Clinical Case Report

Thoracic neuroblastoma presenting as recurrent empyema

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ABSTRACT

Neuroblastoma is the most common intra-abdominal and extracranial solid tumour in children, accounting for 7%–8% of all childhood cancers. It is a malignant tumour of the autonomic nervous system derived from the neural crest. Most children with neuroblastoma have distant metastatic disease at the time of diagnosis. Pulmonary metastasis at the time of diagnosis is rare, and rarer is the presence of associated pleural effusion. We present the case of a child with recurrent empyema, who was diagnosed to have a thoracic neuroblastoma.


INTRODUCTION

Most children with neuroblastoma have distant metastases at the time of diagnosis.1 Pulmonary metastasis at the time of diagnosis is rare, as is the presence of an associated pleural effusion. The common symptoms are non-specific and include abdominal distension, abdominal pain, fever, constipation, diarrhoea, decreased appetite, loss of weight or neurological symptoms (paraparesis, myoclonic jerks, opsoclonus). We managed a child who presented with recurrent empyema, and was later diagnosed to have a thoracic neuroblastoma.

THE CASE

A 2-year-old boy from Nepal presented with respiratory distress and fever for 2 weeks. There was a history of similar complaints 6 months ago. A chest X-ray at that time showed an opaque left hemithorax. An intercostal tube was inserted and 125 ml of purulent fluid was drained. A diagnosis of pneumonia with left-sided pleural effusion was made. The chest X-ray following drainage of the effusion showed partial clearance with persistence of upper- and middle-zone opacities. The child improved after receiving intravenous antibiotics. Six months later the symptoms reappeared. A chest X-ray showed an opaque left hemithorax. With a diagnosis of recurrent empyema, the child was started on antibiotics and a contrast-enhanced CT (CECT) scan of the chest was done. It showed collapse and consolidation of the left lung with pleural thickening and minimal fluid in the pleural cavity. The child was referred to our centre for further management.

At admission, the child had a pulse rate of 140/minute, respiratory rate of 60/minute with subcostal and intercostal retraction and his blood pressure was 100/50 mmHg. Oxygen saturation (SpO₂) on room air and with oxygen supplementation using a face mask were 89% and 97%, respectively. Pallor was present without clubbing, lymphadenopathy or organomegaly. Anthropometry was within normal limits. There was fullness on the left side of the chest with grossly decreased air entry and a dull percussion note. Examination of other systems was normal. Differential diagnoses included loculated empyema, tuberculosis, non-resolving pneumonia and immune deficiency.

The total white blood cell count was 15 400/cmm with 75% polymorphonuclear cells. A chest X-ray showed an opaque left hemithorax with mediastinal shift to the right (Fig. 1A). Renal and liver function tests, and evaluation for immunodeficiency including HIV infection were within normal limits. The intercostal tube drained 50 ml of pus mixed with blood. Biochemical examination of the pleural fluid revealed protein 100 mg/dl, sugar 40 mg/dl and 20 cells, with 80% polymorphonuclear cells. These findings were suggestive of an empyema, but the Gram-stain was negative and the culture was sterile. Pleural fluid cytology was negative for malignant cells. Pleural fluid lactate dehydrogenase level was 485 i.u./L (normal 100–330 i.u./L) and polymerase chain reaction (PCR) for Mycobacterium tuberculosis was negative. Blood culture was sterile. A repeat chest X-ray showed persistence of upper- and middle-zone opacities (Fig. 1B). An ultrasound of the chest showed a lobulated ill-defined mass-like thickening of the left pleura with a mild pleural effusion causing collapse of the left lung. Ultrasound of the abdomen was normal. A CECT scan of the chest showed a solid mass involving the left hemithorax, possibly pleural-based and encasing the blood vessels but not compressing them (Fig. 1C). There was no calcification and the ribs were not involved. The radiological differential diagnoses included pleuro-pulmonary blastoma, mesothelioma, lymphoma and neuroblastoma. A CECT scan of the abdomen showed a pleural-based lobulated soft tissue mass in the left hemithorax extending and causing encasement of the aorta and involvement of the left crus of the diaphragm with likely involvement of the left adrenal gland.

On 111I MIBG scan, there was increased radiotracer uptake in the chest mass along with enhancement of the left adrenal gland. A positron emission tomography–computed tomography (PET-CT) scan was suggestive of active disease in the lymph nodes (paratracheal, suprACLavicular) and in the underlying collapsed lungs (Fig. 1D). A bone scan showed heterogeneous radiotracer uptake in the left thoracic mass without any evidence of skeletal metastasis. Bone marrow aspiration and biopsy were normal. An ultrasound-guided biopsy of the chest mass was done which showed a malignant small round cell tumour with a diffuse fibrillary background and focal rosette formation. The tumour cells were immunopositive for chromogranin and synaptophysin and negative for CK, myogenin, desmin and MIC-2. The final histopathological diagnosis was neuroblastoma. His urine examination was normal.

The child was started on chemotherapy (cisplatinum, doxorubicin, cyclophosphamide and etoposide) with 5 cycles being given every 4 weeks. Follow-up evaluation showed a normal chest X-ray, normal CECT of the chest and abdomen and a normal PET scan. Consolidation chemotherapy and bone marrow transplantation was planned for this patient.
of clinical presentation when first hospitalized; absence of lung parenchymal disease; and negative bone marrow, urine and cytology results.

A malignant pleural effusion is rare in patients with neuroblastoma. A malignant pleural effusion results in an upgrade of tumour stage in extrathoracic neuroblastoma to stage 4 and in thoracic neuroblastoma to stage 2B. Since survival estimates for INSS stage 4 neuroblastoma and its treatment differ significantly from those of lower stages, detection of a malignant pleural effusion is of importance. Currently, the imaging studies recommended by the INSS for abdominal neuroblastomas are antero-posterior and lateral chest X-rays. CT/MRI scanning is necessary if an abnormality is identified on chest X-rays or if the abdominal disease extends into the chest.

Our patient presented with recurrent empyema. The cause of empyema, whether primary or secondary to the tumour, could not be ascertained since no organism was isolated from the pleural fluid. To conclude, any patient with recurrent pleural effusion/empyema with persisting abnormalities on the chest X-ray should be investigated further to ascertain the underlying cause. (The prognosis of thoracic neuroblastoma does not change whether or not there is an associated pleural effusion.)

REFERENCES


DISCUSSION

To the best of our knowledge, a thoracic neuroblastoma presenting as recurrent empyema has not been reported previously. The unusual aspects of this patient’s illness were the location of the tumour (which is considered to be an uncommon site); the mode