## 소아기 붕괴성 장애의 양상을 보이는 PANDAS 1례

## A CASE OF PANDAS WITH CHILDHOOD DISINTEGRATIVE DISORDER

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요 약:	A (group	oup A beta-hemolytic streptococcus)				
Sydenham					가 ,	Α
		가	. A		. ,	
				가		
			PANDAS(pedia	atric autoimr	nune neuro	psychiatric
disorders associated	with streptococcal	infections)				
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가		PANI	DAS			
, PANDAS가	, ,	/	,		,	
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중심 단어 : A	· PANDAS ·					
М	론					
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	가		mmune neuro	psychiatric	disorders	associated with
(National Institute of Me	ental Health)		streptococcal			1)
,	A				(aroup A	beta - hemolytic
, 가	_		streptococcus		(3 - 5)	Syden
•	·		ham	,		274011

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Osler<sup>2)</sup>
                                                                                         Sokol<sup>21)</sup>
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          Husby 3)
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  Sydenham
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                                 . Sydenham
                   Sydenham
                                                    2. 현병력
   7 - 10)
                  , Swedo<sup>11)</sup>
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                                                                             98 1
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                 , Sydenham
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                  . A
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 13 - 18)
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 <sup>19)</sup> A
                                      - O(antis-
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treptolysin - O,
                  ASO)
                       가
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    PANDAS
                                                                    가
 . Peterson <sup>20)</sup>
                                           ASO
                                anti - DNAse B)
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          가 PANDAS
         Peterson <sup>20)</sup>
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                  anti - DNAse B
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                                         1998
                                                                        Brain
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. 3 risperidone 0.5mg 가 가 2~4 1~2 가 가 가 가 가 가 . 10 가 ASO 733 Brain MRI, . 10 가 Brain 57 SPECT), 30 가 , ASO(790), (FANA)(+, >1:160), 가 (Lupus anticoagulant) 35 risperidone 1mg ESR, CRP, ceruloplasmin, C3/C4 고 찰 가 1908 Heller <sup>23)</sup>. Heller 7.75 , 가 79. 5, 3~4 3 가 . Heller 가 100,000 5. 경과 및 치료 4~8 3 3~4 가 . DSM -ICD - 10 2 가 가 가 가 . 1~2

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          PANDAS
                                               clonal antibody) D8/17
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               Brain MRI, Brain SPECT,
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                                                          ASO
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ASO
        가 790
                  3
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     가
                             1:160
                                                     PANDAS
                                                  . Giedd <sup>26)</sup>
                                                                                       가
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                           , ESR, CRP, cerulo-
                                               PANDAS
plasmin, C3/C4
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                                                                                     (globus
                                                                    , Peterson <sup>20)</sup>
  Α
                                               pallidus)
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                   가
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                                   , PANDAS
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  PANDAS
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edo <sup>1)</sup> 1)
                                      2)
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       3)
                          4) A
                                                          Sydenham
        5)
                                                   PANDAS
                                                              27)
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가

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PANDAS		, Swedo , PANDAS
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PANDAS	,	,
가 Perlmutter	30)	A
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가 PANDAG		. DANDAC 71
PANDAS risperidone	<sup>32)</sup> 가	PANDAS 가 .
	risperidone	References
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rispridone , 1) 가 2) 7	3) 가	1) Swedo SE, Leonard HL, Garvey M, Mittleman B, Allen AJ, Perlmutter S, Louqee L, Dow S, Zamkoff J Dubbert BK (1998): Pediatric autoimmune neuropsychiatric disorders associated with streptococcal infections: clinical description of the first 50 cases. Am J Psychiatry 155: 264-271
ASO(790),	( , >1:160),	2) Osler W(1894): On Chorea and Choreiform Affections. Philadelphia, HK Lewis, pp33-35
, ASO 가7 A	33	3) Husby G, van de Rijin I, Zabriskie JB, Abdin ZH, Williams RC (1976): Antibodies reacting with cytoplasm of subthalamic and caudate nuclei neurons in chorea
5) , ,	PANDAS	and rheumatic fever. J Exp Med 144: 1094-1110 4) Swedo SE, Rapoport JL, Cheslow DL, Leonard HL
	Swedo	Ayoub EM, Hosier DM, Wald ER(1989): High pre-
	, PANDAS	valence of obsessive-compulsive symptoms in patients with Sydenham's chorea. Am J Psychiatry 46: 245-249
,		5) Swedo SE, Leonard HL, Schapiro MB, Casey BJ,
ASO 가	. ,	Mannheim GB, Lenane MC, Rettew DC (1993): Sydenham's chorea: physical and psychological symptoms of St. Vitus's Dance. Pediatrics 91: 706-713
,	,	6) Asbahr FR, Ramos RT, Negrao AB, Gentil V(1999): Case series; increased vulnerability to obsessive compulsive symptoms with repeated episodes of Sydenham's chorea. J Am Acad Child Adolesc Psychiatry

