Case report

Incidental diagnosis of thoracic ganglioneuroblastoma in a 3 years old female with wheezing

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Abstract. Ganglioneuroblastoma (GNB) is a cancer of the peripheral sympathetic nervous system and represents the 30% of cases of Neuroblastoma. When originates from thoracic ganglia it may appear very late, with severe symptoms like respiratory distress or neurological dysfunctions. We present an incidental diagnosis of thoracic GNB, discovered by a chest radiography during a recurrent wheezing unresponsive to bronchodilators. The early diagnosis allowed a precocious treatment probably improving the outcome. (www.actabiomedica.it)

Key words: incidentaloma, ganglioneuroblastoma, recurrent wheezing

Introduction

Incidentaloma is defined as a mass (more often an adrenal tumor) discovered during a radiological examination performed for other reasons. Most of these, found in adults, are benign non-functioning adenomas. On the contrary, in infancy and childhood, the most common incidental mass is the Neuroblastoma (NB): a malignant neural crest tumor that could be aggressive in the rate of its growth and also in its relationship to surrounding structures. Autopsy prevalence of adrenal tumors in the general population is 6% (1, 2) indeed prevalence by Computed Tomography (CT) varies from 2.5 to 4% for CT abdomen, and 4.2% for CT thorax, in adult populations (3, 4). In literature data about Incidentaloma's prevalence in childhood are not established and few cases are described. In this report we present a case of thoracic Ganglioneuroblastoma in a 3 years old female "incidentally" discovered in exams for recurrent wheezing.

Case presentation

XX., female, three-years old came to our Pediatric Emergency Unit because of fever, caught and rhinorrhea. It was the second access in two months for the same symptoms. Examinations revealed dyspnea ($SatO_2$ 95%, tachypnea, subcostal and intercostal retractions), strong bilateral reduction of ventilation and some crackles. Three puffs series with salbutamol (300 mcg) 20 minutes one for another and administration of betamethasone (2,5 mg) did not significantly improve the symptoms. Following internal guide lines, a chest x-ray was performed. It detected lung thickening with well-defined margins arcuate aspect that projected for about 5 cm in the back of the left paramedian cardiac cranio-caudal.

The patient was hospitalized and a Computed Tomography (CT) showed a solid mass in posterior mediastinum at the left cost-phrenic angle compatible with Neuroblastoma. Abdominal ultrasound was negative for abdominal and retroperitoneal masses.

To refine the diagnosis a spinal-thoracic-abdominal Magnetic Resonance Imaging (MRI) was performed clearly showing the location of the mass and its relationship with the surrounding structures (Fig. 1). The MIBG and SPET scintigraphy showed a slight accumulation of the radio-drug in the left posterior mediastinal mass (Fig. 2). It was also performed a Neuron-Specific Enolase dosage resulted mild posi-

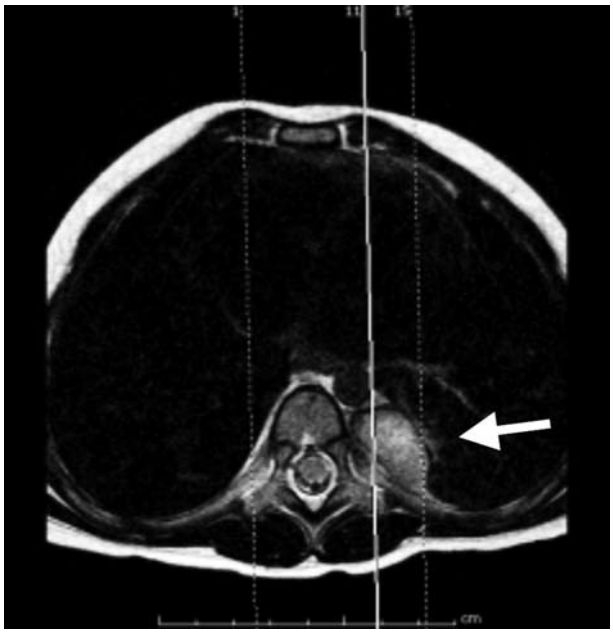

Figure 1. MRI: Solidal fusal neoformation in the left paraspinal mediastinum, between D8-D10 (maximum diameter: 3.5 cm)

