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Peripheral presentation of central facial palsy

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Facial nerve palsy in children may be congenital or due to neoplastic, infectious or traumatic conditions. Although idiopathic (Bell's palsy) facial nerve palsy is the most commonly encountered cause, facial nerve palsy may be the first and only symptom of a severe underlying condition. Previous studies on pediatric populations have reported on a tumor cause of facial nerve palsy in 2%-12% of cases. One of the most important pitfalls for a clinician is to assume that a child with facial nerve palsy has Bell's palsy and does not need complete and comprehensive evaluation for other etiologies. A case is described of an infant with oculocutaneous albinism who presented with persistent findings of peripheral facial palsy in whom the cause was belatedly identified as a brain tumor following magnetic resonance imaging. The most striking feature in our patient was the peripheral presentation of central facial nerve palsy, and if the magnetic resonance imaging had not been obtained, a potentially serious diagnosis may have been missed. Complete medical history and detailed physical examination should be obtained for every patient presenting with peripheral facial palsy, particularly in infants where cranial tomography or magnetic resonance imaging may be warranted to avoid misdiagnosis.

Keywords: facial paralysis; albinism, oculocutaneous; Bell palsy; brain neoplasms; infant

INTRODUCTION

Facial nerve palsy (FNP) is a common illness with potentially severe consequences that may negatively affect the patient's quality of life. The estimated annual incidence of FNP is 15-40 *per* 100 000 individuals (1). FNP in children may be congenital or due to neoplastic, infectious or traumatic conditions. Although idiopathic (Bell's palsy) FNP is the most commonly encountered cause, FNP may be the first and only symptom of a severe underlying condition (2, 3).

A diagnosis of Bell's palsy should only be made after other causes have been exclusively ruled out, particularly since any delays in diagnosing a neoplastic etiology may have dire consequences. Previous studies on pediatric populations have reported a tumoral cause of FNP in 2%-12% of cases (4). One of the most important pitfalls for a clinician is to assume that a child with FNP does not need complete and comprehensive evaluation (4). Following detailed history and careful physical examination, undertaking imaging modalities such as computed tomography (CT) or magnetic resonance imaging (MRI) is recommended to prevent a misdiagnosis, particularly in infants with peripheral facial palsy (5).

A case is described of an infant who presented with persistent findings of peripheral facial palsy in whom the cause was belatedly identified as a brain tumor.

CASE REPORT

A 45-day-old boy was brought to the emergency department after his mother had noticed that his mouth was drawn to the left side when he cried. His complaints started 5 days before and did not exist at childbirth. He was born by normal vaginal delivery after 39 weeks of gestation to a 34-year-old woman (gravida 3, para 1) following an unremark-

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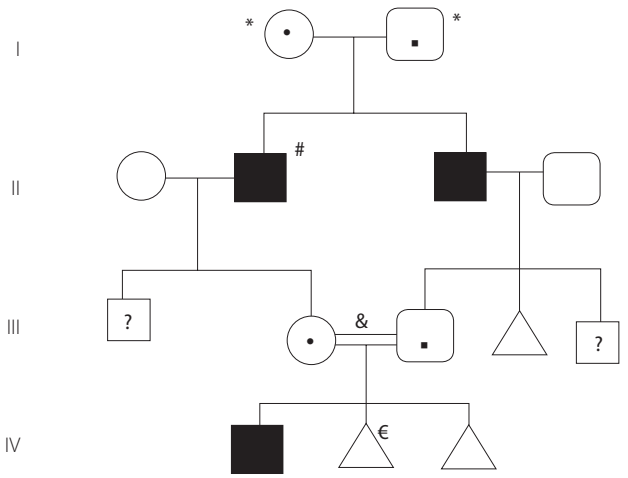

*carrier (autosomal), #affected individuals, €miscarriage, &consanguineous marriage

FIGURE 1. The patient's pedigree (parents were third-degree relatives, and the mother had a history of 2 previous stillbirths)

able pregnancy. He had no history of previous hospitalization. The patient's parents were third-degree relatives, and the mother gave a history of two previous stillbirths of unknown cause. The pedigree is presented in Figure 1.

Physical examination revealed an infant in good general condition with normal vital signs. He was 57 cm (50th percentile) long, weighing 4250 g (25th-50th percentile) with head circumference of 39 cm (50th-70thpercentile). The patient had flattening of the forehead and nasolabial fold on the left side with a sagging eyebrow. He had nystagmus, was unable to close his right eye, and his mouth was drawn to the non-affected side (Figure 2). Spontaneous movement of all four extremities was observed. Physical examination was otherwise unremarkable except for findings consistent with oculocutaneous albinisim (OCA). The characteristic findings of OCA, which involved both the skin as hypopigmentation and the eyes as pendular nystagmus and iris transillumination defects, had been present from birth in our patient. He was also examined by an otolaryngologist and temporal CT was proposed. The findings reported by the ophthalmologist were consistent with our findings. Initial blood workup did not reveal any abnormalities and the results of urinalysis were normal.

