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CASE REPORTS

AWARENESS OF THE PITFALLS IN CHILDREN WITH MUSCULOSKELETAL COMPLAINT

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ABSTRACT

Children with an underlying life-threatening condition may present as nonspecific musculoskeletal complaints. One should alert with a high index of suspicion not to miss or delay the diagnosis and treatment. We reported a patient who suffered from thoracic neuroblastoma and presented with refusal to walk and musculoskeletal pain. A simple chest X-ray picked up the large superior mediastinal neuroblastoma. Subsequent imaging revealed metastatic spinal cord compression, which required emergency surgical decompression.

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KEYWORDS

musculoskeletal pain, bone metastasis, spinal compression, mediastinal mass, thoracic neuroblastoma.

Introduction

Neuroblastoma is the most common extracranial solid tumour in childhood, accounting for approximately 8% of all childhood cancers and the global annual incidence of neuroblastoma is 0.85-1.1 cases per 100,000 children younger than 15 years old.[1] Diagnosis of neuroblastoma can be difficult sometimes as the symptoms may simulate other diseases. The most common musculoskeletal symptom of neuroblastoma is hip pain and neuroblastoma is often incidentally revealed as an abdominal mass in radiologic studies.[2,3,4] We reported a child with thoracic neuroblastoma who presented as a musculoskeletal complaint. This unusual presentation highlighted the importance of including childhood malignancy in unexplained musculoskeletal complaint differential diagnosis.

Case Report

A 4-year-old girl was admitted for bilateral leg pain and refused to walk. She had a fever up to 38.5^0 on the date of admission. Her body weight was 13.1 kg (3^{rd} centile), body height 96 cm (10^{th} centile), blood pressure was 94/65 mmHg, heart rate was 136 beats per minute and respiratory rate was 26 breaths per minute. Her oxygen saturation was 99% at room air. She complained of pain over both thighs and noticed a "limp" on walking in the past week. She complained of nonspecific back pain on and off for the past month. There was no skin rash, no swelling of joints and no history of trauma. The parents were not sure about any precipitating or relieving factors related to the pain and the patient could not describe the pain clearly.

Her past medical history was unremarkable and she was the only child of the family. A private paediatrician assessed her with laboratory tests performed. It showed a normal blood count and normal liver, renal function tests. The C-reactive protein was mildly elevated at 12.8 mg/dL (reference <5 mg/dL), erythrocyte sedimentation rate was elevated at 63 mm/h (reference <20 mm/h). Private ultrasound abdomen did not show any hepatosplenomegaly. The X-rays of hips, knees and ankles were normal.

Physical examination on admission showed that she was alert but was very irritable. She refused to sit up or walk. She had a right lateral neck mass, measured about 3x3 cm in diameter. She tilted her head to the left side at about 45 degrees and refused proper examination of the neck mass, presumably because of pain. Her face was not puffy. Her neck veins were not engorged. There was no stridor, no hoarseness of voice, no suprasternal or subcostal insucking to suggest any respiratory distress. On lower limb examination, there was no erythema, swelling or tenderness over her knees, ankles and upon observation, she could move her hips. Her muscle power was at least against gravity. The knee and ankles jerks were not brisk and plantar reflexes were downward.

Her blood tests showed haemoglobin 10.8 g/dL, WCC 5.5×10^9/L and platelets 175×10^9/L. C-reactive protein was 55 mg/dL. LDH was 1223 IU/L (reference 135-345 IU/L). Ferritin was 775 pmol/L (reference 9-151 pmol/L). There was no blast cell in the peripheral blood smear.

Her chest X-ray was alarming and revealed a large right mediastinal mass (Figure 1). An urgent thorax computerized tomography (CT) scan with contrast showed a large right upper posterior mediastinal mass with features suspicious of neuroblastoma. Superiorly, the mass extended above the thoracic inlet into right lower neck. There was no superior vena cava obstruction, but her right internal jugular vein was compressed and narrowed by cervical lymph node metastasis (Figures 2a and 2b). There was intraspinal invasion with thecal sac narrowing and cord compression at the T3-T4 level (Figures 3a and 3b). An urgent spine magnetic resonance imaging (MRI), showed a T2-T5 epidural tumour with cord compression (Figure 4) and vertebral metastasis at T1-T5, T8, T10, T11, L3, L5 and S1. She had emergent excision of the extradural intraspinal tumour. The tumour pathology confirmed neuroblastoma. She underwent a bone marrow biopsy, staging CT, bone scan and MIBG scan, which confirmed stage 4 neuroblastoma.

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Figure 1. Chest X-ray showed a large right superior mediastinal mass with trachea deviated to the left side. There was a linear calcification in the mediastinum of uncertain origin. The soft tissue over the right neck was increased. There were no bony erosions, no pleural effusion. The cardiac shadow was not enlarged.

