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Primary neuroblastoma: a rare cause of a retropharyngeal mass in a neonate

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Abstract

Neuroblastoma is the most common extra-cranial solid malignancy in children and the most common tumour occurring during infancy. This tumour arises from undifferentiated precursor cells of the sympathetic nervous system. The abdomen (65 per cent) is the most common site for these tumours, followed by the throat (15 per cent), pelvis (5 per cent) and cervical region (5 per cent).

We report a case of primary retropharyngeal neuroblastoma in a three-week-old baby boy presenting with upper airway obstruction

Key words: Neuroblastoma; Pharyngeal Disease

Introduction

A retropharyngeal mass is an uncommon cause of upper airway obstruction in the paediatric patient. Most retropharyngeal swellings are secondary to infection with subsequent abscess formation. Occasionally, a rare or unusual cause of retropharyngeal swelling is found.

Case report

A three-week-old baby boy was brought to hospital with a six-day history of noisy breathing. He was pyrexial with a temperature of 37.5°C. On examination, grade one stridor, 'tracheal tug' and subcostal recession were present. His chest was clinically clear. Examination of his oropharynx revealed a smooth swelling of the right postero-lateral pharyngeal wall.

A fibre-optic nasendoscopy was performed which revealed a retropharyngeal swelling with a normal looking larynx. A lateral neck X-ray showed a large retropharyngeal soft tissue mass extending from the base of skull to the level of the fifth cervical vertebra (Figure 1).

The most likely diagnosis was thought to be a retropharyngeal abscess, but transoral incision and drainage under general anaesthesia did not reveal any pus, and a tissue biopsy of the mass was performed.

A computed tomography (CT) scan was subsequently performed and showed a $3.5 \times 1.7 \times 3$ cm solid mass extending from the anterior margin of the foramen magnum to the level of the fifth cervical vertebra. The nasopharyngeal lumen was almost completely obliterated and the trachea and oesophagus were displaced to the left (Figure 2).

The full blood count, urea and electrolytes were normal and a human immunodeficiency virus (HIV) screen was negative.

A formal examination of the pharynx under anaesthesia was performed and a repeat biopsy was taken as the initial biopsy was inconclusive. The baby remained intubated overnight and developed airway obstruction after extubation the following day. A tracheostomy was performed to establish a definitive airway.

The histology result revealed a neuroblastoma. Because of the size and location of the tumour it was not amenable to surgery and the child was referred to the oncology team for possible chemotherapy. Abdominal, chest and cervical imaging studies alongside bone marrow biopsy revealed the tumour to be confined to the retropharyngeal space with no evidence of distant spread. The tumour responded well to combination chemotherapy (vincristine, cyclophosphamide and doxorubicin) and the patient was able to be decannulated three months later. Subsequent CT scans revealed a small ($1.3 \times 0.9 \times 2$ cm) residual retropharyngeal swelling. At the time of writing the patient was planned to be followed up at regular intervals by repeat CT scanning.

Discussion

If intracranial neoplasms are excluded, 5–10 per cent of all the malignant solid tumours in children occur in the head and neck region.[1] Lymphomas and soft tissue sarcomas are the most frequent neoplasms in children.

Neuroblastoma, a malignant embryonic tumour of the nervous system with neuroblastic differentiation, is the fourth most common malignancy in children.[2,3] This malignancy accounts for 7.8 per cent of childhood cancers worldwide. The majority (70 per cent) of neuroblastomas diagnosed in the head and neck region are metastatic lesions to the cervical lymph nodes, retro-orbital area or cranial bones.[4]

Five to ten per cent of primary neuroblastomas occur in the head and neck region. Primary neuroblastoma of the

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neck usually arises in the cervical sympathetic ganglia.[5] The origin and migration patterns of neuroblasts during fetal development explain the multiple anatomic sites where these tumours occur. They occur predominantly in the first three years of life with the highest incidence in the first six months.[6]

Patients usually present with a smooth, solid, rubbery, painless mass in the neck or pharynx. Dysphagia, stridor, cyanosis, aspiration during feeding and Homer's syndrome can be present.[7]

The patient should undergo a staging workup along with surgical resection or biopsy as appropriate. There are three different staging systems, namely the Children's Cancer Study Group system (CCG), the Paediatric Oncology Group system (POG) and the International Neuroblastoma Staging system (INSS). In general the CCG staging system is based on clinical findings, whereas the POG system is clinicopathologic. The INSS system uses features of both the CCG and POG systems.

Treatment of malignant tumours, especially in children, can be very complex. It necessitates co-operation between oncologists, chemotherapists, radiotherapists and surgeons, with a highly specialized histopathologist as the link between them. Surgical resection plays an important role in the treatment of patients with neuroblastoma. Patients with localized, resectable neuroblastomas have excellent event-free survival rates with surgical excision alone.[8] For patients with regional or metastatic disease, surgery to establish a diagnosis and obtain adequate samples for biologic studies is essential. Typically, in these patients second-look surgery following chemotherapy is employed to attempt a complete resection. The emphasis in the second-look procedure is on achieving as complete a debulking of the tumour as possible without sacrifice of major organ function.

