted for a first resection of a posterior fossa cranial tumor. A diagnosis of medulloblastoma (64%) was made In 20 cases. The tumor was found to be a low grade astrocytoma in 12 cases (36%). No speech disorders or dysarthria had been found preoperativelyl. The CM syndrome occurred in 10 (32%) out of 32 children. All had cerebellar ataxia except two cases. The ataxia was both truncal and appendicular. It was often asymmetrical. Three patients had 6th nerve pareses following surgery and four patients had 7th nerve pareses which resolved completely.

Speech behavior was followed-up every day during the week after surgery in all children. In cases of mutism, they were followed-up at intervals as short as possible for 4 weeks. They were assessed every ten days until the facility for speech was regained.

The speech criteria were,

1- spontaneous language

2- repetition of words and sentences,

3- reading

4- watching anything

5- smelling and interacting with another child in the pediatric room.

These criteria are modified from the Mayo clinic lists of Darley et al (10).

Speech characteristics such as voice, quality, nasality, articulation, speech rate, and respiration were assessed. A neurological examination was performed to exclude any other neurological disease.

The Children Oncology groups (27) has prepared a cerebellar mutism syndrome survey in 2006, but we did not use this as our study began in 1990.

The latency for the development of mutism ranged from 0 to 90 days (mean 15.6 d). Speech returned to normal in eight patients. All of them had always accompanied by cerebellar ataxia.

Brain computed tomography (CT), or magnetic resonance imaging (MRI), or both were studied to determine the preoperative maximum lesion diameter and size of tumor. We collected all information about their postoperative data such as edema, ventricular enlargement, infection, and hemorrhage.

The records of these patients were reviewed to identify patients who developed mutism following the surgical procedure and who were alive a minimum of one year after the surgery.

Except seven cases, these cases had pseudomeningocele or cerebrospinal fluid leaking from the incision location. Four cases were reoperated due to pseudomeningocele in the early postoperative period (12. 8 %). Seven cases had a residual tumor under 2 cm in diameter.

Case 1. A 6-year-old girl presented with a six-month history of headache, nausea and vomiting. Ataxic gait had appeared in the last month. She had dysmetria, ataxic gait, bilateral dysdiadakokinesia, horizontal nystagmus, and severe papilledema. The girl was able to speak fluently. Brain MRI showed a tumor in posterior fossa and the brain stem was infiltrated by the tumor (Figure 1). The patient was operated in the prone position with three point cranial fixation and tumor excision with vermian incision was performed the next day. In the postoperative period, MRI revealed that the tumor has been removed totally (Figure 2). One day after the operation, the patient developed mutism, which lasted for seven weeks. The mutism was not accompanied by long tract signs or cranial nerve palsy. At the end of the third week the mutism regressed, and the speech returned to normal on the postoperative third month. Histopathological diagnosis of the tumor was pilocytic astrocytoma. The patient has been followed-up for 2 years.

Case 2. This 5-year-old right handed girl with a history of headache had sudden vomiting followed by ataxia in May 1993. CT imaging revealed a large

