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Case Report

A case of neuroblastoma presenting only in the form of cervical lymphadenopathy

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Abstract

Neuroblastoma is a cancer of the peripheral sympathetic nervous system, derived from embryonic neural crest cells and is one of the few cancers known to undergo spontaneous regression from an undifferentiated state to a benign tumour. It generally arises in the adrenal medulla or sympathetic chain in a child. Metastasis of neuroblastoma occurs mainly through lymphatic and haematogenous routes. This case report is that of a male child who presented only with cervical lymph node enlargement and histopathological examination diagnosed the lesion as neuroblastoma secondaries to cervical lymph node.

Keywords: Neuroblastoma, adrenal, metastasis, lymph node

1. Introduction

Neuroblastoma is the prototypic example of small blue cell tumour of childhood[1] and is the most common extracranial solid tumour in children accounting for 7-8% of all childhood cancers.[2,3,4] The prevalence of neuroblastoma is about 1 case per 7,000 live births.[2] It is slightly more common in boys with a male to female ratio of 1.1:1.[5] It can arise anywhere throughout the sympathetic nervous system, most common primary site being adrenal gland followed by abdominal (extra-adrenal), thoracic, cervical and pelvic sympathetic ganglia.[2] Approximately 1% of patients present initially with evidence of metastatic disease without a readily identifiable primary lesion.[6] In such cases, immunohistochemistry can aid in the diagnosis as neuroblastoma cells express neuron specific enolase, neurofilaments, chromogranin, synaptophysin and many other neural related antigens.[7,8]

2. Case report

A four and half year old male child presented only with right cervical lymph node enlargement with no other history or clinical findings. On physical examination, the child appeared to be otherwise healthy with normal vital parameters except for the enlarged and matted right cervical lymph nodes measuring 2x2cm in the neck. Systemic examination revealed no findings clinically. The provisional diagnosis of the treating surgeon was tuberculous lymphadenitis. The child was subjected to cervical lymph node excision biopsy and the excised tissue specimen was sent in 10% formalin for histopathological examination. Grossly the specimen comprised of five nodular grey white tissue bits ranging from 2x1.5x1cm to 0.5x0.5x0.3cm. The larger bits were cut into two and all the tissue bits (ten in number) were embedded in five blocks (A,B,C,D,E). After routine processing of the sections, paraffin blocks were made, cut and stained with the haematoxylin and eosin stain and studied under light microscope. Microscopically, all the bits showed lymph node tissue infiltrated with sheets and clusters of small, round cells separated by neurofibrillary matrix (Figure 1) which also formed the centre of rosettes (Homer Wright rosette) (Figure 2). The nuclei were round, deeply staining and slightly larger than lymphocytes. The cytoplasm was scanty with poorly defined cytoplasmic outlines. Areas of necrosis were also observed. Immunohistochemistry for neuron specific enolase (NSE) was done which showed diffuse strong positivity (Figure 3). The final histopathological diagnosis was given as cervical lymph node with secondaries from neuroblastoma. After obtaining the histopathological diagnosis, the treating surgeon asked for an ultrasonography of abdomen of the child which showed a small suprarenal mass on the right side measuring 2x1.5x1.5cms which denoted the primary site of tumour as adrenal gland. No other organs were found to be involved ultrasonographically. Bone marrow aspiration and biopsy was negative for secondaries.

Figure 1: Photomicrograph of lymph node showing lymphoid tissue (thick arrow) with infiltration by small round blue cells separated by neurofibrillary matrix (thin arrow) (H&Ex100).

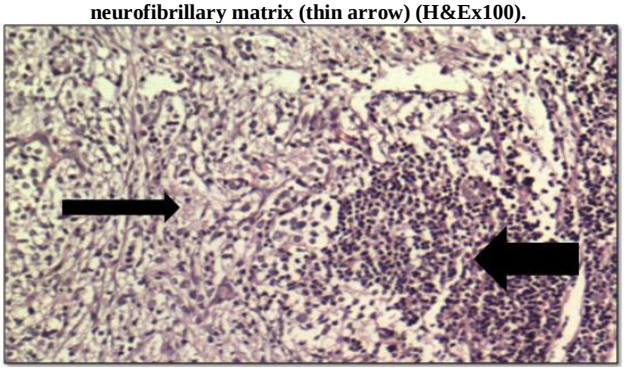


Figure 2: Photomicrograph showing sheets of small round blue cells with scant cytoplasm, round, deeply staining nuclei in a fibrillary matrix which forms the centre of Homer Wright rosette (arrows) (H&Ex400).