Irresectable or intermediate- to high-risk patients should receive multimodality therapy including chemotherapy, surgery and, in selected cases, radiation therapy.

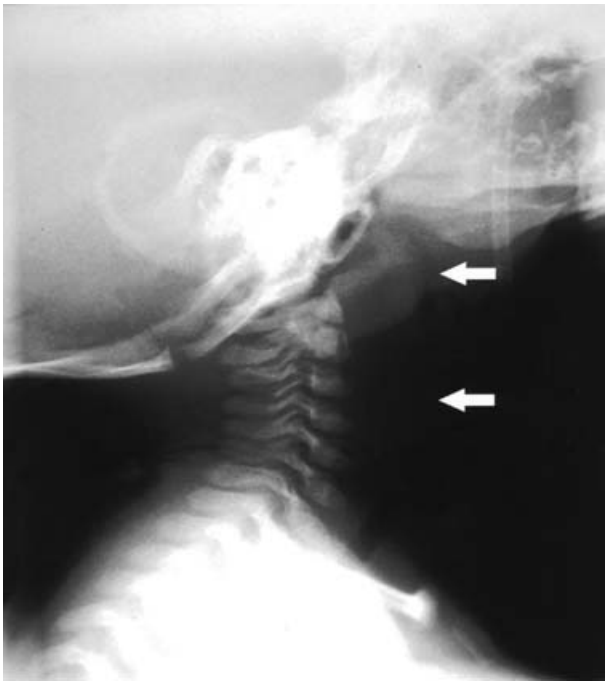

FIG. 1

Lateral projection of the cervical spine demonstrating striking increase in upper cervical prevertebral soft tissue (arrows).

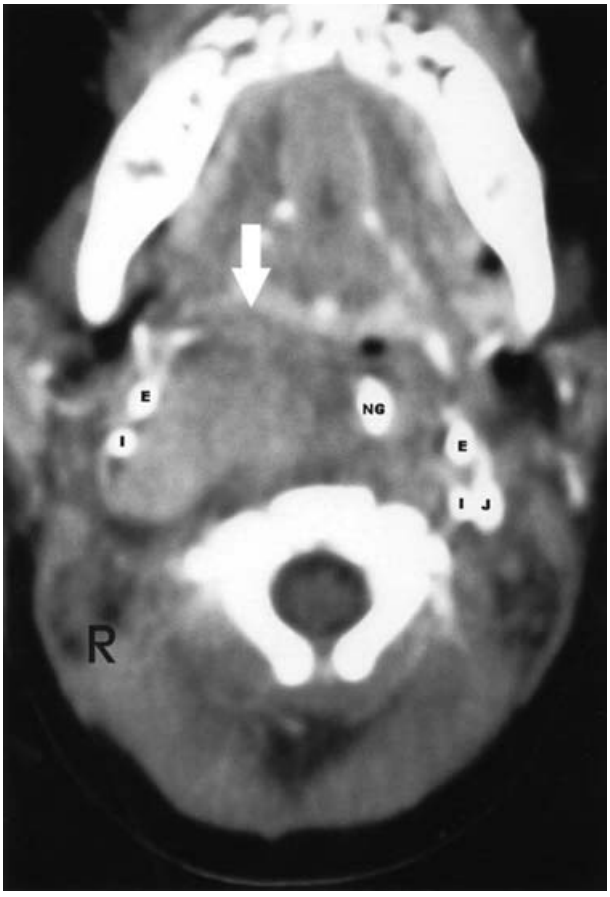

FIG. 2

Computed tomography scan through the oropharynx after administration of intravenous contrast demonstrating an inhomogeneously enhancing, retropharyngeal mass on the right (arrow). Note the ventral and lateral displacement of the right internal (I) and external (E) carotid arteries. The oropharyngeal airway is almost completely obliterated (defined by the nasogastric tube, NG) and shows ventral and left lateral displacement. J = left internal jugular vein.

Elevated catecholamines causing hypertension are present in 90 per cent of patients with adrenal involvement. Markers associated with a poor prognosis are elevated levels of ferritin, serum lactate dehydrogenase (LDH) and serum neuron-specific enolase (NSE).

The differences in outcome for patients with neuroblastomas are striking. Infants younger than one year have a good prognosis, even in the presence of metabolic disease, whereas older patients with metabolic disease fare poorly. Patients with localized tumours (regardless of age) have an excellent outcome (80–90 per cent three-year event-free survival rate).[9]

Conclusion

A retropharyngeal swelling is a rare cause of upper airway obstruction in the neonate. The majority of these swellings are abscesses and are successfully treated with incision, drainage and antibiotics. Occasionally, a tumour of the retropharynx may present in a similar manner. In such cases a tissue biopsy should be performed in order to make a histological diagnosis before deciding on further management.

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