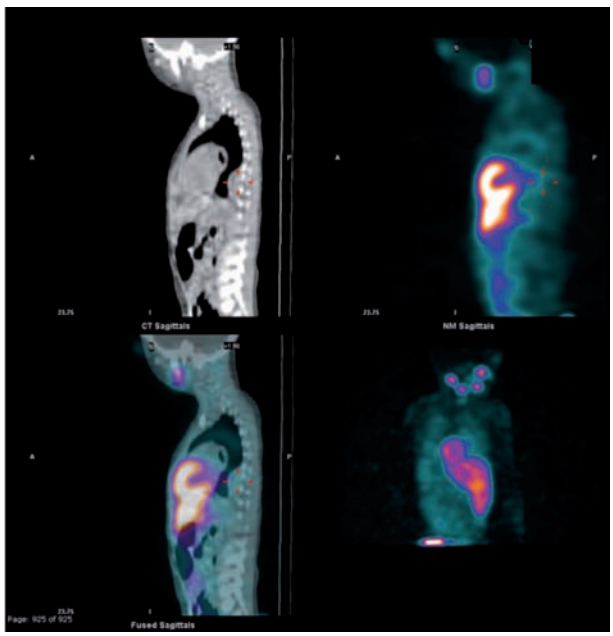

Figure 2. MIBG and SPET scintigraphy: slight accumulation of radio-drug in mediastinal mass

tive (30.5 mcgr/l), like the dosage of Vanilly-mandelic Acid in urine (31.1 mg/day).

The mass was removed through thoracoscopy after extemporary biopsy that shown "Neuroblastic peripheral tumor with many items with ganglionar morphology". Subsequent histological examination allows a diagnosis of classic nodular ganglioneuroblastoma.

After the diagnosis, XX. started the first cycle of chemotherapy with cyclophosphamide, doxorubicin, etoposide, carboplatin following the "NB unresectable" protocol.

The patient followed the therapy for six months, and now she is included in a 5 years follow-up program.

Conclusion and discussion

Ganglioneuroblastoma (GNB) is a cancer of the peripheral sympathetic nervous system and represents the 30% of NB's cases in children (5). It originates from cervical, thoracic or pelvic ganglia and its histological features are more differentiated than NB (undifferentiated small round cells/mature ganglion cells). It has a relative good prognosis even in children older than one year (6, 7).

NB is the most common extra-cranial solid cancer in children, in fact it represents about 8% of childhood malignancies (about 30 new diagnosis per year in Italy - M:F=1,3:1) (8, 9). About 93% is diagnosed in children younger than 6 years and the median age at diagnosis is 2 years. It is also one of the most common tumors in neonates often with prenatal diagnosis (10, 11). Most primary tumors are located in the abdomen, half of which are borne by the adrenal medulla. Other common sites of disease include: neck, chest and pelvis. Signs and symptoms of presentation are highly variable and depend in part on the primary tumor but also by the presence/absence of metastases or paraneoplastic syndromes like cerebellar ataxia or opsoclonus/myoclonus (11). These tumors have an unexplained tendency to metastasize in orbit bones doing orbit bruising and proptosis and a spread to the central nervous system may be observed with the evolution of the disease. Sometimes patients show hypertension, renin-mediated, due to impaired renal vasculature. Finally, 5% of patients < one year present the 4S disease: a small primary tumor (I or II stadium)

with metastases to liver, skin and bone marrow, which almost always spontaneously regress (11).

Only few papers are present in literature about NB's epidemiology in children, in particular about the symptoms at onset. Furthermore works describe groups of few cases, and no general data about pediatric symptoms at onset are described.

A work of the Pediatric Oncology Group (12) studied ninety-six pediatric patients with thoracic NB: they found that 14% of the patients presented with acute respiratory distress at diagnosis.

Another study (13) shows that in a group of 46 patients affected by non-metastatic thoracic Neuroblastomas, 57% of them were symptomatic. The most frequent clinical findings were cough (18%) and thoracic x-ray was the most useful diagnostic method (89%).

The third work we have studied (14) shows that in a group of 43 mediastinal neurogenic tumors the 86% presented symptomatically with: cough (30%), dyspnea (21%), wheezing (11.6%).

All these data, even if are not "universal", show that a mediastinal mass could manifest itself in many different ways, also with respiratory symptoms like cough, wheezing and respiratory distress.

Physicians who work in an Emergency Unit must remember that, back to an asthma-like symptomatology that often do not answer to the therapy, could be the presence of an extrinsic mass that compresses airways.

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