- 38:1522-1525
- 7) Rapoport JL(1989): The neurobiology of obsessive-compulsive disorder. JAMA 260: 2888-2890
- 8) Peterson BS, Riddle M, Cohen D, et al(1993): Reduced basal ganglia volumes in Tourette's syndrome using three-dimensional reconstruction techniques from magnetic resonance images. Neurology 43: 941-949
- 9) Giedd JN, Rapoport JL, Kruesi MJ, Parker C, Schapiro MB, Allen AJ, Leonard HL, Kaysen D, Dickstein DP, Marsh WL, Kozuch PL, Vaituzis AC, Hamburger SD, Swedo SE(1995): Sydenham's chorea: magnetic resonance imaging of the basal ganglia. Neurology 45: 2199-2202
- 10) Peterson BS, Tucker DM(1996): Neuroimaging in developmental neuropsychiatric disorders. In: Child and Adolescent Psychiatry, A Comprehensive Textbook. 2nd ed.(ed.) by Lewis M. Williams & Wilkins. Baltimore, pp.108-109
- Swedo SE(1994): Sydenham's chorea. A model for childhood autoimmune neuropsychiatric disorders. JAMA 272: 1788-1791
- 12) Kerbeshian J, Burd L, Pettit R(1990): A possible poststreptococcal movement disorder with chorea and tics. Dev Med Child Neurol 32: 642-644
- 13) **DiFazio MP, Morales J, Davis R**(1998): Acute myoclonus secondary to a case of acute beta-hemolytic streptococcus infection. J Child Neurol 13: 516-518
- 14) Kiessling LS, Marcotte AC, Culpepper L(1993): Antineuronal anti-bodies in movement disorders. Pediatrics 92: 39-43
- 15) **Kiessling LS, Marcotte AC, Culpepper L**(1994): Antineuronal anti-bodies: tics and obsessive-compulsive symptoms. J Dev Behav Pediatr 15: 421-425
- 16) Swedo SE, Leonard HL, Mittleman BB, Allen AJ, Rapoport JL, Dow SP, Kanter ME, Chapman F, Zabriskie J(1997): Identification of children with pediatric autoimmune neuropsychiatric disorders associated with streptococcal infections by a marker associated with rheumatic fever. Am J Psychiatry 154: 110-112
- 17) Kurlan R(1998): Tourette's syndrome and 'PANDAS'. Will the relation bear out? Neurology 50: 1530-1534
- 18) Singer HS, Giuloiano JD, Hansen BH, et al (1999): Antibodies against a neuron-like (HTB-10 neuroblastoma) cell in children with Tourette syndrome. Biol Psychiatry 46: 775-780
- 19) 정유숙(1998) : A군 연구균 감염이후 Antistreptoly-

