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Presentation of disseminated neuroblastoma mimicking Bell's palsy: a case report

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Abstract

Background Neuroblastoma is an embryological malignancy of neural crest cells that may have diverse presentations owing to direct effects, metastases, or paraneoplastic syndromes. Facial nerve palsy is an extremely rare presentation of neuroblastoma.

Case presentation A previously healthy 1-year-and-10-month-old Sinhalese girl presented with difficulty in closing her left eye for 2 weeks, preceded by deviation of her mouth to opposite side. Initial physical examination was normal, apart from left lower-motor-type facial nerve palsy. Initial laboratory investigations were also normal. A tentative diagnosis of Bell's palsy was made. However, magnetic resonance imaging of the brain was performed owing to the incidental detection of a painless bulge in the left zygomatic region with overlying skin bruising, in the absence of any preceding trauma.

Magnetic resonance imaging of the brain revealed a soft-tissue mass in the left lateral orbital wall, extending to the body of the sphenoid and bulging into the left anterior fossa. Contrast-enhanced computed tomography of the abdomen, performed subsequently, showed an intra-abdominal large paravertebral soft-tissue mass. Histology of the abdominal mass confirmed poorly differentiated neuroblastoma.

Conclusion Facial nerve palsy is well recognized to have a myriad of underlying etiologies, including hematological malignancies, solid tumors, and paraneoplastic syndromes. However, its timely diagnosis is often challenging in the absence of other supportive clinical features. These children need careful and thorough evaluation for malignancies before commencing steroids, especially when underlying malignancy is likely.

Keywords Disseminated neuroblastoma, Bell's palsy, Child, Case report

Background

Neuroblastoma is an embryological malignancy of neural crest cells [1]. Although the adrenal gland is the most common site, neuroblastoma can present in various ways, owing to direct effects, metastases, or paraneoplastic syndromes. Bone pain, limp, scalp lumps, mandibular lumps, and periorbital swelling are among such atypical presentations [2]. Therefore, a high degree of suspicion is required to identify neuroblastoma presenting in an atypical manner. Neuroblastoma has been reported to present very rarely with facial nerve palsy [3]. We present a child with disseminated neuroblastoma who initially presented solely with left-sided lower-motor-neuron-type facial paralysis, mimicking Bell's palsy.

Case presentation

A previously healthy 1-year and 10-month-old Sinhalese girl presented with difficulty in closing of the left eye for 2 weeks. Her mouth was observed to have deviated to right side, which was first noticed a few days before the eye involvement. There was no preceding

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history of facial trauma nor clinically apparent viral or bacterial infections. The parents did not seek medical advice initially, since the child was otherwise well and active.

On examination, left-sided lower-motor-neuron-type facial nerve paralysis was noted, with incomplete eye closure and deviation of the mouth to the opposite side. Lacrimation was, however, not impaired. Other cranial nerve examinations and neurological examination of the upper and lower limbs were normal.

The full blood count revealed a white blood cell (WBC) count of 7.4×10^3(normal range: $5–15\times10^3$), with neutrophils at 31% (normal range: 25–70%), lymphocytes at 64% (normal range: 30–70%), hemoglobin at 9.6 g/dL (normal range: 11–14 g/dL), and platelets at 200×10^9 (normal range: $150–450\times10^9$/L). The laboratory results showed that C-reactive protein (CRP) was 2.1 mg/L (normal range: 0–3 mg/L) and erythrocyte sedimentation rate (ESR) was 7 mm in the first hour. Serum electrolytes were within normal limits, with sodium at 141 mmol/L (normal range: 138–145 mmol/L) and potassium at 4.1 mmol/L (normal range: 3.4–4.7 mmol/L), and renal functions were normal, with serum creatinine at 0.21 mg/dL (normal range: < 0.7 mg/dL) and blood urea at 13 mg/dL (normal range: 11–39 mg/dL). Aspartate transaminase (AST) was elevated, at 155 IU/L (normal range: 15–37 IU/L), with a normal alanine transaminase (ALT) of 33 IU/L (normal range: 14–50 IU/L). Serum uric acid, at 3.8 mg/dL (normal range: 2.6–6 mg/dL), was normal. Lactate dehydrogenase (LDH) was elevated.