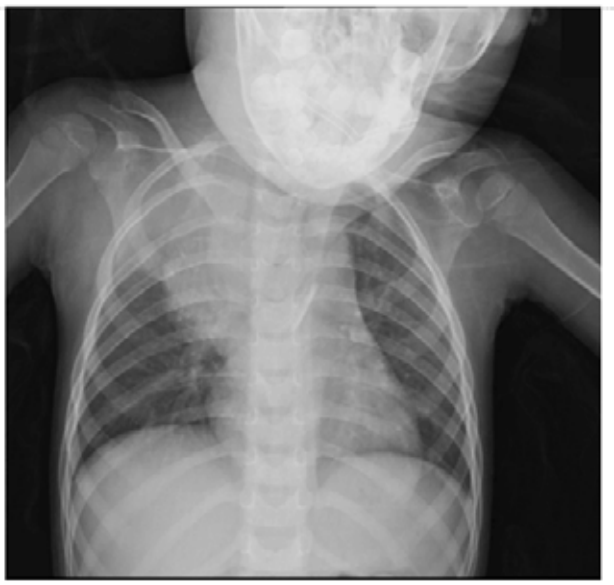


Figure 2a and 2b. CT Thorax. The coronal view of plain CT scan showed a large heterogenous superior-posterior mediastinal mass, measuring 8.4x4.3x7.4 cm with internal calcifications and necrosis (yellow arrow). The enlarged supraclavicular lymph node over the right side of neck, compressing on the right internal jugular vein (red arrow).

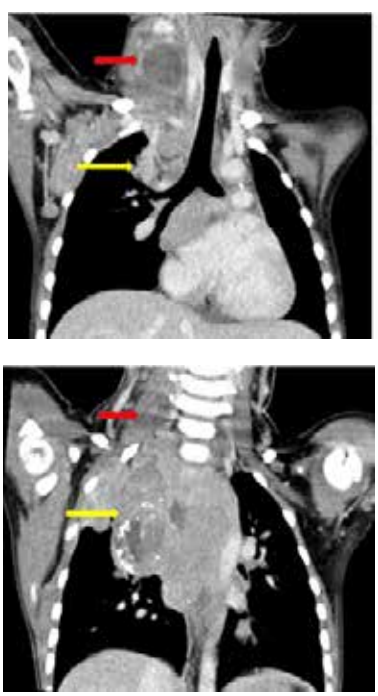


Figure 3a and 3b. CT thorax at the axial view. Figure 3a showed the axial view and lung view of plain CT thorax which showed the compression of the right bronchus by the mass (yellow arrow). Figure 3b revealed the thecal sac and spinal cord were compressed and invaded by metastatic tumor mass (orange arrow). There mass had diffuse calcification and necrosis.

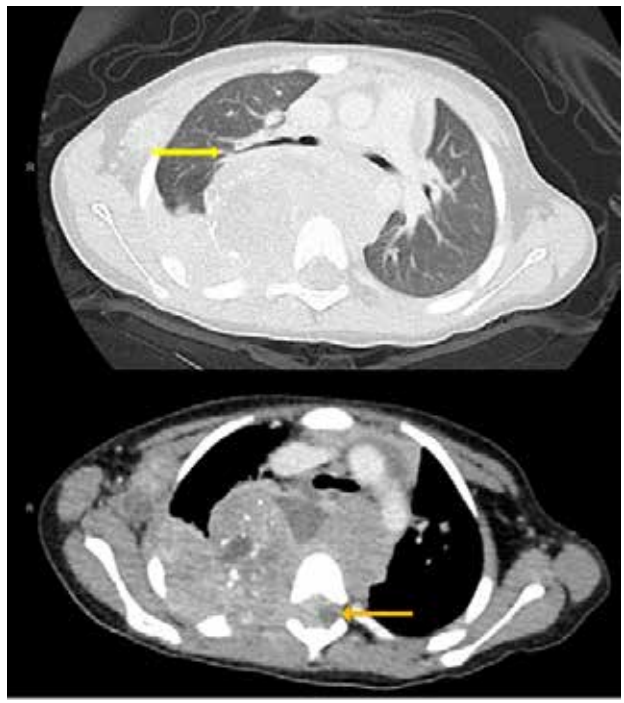
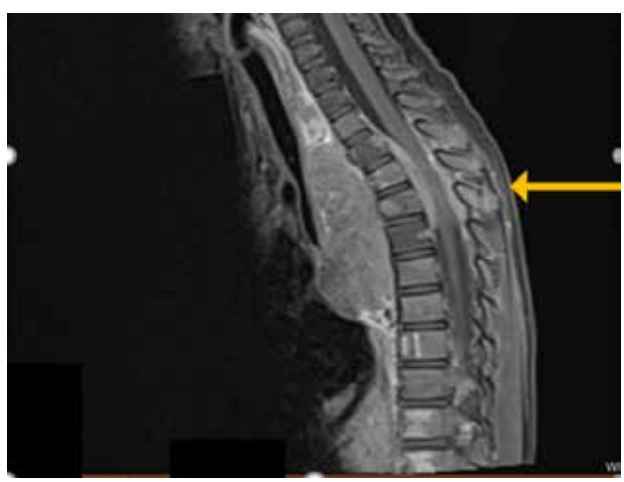


Figure 4. MRI spine sagittal view, showed the metastatic tumour with epidural invasion and cord compression at T2 to T5 (yellow arrow).