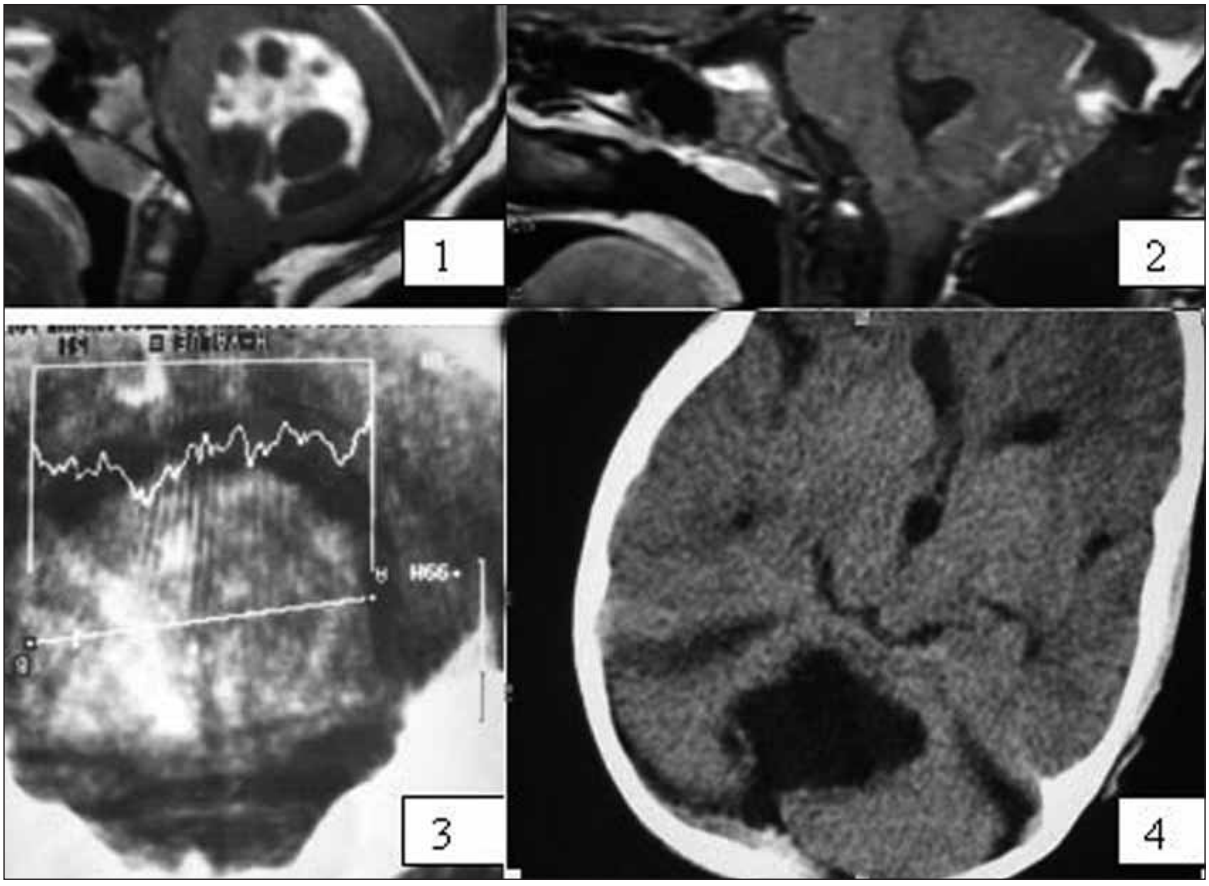

Figure 1: *Preoperative MRI showing the median posterior fossa tumor. The brainstem is compressed by the tumor. A cystic and solid lesion surrounding the tumor anteriorly which is compressing the dorsal midbrain (pilocytic astrocytoma).*
Figure 2: *Postoperative sagittal magnetic resonance image showing no residual tumors.*
Figure 3: *Preoperative computed tomography image (with contrast) showing the median posterior fossa tumor (Medulloblastoma).*
Figure 4: *Postoperative computed tomography showing the posterior median approach leading to the tumor. There is no residual tumor in the early postoperative period.*

midline posterior fossa tumor (Figure 3). The patient had severe hydrocephalus and was taken urgently to the operating room for a posterior fossa decompression and gross total resection of the cerebellar tumor. Surgical pathology reported the contents to include medulloblastoma. The postoperative CT did not show any residual tumor (Figure. 4). The patient was extubated on the first postoperative day and was initially nonverbal, displaying signs of cerebellar mutism. After 35 days, she began to have slurred speech, indicating a shift in phonation to ataxic dysarthria. She continued to show improvement in her phonation and articulation. The patient was discharged home with plans to follow-up with a speech therapy. The patient died after two years.

Statistical analysis

The tumor size, type, incision site were analyzed. The CM was correlated with these parameters by linear regression analysis. The Pearson correlation coefficient was used to determine the level of statistical significance. The dependence of the CM and the relation to the tumor parameters were analyzed using the Mann-Whitney U-test. The p values were used in the exact analyses for the statistical significance. The tumor type and type of incision variables were dischotomised as follows: tumor type was divided into medulloblastoma versus other tumor type (astrocytoma). Tumor site was defined as tumor localization in the cerebellar midline with or without tumor extension into the cerebellar hemisphere, versus tumor localization.

RESULTS

The CM syndrome occurred in 10 (32%) out of 32 children. This is a high rate as the tumors were bigger at the time of diagnosis in our series. They had both medulloblastoma and low grade astrocytoma. CM occurred in 6 of 20 patients with a medulloblastoma (30 %) and 4 of 12 (33 %) boys with a low-grade astrocytoma. Invasion of the brain stem and of the cerebellar peduncle was present in 5 patients. Ventricular shunting was performed in four cases in the postoperative period. The size of the medulloblastoma ranged from 4.1 to 7.5 cm (mean 4.5 cm). The largest tumor was a medulloblastoma in the median location in the posterior fossa. The size of the astrocytoma ranged from 3.4 to 7.8 cm (mean 5.6 cm). The duration of CM varied from 1 day to 6 months.