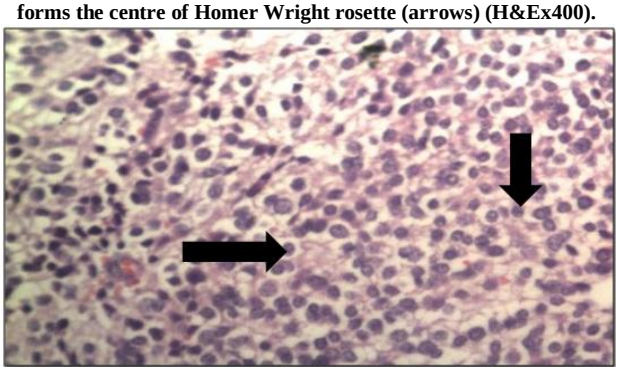
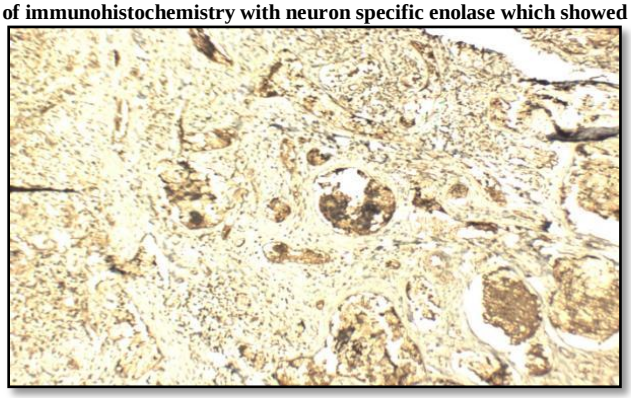


Figure 3: Photomicrograph of immunohistochemistry with neuron specific enolase which showed strong positivity (H&Ex400)



3. Discussion

Most primary neuroblastomas occur within the abdomen (65%).[2]Neuroblastoma commonly metastasizes to bone (60%), regional lymph nodes (45%), orbit (20%), liver (15%), intracranially (14%), and lung (10%).[9] Regional lymph node metastasis was noted in up to 35% of patients with apparently localised tumours. Spread of tumour to lymph nodes outside the cavity of origin is considered to be INSS (International Neuroblastoma Staging System) Stage IV disease.[2]

Presenting signs and symptoms of children with neuroblastoma reflect both the location of the primary tumour and the extent of disease. Patients with localised disease are often asymptomatic, while those with metastatic disease typically appear ill at presentation with systemic symptoms, including fever and bone pain secondary to tumour dissemination.[10]

Even though regional lymph nodes are first to be involved, the present case did not show any regional lymphadenopathy on ultrasound except a small tumour in the suprarenal region on the right side which was the primary. The usual presentation of neuroblastoma is in the form of an abdominal mass first noted by the parents. Rare cases have been reported associated with watery diarrhoea, Cushing's syndrome, Horner's syndrome (in cervical and mediastinal primary tumours) or other paraneoplastic syndromes. There are no case reports of adrenal neuroblastoma presenting only as cervical lymphadenopathy without a palpable mass in the abdomen or abdominal symptoms or any other endocrine syndromes.[11]

In the present case, the primary tumour detected by ultrasonography in the adrenal gland was removed surgically and the patient had an uneventful postoperative period; however the child failed to report for further follow-up.

In conclusion, since there are not many reports of neuroblastoma secondaries presenting as cervical lymphadenopathy with no palpable primary tumour within the abdomen or any other symptoms, it was decided to publish this case to bear in mind the differential diagnosis of neuroblastoma in a child presenting with cervical lymphadenopathy.

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