Discussion

Neuroblastoma is malignant embryonal tumor derived from neural crest cells which form the adrenal medulla and the sympathetic nervous system. Neuroblastoma is predominantly a childhood malignancy, with more than 90% of cases diagnosed in infants and children under 5-years old and the median age is 2 years. Age >18 months is a poor prognostic factor. Compared with 41% of infants, about 80% of patients diagnosed after one year of age have stage 3 or 4 diseases.[5] Neuroblastoma accounts for approximately 15% of childhood cancer mortality.[1]

The clinical presentations of neuroblastoma are extremely variable, including asymptomatic, fever, failure to thrive, abdominal pain, abdominal mass, diarrhoea, sweating, hypertension, opsoclonus, myoclonus, Horner syndrome, paraplegias, limp and

bone pain. Bone marrow infiltration, presenting in 80% of metastatic patients, could be very painful. Some studies reported that about 22% of presentations of neuroblastoma mimic a variety of orthopaedic conditions, ranging from a limp to paralysis of the lower limbs.[2,3,4] Children who refuse to bear weight may be because of pain, neuromuscular weakness, or specific mechanical factors. The most critical clinical dilemma is distinguishing life-threatening causes such as septic disease, malignant disease or neurologic emergency from those benign conditions.[6] For children with rheumatic disease, the pain is usually low to moderate intensity, usually in the morning with the stiffness of joints. In young children, less than 5 years old, atypical pain without an origin may warrant a urine test and radiologic studies to rule out neuroblastoma. In some previous studies, neuroblastoma origins from the abdomen was also an important differential diagnosis of atypical musculoskeletal pain over the lower limbs and abdomen ultrasound is a sensitive screen image.[2,3,4]

However, we should also be aware of neuroblastoma originating from extra-abdominal sites since a prompt diagnosis help to ensure well planning of diagnostic and treatment process, minimise irreversible neural damage and may improve survival rate. While about 72% of primary neuroblastoma arises in the abdomen, thoracic neuroblastoma only accounts for 13%.[7] Thoracic neuroblastoma was often incidentally found as a mediastinal mass on chest X-ray done for a patient with respiratory symptoms, such as cough, dyspnoea, dysphagia and lung infections. Our patient had thoracic neuroblastoma with a tricky presentation, which general practitioners or paediatricians could have easily overlooked as she presented with a musculoskeletal complaint instead of respiratory symptoms. Musculoskeletal examinations were essentially normal in this child. The focus had been on her thigh pain, leg pain, back pain and limping since her initial presentation to the private sector. With a history of fever at admission, an infective origin of arthritis would be a differential diagnosis. The pain she experienced was probably neuropathic pain secondary to her cord compression. Her "refusal to walk" was due to muscle weakness and neuropathy instead of "muscle pain".

An unyielding musculoskeletal examination and a neck mass alerted us to rule out more sinister causes for the patient's complaint. The girl's chest X-ray revealed a large superior mediastinal mass with trachea deviation and the presence of nonspecific calcification also pointed to the possibility of a malignant tumour, though teratoma could be a differential diagnosis.

There was a medical emergency to exclude superior mediastinal syndrome. We had to assess the severity of the mass effect urgently. Its compression over the superior venous cava might cause poor venous return and jeopardise the patient's cardiac output. The compression over the trachea and bronchi might cause respiratory distress and failure requires ventilatory support. Hence, an urgent assessment for the cardiopulmonary function of the child was needed. It was fortunate that the cardiopulmonary function of this child was preserved. Besides the mediastinal mass syndrome, thoracic neuroblastoma frequently complicates with metastatic bony secondaries causing spinal cord compression, which is also an oncology emergency as in our patient.

A recent national-wide study over 23 years in Denmark showed that 7% of the 3895 children with all types of cancer had musculoskeletal diagnoses six months before detecting the cancers.[8] The symptoms could be related to direct malignant cell invasion to the muscle, bone or the bone marrow, intra or periarticular bleeding or even paraneoplastic reactions. Whereas, a 7-year review over 3528 children presented to a paediatric rheumatology centre, 0.25% of patients were finally diagnosed to have an underlying malignancy with acute leukaemia as the most common cancer detected.[6] A musculoskeletal complaint can represent a broad spectrum of causes, including rheumatological, trauma-related, chronic disease features and malignancies.[9] Paediatricians have to be familiar with these unusual presentations and be able to triage the life-threatening disease.

Conclusion

We presented a child with musculoskeletal pain due to thoracic neuroblastoma and metastatic spinal cord compression. The initial presentation and chief complaints may be misleading. One has to be alert about this unusual presentation and includes malignancy as a differential diagnosis in childhood musculoskeletal pain.

Compliance with Ethical Standards
Funding : None
Conflict of Interest : None

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