An assessment by the ophthalmologist revealed normal findings in the fundi. A tentative diagnosis of Bell's palsy was considered on the basis of normal hematological parameters and the absence of any abnormal cells in the blood picture. However, before commencement of steroid therapy, the clinicians decided to perform magnetic resonance imaging (MRI) of the brain to rule out space-occupying lesions in the brain, as a painless bulge with an overlying bruise at the left zygomatic region was noted. The parents denied any history of falls or trauma and recalled that the lump had been present for the previous week, with no evidence of progression in size.

MRI of the brain showed an area of thickening with a large soft-tissue component in the lateral orbital wall (2.2×3.1 cm), with evidence of periosteal reaction. The soft-tissue thickening involved the body of sphenoid, and it extended to the superolateral wall of the orbit, impressing on the left globe (Fig. 1). It was also bulging superiorly into the left anterior fossa, impressing on the inferior part of frontal lobe, and bulging laterally into the temporal region. Signal change was present in left mastoid air cells. The radiological appearance of the lesion was in keeping with either rhabdomyosarcoma or neuroblastoma.

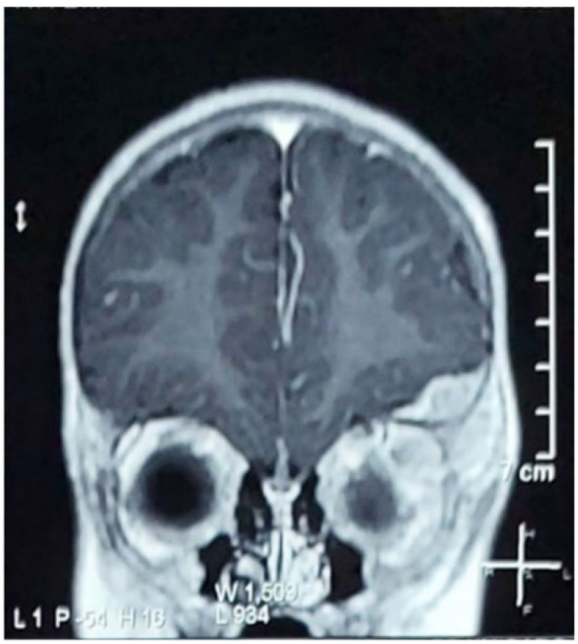

Fig. 1 Magnetic resonance imaging of the brain showing a soft-tissue mass arising from left lateral orbital wall, bulging laterally into left temporal region, and impressing on the inferior part of frontal lobe superiorly

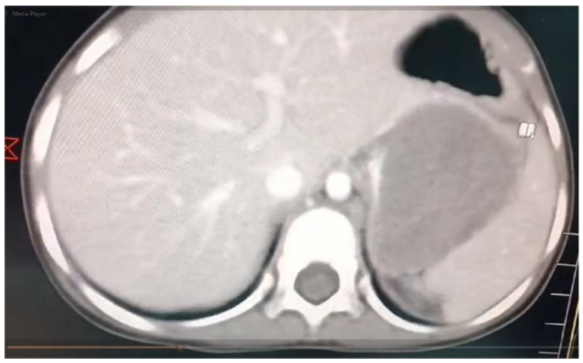

Fig. 2 Contrast-enhanced computed tomography of the abdomen showing a paravertebral soft-tissue mass lesion of 6.2 cm × 4.5 cm × 9 cm

The child soon underwent contrast-enhanced computed tomography (CECT) of the abdomen and chest (Fig. 2). CECT of the abdomen showed a paravertebral soft-tissue mass lesion of 6.2 cm × 4.5 cm × 9 cm with necrotic areas and calcific foci. The abdominal mass displaced abdominal structures; however, there was no evidence of local infiltration. Multiple areas of periosteal bony reaction were present in the pelvis, and bilateral common iliac and external iliac nodes were enlarged. Paravertebral soft-tissue masses extending

into adjacent spinal canal were noted on CECT of the chest.

A computed tomography (CT)-guided biopsy of the paravertebral mass was subsequently performed. The histology of the mass showed fragments of a poorly differentiated small blue cell tumor, with prominent neutrophils in the background, in keeping with a deposit of neuroblastoma. Calcifications, ganglions, and rhaboid differentiation were absent. Bone marrow involvement was excluded through bone marrow biopsy. The child was referred to the pediatric oncologist for treatment.