Analysis model calculations in linear regression showed that the risk of developing CM was increased 6.7 times ($p<0.003$) if the tumor site was cerebellar midline versus tumor localization laterally in the cerebellar hemisphere. In the medulloblastoma group, each increase of tumor size by 1 cm meant a multiplication of the odds to develop CM by 1.53 ($p<0.04$). It may therefore be concluded that size only has an effect in the medulloblastoma group, or in other words: medulloblastoma is a risk factor for CM only when the tumor size is bigger than 5 cm.

DISCUSSION

CM after posterior fossa surgery was first reported by Rekate et al. and Yonemasu in 1985 (25, 35). Mutism is the total loss of ability to speak without any symptoms related to aphasia and without loss of consciousness. The cause of this syndrome or its exact mechanisms are still unknown, but it has been associated with larger midline tumors. CM has been documented as a complication of resective surgery for posterior fossa tumors, especially in children. However, there are only few reports of posterior fossa mutism that are not due to neuro-trauma or neuro-surgical trauma (2, 20, 21). Other causes of CM are infections and vascular accidents (2,14,20,23,25, 26, 32,33). The reason could be damage (ischemic or edematous) to the dentate nucleus and dentatothalamocortical and dentorubrocortical pathways with the possible role of the cerebellum in cognition and in higher functions. This damage has been proposed to develop secondary to postoperative spasm of the cerebellar arteries (17).

CM has an estimated incidence of 10% to 25 % (1-3,5-12, 23-34). The patient has cessation of speech 24 to 72 hours postoperatively, increased emotional lability, pseudobulbar palsies, and possibly hemiparesis. The emotional lability and weakness subside over several weeks, but the mutism may persist for months. There is often some residual dysarthria. Resection of the vermis may play a role, buteven extensive resection of the vermis caused very little neurological deficit (2). Similarly, Pollack reported that there is no correlation between the size of the tumor and length of vermian incision and the possibility of development of mutism (24). Injury to dentate nucleus or the cerebellar peduncles has also been theorized as a causative factor. Kusano reported that the cerebellar mutism may have been caused by

bilateral damage to the dentate nuclei and not by unilateral damage (18).

The Children's Oncology Groups' (27) recent study is the first large-scale, multiinstitutional prospective study of CMS in children who have undergone medulloblastoma surgery. Despite some limitations in diagnosis, their study confirms that, far from being a rare problem, mutism occurred after nearly one quarter of posterior fossa craniotomies for both average-risk and high-risk medulloblastoma and was judged to be of moderate or severe intensity in more than 90% of their patients in whom it was identified. Although there was a negative correlation with cerebellar hemisphere location, the invasion of the brainstem was the only risk factor that correlated positively with CMS in both trials and this finding may suggest a neuroanatomical locus for the mutism. The patients had both cerebellar ataxia and cerebellar mutism.

What is the cause? The fact that vermian lesions (particularly of the inferior lobules) are correlated to different degrees of behavioral disturbances and cerebellar ataxia that range from irritability to a general tendency towards avoidance that may even reach the stage of transitory autism supports the role of the vermis as a cerebellar limbic system (4). Furthermore, the fact that lesions of the cerebellar hemispheres lead to deficits in complex mental activities supports the role of these hemispheres in the modulation of thought, language and executive abilities. Although this conclusion needs to be approached with caution, the two cerebellar hemispheres may have a right-left specialization similar to that of the cerebral hemispheres (26). More recently, Doxey et al. reported in their group of patients of 20 children with mutism following posterior fossa tumor resection that the mutism recovered in all, but there were other residual neurologic deficits, particularly ataxia, in all patients (11). In contrast, Steinbok et al reported in their group of patients of seven children with mutism that following posterior fossa tumor resection, the mutism did not recover except in one case. The long-term outcome of the associated neurological deficits was determined in this study (30), and some of these, particularly the ataxia, tended to persist. This result is not consistent with our cases where ataxia and another neurological deficits such as cranial nerve pareses were resolved.

CM is a rare postoperative phenomenon that occurs predominantly in children after posterior fossa surgery. CM usually has a self-limiting course and a favorable prognosis for resolution.

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