

Acute Myeloid Leukemia with Initial Presentation of Facial Palsy and Exophthalmos

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INTRODUCTION

Facial palsy is a relatively common disease and most of the patients have good outcome. The incidence of facial palsy is 15~40 per 100,000 inhabitants⁽¹⁾. The incidence of facial palsy in children is relatively low. Bell's palsy is the most common cause of facial palsy, the incidence of Bell's palsy is 20.2 per 100,000 populations in United Kingdom.⁽²⁾ However, some unremarkable initial presentation of facial palsy, mimicking Bell's palsy, is caused by a specific disease. We reported a 2-year-8-month-old girl with initial presentation of unilateral facial palsy and exophthalmos followed by rapid progression to bilateral side. The patient was previously diagnosed as Bell's palsy and was treated with steroids, and ultimately proven to have acute myeloid leukemia (AML). This rare presentation of acute myeloid leukemia should be kept in mind as a possibility in a patient with facial palsy and exophthalmos.

CASE REPORT

A 2-year-8-month-old girl was brought to our hospital due to asymmetrical facial expression, including decreased left frontal crease, left droopy eyelid and diminished left nasolabial fold, accompanied with left otalgia since 4 days ago. The patient also had bilateral protruding eyes for 9 days. Fever followed by cough and rhinorrhea were

associated. She visited a local clinic, and took antibiotic agent for left acute otitis media. An endocrine specialist for exophthalmos was consulted, too. Thyroid-stimulating hormone and free thyroxine were checked, and the data were within normal range. Oral prednisolone was prescribed under the diagnosis of Bell's palsy. Two days later, right facial palsy and drooling from mouth angle was observed. On the day of visiting our clinic, physical examination revealed bilateral peripheral type facial palsy and bilateral slight limitation of lateral external ocular motion. There was no obvious hepatosplenomegaly or lymphadenopathy.

On admission, fever flared up to 38.1°C. Laboratory findings revealed anemia with hemoglobin 5.4 gm/dL and mean corpuscular volume 109.9 fl. White blood cell count was 254200/μL, with 86% blasts, 8% lymphocytes, 2% neutrophils, 2% monocytes, 0% bands, 0% eosinophils, 0% basophils and 2% promyelocyte. Platelet count was 31,000/μL. Cerebrospinal fluid analysis was normal without leukemic cells or biochemical abnormalities. Brain CT showed hypertrophy of both lateral rectus muscles, which indicated leukemic infiltration in both orbital fossae (Figure). AML was diagnosed and the chromosome analysis of the leukemic cells revealed 47,XX,+6,t(8;21)(q22;q22).

The patient received transfusion of packed red blood cells and prestorage leukocyte-reduced apheresis platelets

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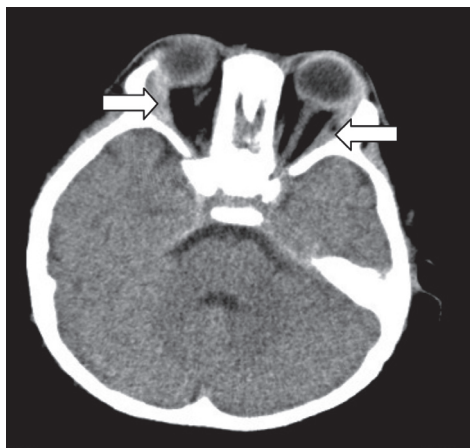


Figure. Both lateral rectus muscles became hypertrophy with heterogenous hyperdensity (as arrow), indicating leukemic infiltration, as known as granulocytic sarcoma or chloroma.

for anemia and thrombocytopenia on admission. The next day, she started TPOG-AML-97A induction therapy. Fever subsided after treatment. Exophthalmos and limitation of external ocular motion recovered quickly after 1 week chemotherapy. MRI was arranged twice but was not done successfully due to poor sedation and cooperation. The bilateral facial palsy, drooling and chewing difficulty improved gradually 5 months later. The patient was still under anti-leukemic therapy.

DISCUSSION

Bell's palsy is the most common cause of facial palsy in children, accounting for 39.5-78.6% of all the cases⁽²⁻⁴⁾. Nonetheless, clinicians need to figure out what is the cause of facial palsy and should not make a diagnosis of Bell's palsy until there is no evidence of other acquired conditions. Causes of acquired facial palsy include trauma, congenital, metabolic, infectious and inflammatory diseases, vascular abnormalities, and neoplasms.

Our patient visited neurologist for an unremarkable peripheral facial palsy as the chief complaint, which is a pitfall for clinicians to make a diagnosis of Bell's palsy. However, initial presentation with peripheral facial palsy and exophthalmos may indicate a more complications of central nervous system leukemia.

From 1984 to 2013, excluding the relapse of leukemia

and adult patients, eleven leukemic children with facial palsy as presenting symptoms were reported⁽⁵⁻¹²⁾. These patients are summarized in Table. The ages of the children ranged from 9 months to 17 years. The duration from the appearance of facial palsy to the diagnosis of leukemia varied from 1 day to 1 month, and was less than 1 week in seven cases. About the associated symptoms, seven patients had chloroma (or granulocytic sarcoma), six patients had otitis media or mastoiditis, four patients had fever. About the type of leukemia, eight patients had AML, and the other three had acute lymphoblastic leukemia (ALL). Three patients had bilateral facial palsy. About the prognosis of facial palsy, seven patients recovered, two patients improved, and one patient had no improvement.