Discussion

Facial nerve palsy may occur owing to diverse etiologies with idiopathic Bell's palsy, trauma, and infections, which are among the most common causes reported [4]. Facial nerve palsy due to neoplastic lesions is rare. Local neoplasms such as rhabdomyosarcoma [5], Burkitt's lymphoma [6], neurinoma [7], and intracranial hemangioma [7] have been reported to cause facial nerve palsy in children, as has invasion of leukemic cells. Slow progression beyond 3 weeks, lack of return to normal by 6 weeks, single-branch involvement, bilateral palsy or unilateral and recurrent nerve palsy, and other nerve involvement are features suggestive of a possible neoplastic etiology.

Neuroblastoma is the most common extracranial solid tumor in childhood. It is a malignancy of neural crest cells. Atypical presentations have been reported, including Hutchinson syndrome, where bone and bone marrow metastases cause irritability or a limp in young children. The most common site of neuroblastoma is in the adrenal glands. Neuroblastoma of the head and neck is rare [8].

Although exceptionally rare, primary neuroblastoma of the facial nerve also has been described in one case report, in which it was located in the intratemporal portion of the facial nerve [9]. Given the resemblance of the presentation of that reported child to Bell's palsy, the child was initially commenced on steroid therapy. There had been over 2 months' delay in the suspicion of the possibility of a malignancy since the initial blood workup was normal, and no follow-up was arranged with the pediatrician. The possibility of malignancy was reconsidered only when the child re-presented with a mass over their right preauricular region, with ipsilateral hearing loss. However, in our patient, the final diagnosis was made 7 days after the initial medical checkup; this was owing to the presence of a small lump adjacent to the right zygomatic region with no plausible etiology and the consideration for performing neuroimaging on the initial patient encounter.

The metastasis of neuroblastoma causing recurrent facial nerve palsy has also been documented with an abdominal primary tumor [10]. Hearing impairment, bulging of tympanic membrane, or loss of stapedial reflex were associated with the facial nerve palsy in the prevailing documented presentations[3].

The facial nerve arises from the brainstem and passes laterally at the cerebellopontine angle to enter the internal auditory canal through the internal acoustic meatus [11]. After forming the geniculate ganglion and giving off the nerve to the stapedius and chorda tympani, it exits through the stylomastoid foramen and traverses the parotid gland to divide into branches. Therefore, when considering the course of the facial nerve and the location of soft-tissue mass, the facial nerve paralysis cannot be attributed to a direct effect by the mass. Two potential causes for this presentation could be manifestations of paraneoplastic syndrome and the presence of micrometastasis, which are not seen in MRIs of the brain. However, the precise mechanism for facial nerve palsy in this child remains to be explained.

Neuroblastoma is well known to be associated with paraneoplastic syndromes in children 13]. The presence of anti-Hu antibodies, which is associated with paraneoplastic neurological disorders, has been reported in children with neuroblastoma [12, 13]. Even though the common presentations are cerebellar ataxia and opsoclonus myoclonus, facial nerve paralysis as a paraneoplastic presentation of neuroblastoma should be considered in this patient, although no similar case reports have been described in literature.

Conclusion

Facial nerve palsy is well recognized to have a myriad of underlying etiologies, including hematological malignancies, solid tumors, and paraneoplastic syndromes. However, timely diagnosis is often challenging in the absence of other supportive clinical features. These children need careful and thorough evaluation for malignancies before commencing steroids, especially when underlying malignancy is likely.

Abbreviations

CECT Contrast-enhanced computed tomography
MRI Magnetic resonance imaging

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Not applicable.

Author contributions

KD, SG, and GJ participated in the literature survey, the making of the diagnosis, and the management of the child's case. SG and KD wrote the manuscript. KD edited the manuscript. All authors read and approved the final manuscript.

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Availability of data and materials

The data that support the findings of this case report are available from the Medical Records Department, North Colombo Teaching Hospital, but restrictions apply to the availability of these data, which were used under license for

the current report and thus are not publicly available. Data are, however, available from the authors upon reasonable request and with permission of the Medical Records Department, North Colombo Teaching Hospital, Sri Lanka.

Declarations

Ethics approval and consent to participate
Not applicable.

Consent for publication
Written informed consent was obtained from the patient's legal guardian for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Competing interests
The authors declare that there are no competing interests regarding the publication of this paper.

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