

CASE REPORT-IV

Ewing's Sarcoma Presenting as Pyrexia of Unknown Origin in a Child with Cerebral Palsy

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ABSTRACT

Pyrexia of unknown origin is common in clinical practice. We report an eight year old boy suffering from cerebral palsy who presented with fever and nonspecific bone pain of six months duration. His physical examination revealed findings of spastic diplegia. Laboratory work up including blood biochemistry, hemogram and peripheral smear examination were within normal limits. His bone marrow examination showed malignant round cell tumour. Immunohistochemical stain for MIC-2 was positive and suggested a diagnosis of Ewing's sarcoma. Bone scan revealed increased tracer uptake in fifth lumbar vertebrae and left iliac crest. A final diagnosis of Ewing's sarcoma, originating from fifth lumbar vertebrae with bone metastasis was made. Such tumours have an overall poor survival rate in which high dose chemotherapy with peripheral stem cell rescue may be a future alternative.

INTRODUCTION

Patients with pyrexia of unknown origin are commonly encountered in clinical practice. In spite of a greatly increased armamentarium of diagnostic tests, in 43% of cases etiology is not clear. Malignancy is attributed as a cause in 2% of cases.¹ Ewing's sarcoma is a common bone tumour malignancy in children and young adults. We report a case of Ewing's sarcoma in a child with cerebral palsy presenting as pyrexia of unknown origin.

CASE: An 8 year old boy with cerebral palsy was admitted with a history of evening rise of temperature 102-103°F, without chills and rigors of 6 months duration, along with a vague intermittent back pain of 3 months duration. There were no symptoms related to pulmonary, gastrointestinal, urinary, and rheumatologic and central nervous systems. There was no history of contact with tuberculosis. He was born out of non consanguineous marriage as twin I (fraternal), preterm, spontaneous vaginal delivery at hospital with a birth weight of 1kg, cyanosed at birth; required oxygen for 2 hours, and Neonatal Intensive care unit stay for 4 days. Later gross motor milestones delayed, but social adaptive milestones were appropriate. Examination revealed a conscious, cooperative child with malnourished appearance. There was mild pallor, kyphosis, scissoring gate, and spastic diplegia. There was no significant lymphadenopathy, bony tenderness, skin rash or visceromegaly.

Investigations revealed hemoglobin 8.2gm/dl, total leukocyte count 3500/mm³, differential counts N37%,L60%,E3% and platelets 237,000/mm.³ Blood culture & urine culture were sterile. Widal test was negative. Liver and renal function tests were normal. X-ray film of chest, ultra sonogram abdomen, and contrast enhanced computed tomogram of chest & abdomen all were within normal. Antinuclear antibody and rheumatoid factor were negative.

Bone marrow performed from left iliac crest showed malignant round cell tumour infiltration. Immunohistochemical stain for MIC-2 positive, conclusive of Ewing's sarcoma. Bone scan revealed increased tracer uptake in fifth lumbar vertebrae and left iliac crest. So

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diagnosis of Ewing's sarcoma metastasis with probable primary in L5 vertebrae was made. Parents were offered treatment, but opted out in view of financial constraints.

DISCUSSION

This case is reported for highlighting three issues. One is the rarity of Ewing's sarcoma (ES) presenting as pyrexia of unknown origin and the practical difficulty in diagnosing at an early stage in a patient with previous neurological deficit; and lastly association of ES in one of twin child and with cerebral palsy.

Ewing's sarcoma of bone is after osteosarcoma the most common malignant bone tumour in children and young adults. In a series of 140 patients at the Mayo's clinic for Ewing's sarcoma, 96% patients presented with pain, 61% with a palpable mass, 16% with a pathological fracture and 21% with fever.²

Often a delay occurs between initial symptoms and diagnosis. In a report from Denmark, the duration of symptoms before diagnosis averaged 9.6 months, with a range of 4 weeks to 4 years.⁴ Common reasons for delay were intermittent nature of pain and pelvis localization of tumour site. The clinical course of ES was not steadily progressive but intermittent, which often misleads the doctor into believing that the condition was temporary.⁵ In our case besides intermittent back ache and localization of tumour at lumbar vertebrae; previous neurological deficit also contributed for a delay in diagnosis.

Ewing's sarcoma has been reported in siblings, although the incidence is very low.⁵ Re-

ports of association with skeletal abnormalities, genitourinary abnormalities, Down syndrome, and hereditary retinoblastoma exist, but are uncommon suggesting chance occurrence rather than a biologic basis.^{6,7} In a pub med review, according to best of our knowledge no case of ES has been reported in a cerebral palsy child and one of a twin.

CONCLUSION

Our experience with this case emphasizes the need to consider Ewing's sarcoma in any young child with pyrexia of unknown origin along with intermittent bone pain. Early diagnosis is highly desirable as early treatment dramatically affects prognosis.

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