Although ALL is 7-fold more prevalent than AML in children⁽¹³⁾ AML is 2.67-fold to ALL in children with facial paralysis according to this review. In addition, the most of these patients with AML (seven of eight) had chloroma. The chloromas were located close to the middle ear and infiltrated the facial nerve or surrounding temporal bone, mastoid bone and middle ear according to pathologic finding of operation⁽⁵⁻⁸⁾ or brain image⁽⁸⁻¹⁰⁾.

There are at least two possible explanations for the association between facial palsy and leukemia. One is a direct infiltration of the nerve with leukemic cells, and the other relates to a common etiologic infectious factor such as Epstein-Barr virus or human T-cell lymphotropic virus⁽⁵⁾. According to reviewed article, the infiltration of leukemic cells would not be obviously delineated through image study. Our patient had difficulty in MRI study due to unstable condition. Thus the brain CT may not trace leukemic cell exactly as technical limitation.

Leukemic cell infiltration at any site of the facial nerve or occurrence of chloroma in temporal bone may cause facial palsy. The entity of AML rather than ALL is a localized tumor composed of immature cells of the granulocytic line, it may be part of the reason why there is a higher prevalence of AML with facial palsy than ALL⁽¹⁴⁾.

We suggest a thorough physical examination, blood pressure, otoscopy examination and complete blood cell count when confront pediatric patients with facial palsy. In those with atypical associated symptoms or signs, e.g. bilateral facial palsy, exophthalmos, or other neurologic deficits, may especially need more detailed investigations. Children who diagnosed as Bell's palsy must be followed-

Table. Characteristics of leukemic children with facial palsy

Patient No.	Age at Diagnosis	Interval* Leukemia	Type of Findings	Additional facial palsy	Prognosis of No.	Ref.
1	13 yr	1 week	AML	Chloroma, mastoiditis	recovered	3
2	5.5 yr	1 day	AML	Chloroma, mastoiditis	recovered	4
3	6 yr	1 day	AML	Chloroma, otitis media, fever	Recovered	5
4	2Y1M	10days	AML	Chloroma, fever	Recovered	6
5	17 yr	1 month	AML	Chloroma, paraplegia	Recovered	7
6	11 mo	5 days	ALL	Bil. Facial palsy, Mastoiditis	Recovered	8
7	11 mo	5 days	AML	Ataxia	Not improved	8
8	6 yr	1 day	ALL	Prolonged fever	Improved	8
9	9 mo	3 weeks	AML	Chloroma, otitis media, FTT	Mild improved	9
10	13 yr	1 month	ALL	Bil. Facial palsy, nose bleeding, purpura	Recovered	10
11	2Y8M	4 days	AML	Bil. Facial palsy, Chloroma, otitis media, fever, exophthalmos,	Recovered	This article

*: Interval from onset of facial palsy to diagnosis of leukemia

Abbreviations: ALL: acute lymphoblastic leukemia, AML: acute myeloid leukemia, FTT: failure to thrive.

up and reevaluated if any abnormal symptoms or signs develop.

REFERENCES

- May M, Klein SR. Differential diagnosis of facial nerve palsy. *Otolaryngol Clin North Am* 1991;24:613-641.
- Rowlands S, Hooper R, Hughes R, Burney P. The epidemiology and treatment of Bell's palsy in the UK. *Eur J Neurol* 2002;9:63-67.
- Peitersen E. Bell's palsy: The spontaneous course of 2,500 peripheral facial nerve palsies of different etiologies. *Acta Otolaryngol* 2002;549:4-30.
- Shih WH1, Tseng FY, Yeh TH, Hsu CJ, Chen YS. Outcomes of facial palsy in children. *Acta Otolaryngol* 2009;129:915-920.
- Todd NW Jr, Bowman CA. Acute myelogenous leukemia presenting as atypical mastoiditis with facial paralysis. *Int J Pediatr Otorhinolaryngol* 1984;7:173-177.
- Levy R, Har-El G, Segal K, Sidi J. Acute myelogenous leukemia presenting as facial nerve palsy: A case report. *Int J Pediatr Otorhinolaryngol* 1986;12:49-53.
- Zappia JJ, Bunge FA, Koopmann CF Jr., McClatchey KD. Facial nerve paresis as the presenting symptom of leukemia. *Int J Pediatr Otorhinolaryngol* 1990;19:259-264.
- Kaufman BA, Jones L, Zutter MM, Park TS. Megakaryoblastic leukemia presenting as a temporal bone granunocytic sarcoma. *J Neurosurg* 1993;79:128-131.
- Eser B, Cetin M, Kontas O, Unal A, Er O, Coskun HS, Altinbas M. Facial nerve paralysis and paraplegia as presenting symptoms of acute myeloid leukemia. *Jpn J Clin Oncol* 2001;31:86-88.
- Krishnamurthy S, Weinstock AL, Smith SH, Duffner PK. Facial palsy, an unusual presenting feature of childhood leukemia. *Pediatr Neurol* 2002;27:68-70.
- Bilavsky E, Scheuerman O, Marcus N, Hoffer V, Garty BZ. Facial paralysis as a presenting symptom of leukemia. *Pediatr Neurol* 2006;34:502-504.
- Buyukavci M, Tan H, Akdag R. An alarming sign for serious diseases in children: Bilateral facial paralysis. *Pediatr Neurol* 2002;27:312-313.
- Tubergen DG, Bleyer A. The leukemias. In: Behrman RE, Kliegman RM, Jenson HB, eds. *Nelson textbook of pediatrics*, 17th ed. Philadelphia: WB Saunders; 2003:1694-1698.
- Kurabayashi H, Miyawaki S, Naruse T, Kubota K. Bilateral tympanic cavity infiltration with effusion in a patient with acute myeloblastic leukemia. *Annals of hematology* 1989;58:45-46.