- sin-O 혈중 농도 증가가 뚜렛씨 장 애의 증상악화에 미치는 영향에 대한 임상 연구. 소아청소년정 신의학 9:209-217
- 20) Peterson BS, Leckman JF, Tucker D, Scahill L, Staib L, Zhang H, King R, Cohen DJ, Gore JC, Lombroso P(2000): Preliminary findings of antistreptococcal antibody titers and basal ganglia volumes in tic, obsessive-compulsive, and attention deficit/hyperactivity disorders. Arch Gen Psychiatry 57: 364-372
- 21) Sokol MS(2000): Infection-triggered anorexia nervosa in children: clinical description of four cases. J Child Adolesc Psychopharmacol 10(2): 133-45
- 22) Mathew SJ(2001): PANDAS variant and body dysmorphic disorder. Am J Psychiatry 158(6): 963
- 23) Heller T(1908): Dementia infantilis. Zeitschrift fur die Erforschung und Behandlung des Jgenlichen Schwachsinns 2: 141-165
- 24) 조수철(1999) : 소아정신질환의 개념. 서울대학교 출판부, pp96-99
- 25) **Singer HS, Giuloiano JD, Hansen BH, et al** (1998): Antibodies against human putamen in children with Tourette syndrome. Neurology 50: 1618-1624
- 26) Giedd JN, Rapoport JL, Garvey MA, Perlmutter S, Swedo SE(2000): MRI assessment of children with obsessive compulsive disorder or tics associated with streptococcal infection. Am J Psychiatry 157: 281-283
- 27) Garvey MA, Perlmutter SJ, Allen AJ, Hamburger S, Lougee L, Leonard HL, Witowski E, Dubbert B, Swedo SE(1999): A pilot study of penicillin prophylaxis for neuropsychiatric exacerbations triggered by streptococcal infections. Biol Psychiatry 45: 1564-1571
- 28) Allen AJ, Leonard HL, Swedo SE(1995): Case study: A new infection-triggered, autoimmune subtype of pediatric OCD and Tourette's Syndrome. J Am Acad Child Adolesc Psychiatry 34: 307-311
- 29) Tucker DM, Leckman JF, Scahill L, Wilf GE, La Camera R, Cardona L, Cohen P, Heidmann S, Goldstein J, Judge J, Snyder E, Bult A, Peterson BS, King R, Lombroso P(1996): A putative poststreptococcal case of OCD with chronic tic disorder, not otherwise specified. J Am Acad Child Adolesc Psychiatry 35: 1684-1691
- 30) Perlmutter SJ, Leitman SF, Garvey MA, Hamburger S, Feldman E, Leonard HL, Swedo SE(1999): Therapeutic plasma exchange and intravenous immunoglobulin for obsessive-compulsive disorder and tic disorders in children. Lancet 354: 1153-1158

- 31) Leonard HL, Swedo SE(2001): Paediatric autoimmune neuropsychiatric disorders associated with streptococcal infection(PANDAS). Int J Neuropsychopharmacol 4: 191-198
- 32) Kleinsasser BJ, Misra LK, Bhatara VS, Sanchez JD(1999): Risperidone in the treatment of choreiform movements and aggressiveness in a child with "PANDAS". S D J Med 52(9): 345-347

- ABSTRACT -

— Korean J Child & Adol Psychiatr 12: 149~156, 2001 —

## A CASE OF PANDAS WITH CHILDHOOD DISINTEGRATIVE DISORDER

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After the fact that obsessive-compulsive symptoms or tic symptoms are common in Sydenham's chorea which is a sequale of rheumatic fever produced by group A beta-hemolytic streptococcus was reported, the association between group A beta-hemolytic streptococcus and a subgroup of obsessive-compulsive disorder (OCD) or tic disorder has been attentioned. This subgroup shared a unique clinical course, characterized by an abrupt onset of symptoms and/or dramatic exacerbations. And this subgroup was distinguished by pre-pubertal onset of symptoms, neurological abnormalities (choreiform movements and a unique pattern of motoric hyperactivity), as well as by relapsing and remitting symptom course. Acronym PANDAS (pediatric autoimmune neuropsychiatric disorders associated with streptococcal infection) was used to denote a subgroup of OCD or tic disorder patients with these clinical characteristics. Then, there was a report suggesting that PANDAS category include some of attention-deficit/hyperactivity disorder (ADHD) and were two case reports of anorexia nervosa and body dysmorphic disorder with characteristics of PANDAS.

This case is a patient who developed normally until age of 7, but after pharyngeal inflammation with high fever, he showed disturbance of cognition, social relationship, and language and communication, as well as tic symtoms and abnormal movement on face, hand, and foot.

We report this case with review of literatures, because we think that this case belongs to the PANDAS category. Based on this observation, we suggest that PANDAS category include some of childhood disintegrative disorder as well as OCD, tic disorder, and ADHD.

KEY WORDS: Group A beta-hemolytic streptococcus · PANDAS · Childhood disintegrative disorder.