Although temporal CT scan was normal, subsequent brain MRI revealed the presence of a cystic lesion resembling an arachnoid cyst, 22x15x14 mm in size, at the level of the posterior fossa anterior to the right cerebellum, displacing the medulla spinalis left laterally to the level of the bulbus. Medially to this lesion, a midline posterior mass lesion, 19x24x19 mm in size, resembling an ependymoma was seen, probably originating from the fourth ventricle and resulting in irregularity and dilatation of the ventricle (Figure 3). After referral to a neurosurgeon, the mass was removed surgically with a histopathologic diagnosis of ependymoma. The patient was referred to pediatric oncology department for treatment. Twenty days after surgery, he was able to close his eyes; two months after surgery, his mouth was slightly drawn to the left side when he cried. Except for his OCA findings, his neurologic examination was normal.

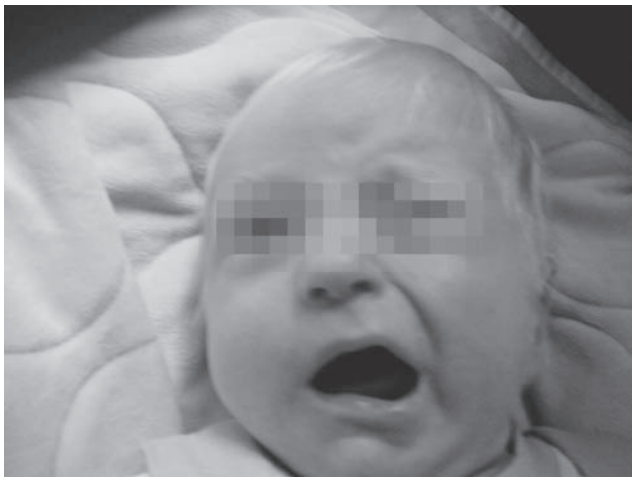

FIGURE 2. A photograph of the patient obtained on presentation

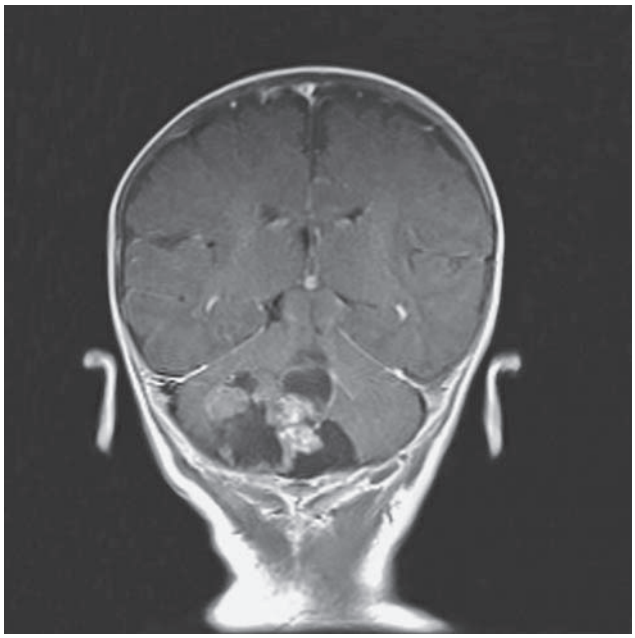

FIGURE 3. Brain MRI scan showing a mass originating from the fourth ventricle, later identified to be an ependymoma (coronal T1 weighted sequences after intravenous contrast)

DISCUSSION

The facial nerve is a mixed-complex nerve containing motor, parasympathetic, special sensory (taste), and sensory components (5). Its motor nucleus lies deep within the re-

ticular formation of the pons, where it receives input from the precentral gyrus of the motor cortex. The motor fibers innervate the muscles of facial expression, the posterior belly of the digastric muscle, the stylohyoid muscle, and the stapedius muscle. Upper motor neuron (supranuclear) tracts descend through the internal capsule and synapse on neurons in the facial motor nucleus. Neurons in the dorsal aspect of the facial motor nucleus supplying the upper face receive inputs from both sides of the cortex, thus exhibiting bilateral innervation, whereas tracts to the lower face mainly receive contralateral input (5). Accordingly, a lower motor lesion of the facial nerve or peripheral facial nerve palsy is usually considered when forehead muscles are involved, and there is a tendency not to obtain cranial imaging in patients with a presumed 'peripheral' presentation. However, this may be misleading since an injury to the facial nerve at any level between the pons and the internal capsule may also result in symptoms and signs consistent with peripheral facial nerve palsy (5).

