



POSTER PRESENTATION

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Neuroblastoma versus systemic JIA – a diagnostic dilemma

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Purpose

Children with malignancies, including neuroblastoma (NB), may present to their primary physician with complaints that mimic those seen in systemic juvenile idiopathic arthritis (sJIA). The purpose of this investigation was to identify significant distinctions between patients with systemic JIA and neuroblastoma at the time of disease presentation.

Methods

A retrospective chart review was completed on all patients less than age 18, between 1990 and 2009 at Pennsylvania State University Children's Hospital, with a diagnosis of either sJIA or NB. The following variables

were identified at the time of presentation; gender, age, race, time to diagnosis, presenting signs and symptoms, neuroblastoma stage, LDH, uric acid, sedimentation rate, CRP, platelet count, hemoglobin, peripheral white blood cell count, percentage of neutrophils and lymphocytes, ferritin, and d-dimer. Other categorical information included whether or not the patients had gait disturbance or extremity pain, fever, rash, abdominal mass, arthritis, fatigue, or weight loss. Individual logistic regression models were evaluated for each variable, with sJIA or NB as the outcome. Given that patients with NB and extremity pain may be referred to orthopedics or rheumatology first, instead of a timely referral to hematology/oncology, a subgroup analysis was completed for those patients.

Table 1 Presentation characteristics of SJIA versus NB patients

Variables	sJIA (65)	Neuroblastoma (89)	
Age (years)	7.2 ± 4.7	3.0 ± 3.5	<0.0001
Gender	Female 51% Male 49%	Female 42% Male 58%	0.26
Race	White 91% Black 6% Hispanic 3%	White 85% Black 7% Hispanic 3% Other 5%	0.86
Fever present (0=no, 1=yes)	45/45 (100%)	12/71 (18%)	<0.0001
Time to diagnosis (months)	1.5 (65)	1.2 (78)	0.18
Arthritis	51/65 (78%)	1/71 (1%)	<0.0001
Rash	53/64 (83%)	2/71 (3%)	<0.0001
Abdominal mass	1/65 (2%)	29/71 (41%)	0.0001
Weight loss	11/65 (17%)	5/71 (7%)	0.07
Gait disturbance or extremity pain	45/65 (41%)	13/73 (18%)	<0.0001

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Table 1 Presentation characteristics of SJIA versus NB patients (Continued)

Neuroblastoma stage	n/a	Stage 1 = 19/62 (31%) Stage 2 = 3/62 (5%) Stage 3 = 9/62(14%) Stage 4 =31/62 (50%)	n/a
LDH (units/L)	637 ± 530 (35)	1842 ± 2410 (46)	0.005
Uric Acid (mg/dL)	3.3 ± 1.2 (22)	5.1 ± 2.9 (34)	0.005
Crp (mg/dL)	14.9 ± 25.3 (26)	10.2 ± 7.2 (4)	0.71
ESR (mm/hr)	81.6 ± 36.3 (58)	61.7 ± 46.0 (13)	0.09
PLT count	464,016 ± 173,148 (62)	327,419 ± 136,488 (62)	<0.00001
WBC	18,881 ± 11,449 (63)	10,484 ± 5,892 (68)	<0.00001
HGB (g,dL)	10.2 ± 1.80 (62)	10.3 ± 2.46 (67)	0.97
Neutrophil %	73 ± 14 (57)	0.49 ± 0.17 (64)	<0.00001
Lymphocyte %	17 ± 10 (47)	40 ± 17 (64)	<0.00001
Feritin (ng/mL)	4,016 ± 6, 135 (50)	159 ± 205 (29)	0.001
D-Dimer elevated	34/36 (94%)	2/2 (100%)	0.73
Aldolase	15.6 ± 11.4 (28)	8.85 ± 1.01 (2)	0.42

Results

There was a total of 65 sJIA and 89 NB patients. Demographically, sJIA patients were significantly older (7.2 vs. 3.0 years of age; $p < 0.0001$). Symptomatically, the sJIA patients had fever, rash, arthritis, and extremity pain or gait disturbance (41% vs. 18%) more frequently ($p < 0.0001$ for all). Laboratories were also significantly different between the groups. NB patients had higher LDH and uric acid levels, while sJIA patients had higher platelet counts, total peripheral WBC, had relative neutrophilia ($p < 0.0001$ for all), and had higher ferritin levels than the NB patients ($p = 0.001$), see Table 1. In the subgroup of 58 children with extremity pain or gait disturbance, 45 (78%) had sJIA and 13 (22%) had NB; see Table 2. NB patients had higher LDH ($p = 0.01$) and lower hgb levels ($p = 0.003$), while sJIA patients had a relative neutrophilia (76% versus 51%; $p < 0.0001$).

Table 2 Laboratory values for sJIA and NB subjects with gait disturbance or extremity pain

Laboratory	sJIA	NB	p-value
LDH	625 ± 522 (28)	1365 ± 1073 (9)	0.01
Uric Acid	3.2 ± 1.1 (20)	4.0 ± 1.2 (8)	0.14
CRP	9.9 ± 7.8 (22)	10.2 ± 7.3 (4)	0.95
ESR	84 ± 32 (42)	100 ± 32 (6)	0.24
Hgb	10.4 ± 1.7 (43)	8.5 ± 2.4 (12)	0.003
Neutrophil %	76 ± 12 (40)	51 ± 10 (11)	<0.0001
Lymphocyte %	15 ± 9 (40)	37 ± 12 (11)	<0.0001
Ferritin	4554 ± 6890 (37)	555 ± 135 (3)	0.34
d-dimer abnormal	27/20	1/1	0.79
PTT	35 ± 6 (30)	31 ± 4 (5)	0.15
Aldolase	16 ± 2 (24)	9 ± 8 (2)	0.44

Conclusion

There are significantly different demographic, presenting symptoms, and laboratories at the time of presentation of patients who are ultimately diagnosed with sJIA or NB. In particular, there are discerning laboratories that may help facilitate an appropriate and timely referral to oncology when extremity pain or gait disturbance are among the initial signs and symptoms.

Disclosure

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