Short report

Transient dystonia as a complication of varicella

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SUMMARY A case of transient post-varicella lingual-mandibular dystonia is presented. This case was compared with the eight previously reported instances of involuntary movement disorders which rarely follow varicella infection.

Neurological complications of varicella have been recognised since the mid-nineteenth century. These include meningoencephalitis, encephalitis, acute cerebellar ataxia, myelitis, Guillain-Barré syndrome and Reye's syndrome.¹ Amongst patients with the encephalitic form, several cases of chorea, athetosis, tremor and "spasms" of the face, extremities and trunk have been described.²⁻⁵ We recently examined a patient who developed transient lingual-mandibular dystonia following varicella infection.

Case report

The patient was a 17 year old right-handed Hispanic girl who developed typical varicella skin lesions following two days of malaise, headache and fever. On the fifth day following the eruption of the skin lesions, the patient developed involuntary upward eye deviation, tongue protrusion, jaw opening and inability to speak, persisting for four hours. Within 12 hours, the same phenomenon, save for eye deviation, returned for nine hours and was complicated by jaw dislocation. Treatment with intravenous diphenylhydramine was unsuccessful. A similar episode recurred for two hours the following day. Some pharyngeal constriction was also noted. All involuntary movements were exacerbated by attempting to speak.

There was no history of any drug ingestion, particularly neuroleptics, antihistamines or proprietary "cold" remedies.

Past medical, perinatal and developmental history were unremarkable. Family history was not significant for any neurological disease.

General physical examination was normal, save for crusted varicella skin lesions. Neurological examination was remarkable only for the dystonic movements described above.

Routine laboratory studies, including electrolytes, BUN, glucose, liver enzymes, serum ammonia, and full blood count, were all normal. Two cerebrospinal fluid samples were obtained; one with commencement of the involuntary movements and the second, 2 days later. The cerebrospinal fluid on both occasions was acellular with normal protein, glucose, and gamma-globulin levels. Viral cultures of the fluid were negative. An electroencephalogram was normal.

Two months later, the patient had remained well without further dystonic episodes. She has since been lost to follow-up.

Discussion

Neurological complications of varicella occur rarely, with an incidence less than one per 1000 cases.^{2,6,7} The development of abnormal involuntary movements in these cases is an even rarer occurrence, having been noted in seven of 120 cases reviewed by Underwood⁵ and three of 59 cases surveyed by Appelbaum, *et al.*² Little clinical description is available from the series of Appelbaum, *et al.*, so comparison with the current case is difficult.² Fortunately, six of the cases that developed involuntary movements following varicella that were reviewed by Underwood were previously reported in detail by others.^{5,7-11} Racaz and Lartigue¹² and Swan⁴ each reported one additional case of involuntary movements following varicella in sufficient detail. Therefore, there are eight previously reported descriptions of movement disorders following varicella which can be classified by current nomenclature. Two of the cases^{5,9} developed transient head tremor. Two cases of chorea were noted, one case was generalised¹¹ and the other was unilateral.¹² In both instances the chorea was noted to persist long after the acute illness. One case of persistent hemiballism was noted.⁸

Particular note should be made of Menko's case³ as

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the clinical features were strikingly similar to those of the current one. The patient was a 4 year old girl who developed prominent involuntary facial grimacing, accompanied by chorea-athetoid movements of the upper extremities 12 days after the exanthem. Within 2 days, tremor of the head and upper extremities, increased grimacing with involuntary eye closure, rhythmic tongue protrusion and slowed speech were noted. No other neurologic abnormalities were noted. All symptoms resolved in the subsequent 4 weeks.

The case of Sendrail and Dudeviant¹⁰ was a 7 year old boy who developed peri-oral grimacing, trismus, involuntary neck extension, impaired speech, choreoathetosis of the upper extremities and probable diaphragmatic chorea several days after the cutaneous eruption. This child also made a complete recovery. The authors coined the term "striatal varicella" to describe the clinical observations.

Swan's⁴ case was a 6 year old boy who developed an altered mental status with seizures 7 days after the onset of varicella. Examination of the spinal fluid revealed a lymphocytosis with normal protein. Approximately a week thereafter, the patient developed recurrent episodes of neck, spine, and upper extremity extension with bilateral flexion at the hips and knees without alteration in consciousness. Over the next week, the movements continued, accompanied by pyrexia and inability to clear secretions resulting in the patient's demise. Neuropathological examination demonstrated a scattered perivascular leptomeningeal lymphocytic infiltrate and a severe necrotising perivascular inflammatory infiltrate, with some demyelination, predominantly affecting the rostral putamen and caudate nucleus.

The above three cases were all characterised by dystonic movements.¹³ Ours may be readily added to this group, as it was very clearly a case of sustained involuntary contractions. Therefore, it appears that dystonia can be a rare neurological complication of varicella. In fact, dystonia may be one of the most common movement disorders to arise as a complication of varicella, as suggested by review of the literature. Presumably, the development of dystonia or other movement disorders associated with varicella does represent a low grade inflammatory infiltration

of the striatum that can occur without leptomeningeal involvement. The occurrence of a movement disorder with varicella encephalitis does not imply a grave prognosis for most patients.^{1 2 6 7}

References

- 1 McKendall RR, Klawans HL. Nervous system complications of varicella-zoster virus. In: Vinken PJ, Bruyn GW, eds. *Handbook of Clinical Neurology*, vol 34. Amsterdam, Netherlands: Elsevier Science Publishers, 1978:161-83.
- 2 Appelbaum E, Rachelson MH, Dolgopul UB. Varicella encephalitis. *Am J Med* 1953;15:223-30.
- 3 Menko MLHS. Choreiforme bewegungen nach varicellen. *Deutsche Medizinische Wochenschrift* 1899;25:745.
- 4 Swan C. A case of post-varicellar encephalitis showing bilateral softening of the neostriatum and terminal "tetanoid chorea" (Gowers). *Med J Aust* 1946; 1:697-702.
- 5 Underwood EA. The neurological complications of varicella. A clinical and epidemiological study. *Br J Child Dis* 1935;32:83-107,177-96,241-83.
- 6 Boughton CR. Varicella-zoster in Sydney: II. Neurological complications of varicella. *Med J Aust* 1966;2:444-7.
- 7 Johnson R, Milbourn PE. Central nervous system manifestations of chickenpox. *Can Med Ass J* 1970; 102:831-4.
- 8 Babonneix L, Adeline, Colombe. Un cas d'encephalite varicelleuse. *Gaz Med France*.1928;2:174-5.
- 9 Gautier P, Monedjikova V. Un cas d'encephalite varicelleuse. *Bull et Mem de la Soc. Med des Hop de Paris*. 1931;55:332-5
- 10 Sendrail M, Dudeviant O. Les formes choreoathetotiques de l'encephalite varicelleuse. *Bulletins et Memoirs de la Societe. Med des Hop de Paris* 1932;56:358-63.
- 11 Wilson RE, Ford FR. Nervous complications of variola, vaccinia, and varicella with report of cases. *Bull Johns Hopkins Hosp* 1927;40:337-53.
- 12 Rocaz, Lartigue. Varicelle et encephalite. *J Med Bourdeaux* 1921;92:326.
- 13 Fahn S, Eldridge R. Definition of dystonia and classification of dystonic states. In: Eldridge R, Fahn S, eds. *Advances in Neurology*, vol 14. New York: Raven Press, 1976:1-5.