Although not a life-threatening condition, FNP remains relatively common with severe negative effects on the patient's quality of life. The etiology of facial palsy may be classified into infectious causes such as otitis media, chickenpox, herpes zoster oticus, mumps and mononucleosis, and noninfectious causes including congenital disorders, trauma, neoplasms, metabolic and autoimmune diseases. When the cause cannot be identified, FNP is considered idiopathic (Bell's palsy) (4).

In a study by *May et al*. (6) on 170 pediatric cases of FNP spanning a period of 17 years, Bell's palsy was the final diagnosis in 42% of patients, followed by trauma (15%), otitis media (12%), congenital conditions (7.6%), birth trauma (5%) and neoplasia (4%). Identification of the cause requires review and exclusion of many potential causes in the differential diagnosis.

Neoplasias remain one of the most important etiologies for FNP, since a late diagnosis often leads to delayed treatment (4). Tumors that have been implicated as a cause of FNP may be intracranial or extracranial causes. Common intracranial tumors include astrocytoma, neuroblastoma, ependymoma, acoustic neuroma, and pontine glioma (4). Ependymomas, which are derived from ependymal cells that traverse the central nervous system, are the third most common brain tumors in children. In this age group, ependymomas typically arise within the fourth ventricle, often growing and filling the whole ventricle (7). In a study by *Morrris et al*. (8), tumors (including infratentorial ependymomas) were identified as the cause of FNP in 34 out of 96 children (35%). The tumor that was believed to be the cause of FNP in our patient arose from the fourth ventricle and following surgical resection was identified to be an ependymoma.

Albinism is an autosomal recessive disorder of melanin synthesis characterized by the congenital absence or reduced production of melanin. In OCA, the lack of pigmentation in the skin, hair, eye and optic nerves poses an increased risk of cutaneous neoplasms (9). For example, actinic keratosis has been reported to eventually develop in almost all albinos. Current data also suggest that non-melanoma skin cancer, particularly squamous cell carcinoma, may be more aggressive in patients with albinism compared to the general population (10).

Although OCA has been reported to occur in association with other tumors (besides skin cancer), to the best of our knowledge, our case represents the first association of OCA with a brain tumor, a conjunction that we believe to be incidental. The most striking feature in our patient was the peripheral presentation of central FNP, and if an MRI had not been obtained, a potentially serious diagnosis may have been missed.

Complete medical history and detailed physical examination should be obtained for every patient presenting with peripheral facial palsy, particularly in infants where cranial CT or MRI may be warranted to avoid a misdiagnosis.

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SAŽETAK

Periferna manifestacija središnje paralize facijalisa

Ömer Bektaş, Funda Kurt, Banu Katlan, Veli Korkmaz, Emine Suskan, Gülhis Deda

Paraliza facijalisa kod djece može biti prirođena ili uzrokovana neoplastičnim, infektivnim ili traumatskim stanjima. Iako se kao najčešći uzrok susreće idiopatska (Bellova pareza) facijalnog živca, paraliza facijalisa može biti prvi i jedini simptom neke teže osnovne bolesti. Prijašnja ispitivanja u dječjoj populaciji opisuju tumor kao uzrok paralize facijalisa u 2%-12% slučajeva. Jedan od najtežih propusta za kliničara može biti da jednostavno pretpostavi kako dijete s paralizom facijalisa ima Bellovu parezu, pa nije potrebna potpuna i sveobuhvatna procjena druge moguće etiologije. Ovdje se prikazuje slučaj dojenčeta s očno-kožnim albinizmom, koje je došlo dugotrajne periferne paralize facijalisa i u kojega je magnetskom rezonancijom sa zakašnjenjem utvrđen tumor mozga kao uzrok ovoga stanja. Kod našega bolesnika je najizrazitija bila periferna manifestacija središnje paralize facijalisa; da nije napravljena magnetska rezonancija, ova potencijalno teška dijagnoza mogla se previdjeti. Kod svakog bolesnika koji očituje perifernu paralizu facijalisa treba uzeti potpunu medicinsku anamnezu i napraviti temeljit fizikalni pregled, naročito kod dojenčadi gdje se tomografijom ili magnetskom rezonancijom glave mogu zasigurno izbjeći krive dijagnoze.

Ključne riječi: paraliza facijalisa; albinizam, očno-kožni; Bellova pareza; tumori mozga; dojenče