Case 2051

B-cell non-Hodgkin's lymphoma

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Section: Paediatric radiology
Imaging Technique: CT

Case Type: Clinical Cases
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Patient: 4 years, female

Clinical History:

The patient presented with a one-month history of general weakness, decreased appetite, fever, cervical lymph node enlargement, and right-sided exophthalmos. Laboratory tests showed leukocytosis and neutrophilia.

Imaging Findings:

CT with intravenous contrast medium showed a tumour mass in the left infratemporal fossa extending into the left maxillary sinus, spreading to the oral and nasal cavities, involving the posterior pharyngeal wall and extending into the middle cranial fossa with destructive lesions involving the bones of the skull base, maxilla and hard palate. Both orbits were involved due to enlargement and abnormal enhancement of the lateral rectus muscles resulting in bilateral proptosis. Large nodal masses was noted on the left side of the neck and under the head of the pancreas with enlarged paracaval lymph nodes. Multiple non-enhancing lesions were visualised in the enhanced and enlarged kidneys. Enlarged paravertebral soft tissues and lytic destructive lesions involved the vertebral bodies Th6-Th8, Th11-Th12, S1-S2 and the right ischium and pubis bones. There was a large epidural soft tissue mass on the level S1-S2 which deviating the thecal sac to the right and involved the nerve roots. Also infiltration of the right obturator internus muscle was seen.

Discussion:

Lymphomas are primary neoplasms of the immune system and arise within lymphoid tissue [1]. There are two main types of lymphoma. Hodgkin's lymphoma or Hodgkin's disease was identified in 1832 by Dr Thomas Hodgkin. All other types of lymphoma are called non-Hodgkin's lymphoma (NHL). Because lymph tissue is found in many parts of the body, lymphoma can start nearly anywhere. The lymph nodes become enlarged and the cancer can spread through the lymph system. In children lymphoma is the third most frequent malignancy, following behind leukaemia and central nervous system neoplasia.

NHLs are a much more heterogeneous group of malignant tumours than Hodgkin's lymphomas. A working
formulation divides the NHLs into low-, intermediate-, and high-grade groups. Low-grade lymphomas are usually follicular, while the intermediate- and high-grade tumours are usually diffuse [2]. The NHLs of childhood, in contrast with those of adults, are usually diffuse, extranodal, high-grade tumours. To eliminate the confusion created by multiple classification schemes, three primary subtypes of high-grade NHL have been defined: small noncleaved cell (SNCC), lymphoblastic, and large cell. The SNCC NHLs (Burkitt and non-Burkitt subtypes) are B-cell tumours that express surface immunoglobulin and contain one of three characteristic chromosomal translocations. Lymphoblastic lymphomas are usually of T-cell origin and may contain a translocation involving a T-cell receptor gene. Large cell NHLs occur as T-cell, B-cell or non-B, non-T-cell phenotypes.

The presenting signs and symptoms of NHL in children are largely determined by disease site and extent. The most frequent primary sites are the abdomen (31.4%), mediastinum (26%), and the head/neck region, including Waldeyer ring and/or cervical lymph nodes (29%). Noncervical lymph nodes are the primary sites in 6.5% of cases with skin, thyroid, epidural space, and bone accounting for the remainder (7%). There is a striking association between histological subtype and disease site. Lymphoblastic NHL usually occurs in the head and neck region or the anterior mediastinum; SNCC primary tumours arise in the abdomen and/or the head and neck; and large cell NHL may present in any anatomical location.

Head and neck primaries are usually painless masses arising from the cervical lymph nodes or tonsils. Mediastinal masses may be associated with pleural effusions, respiratory distress, or superior vena cava syndrome (swelling of the arms, neck, and face). Abdominal masses usually arise from the ileocaecal region and may be associated with abdominal distention, nausea, vomiting, or change in bowel habit, a clinical picture similar to appendicitis or intussusception. Bone marrow involvement may cause anaemia or thrombocytopenia and central nervous system disease may result in headache, increased intracranial pressure, or cranial nerve palsies [3].

The diagnostic and staging work-up of a child with suspected NHL must be expeditious because of the rapid growth rate of these tumours. A tissue diagnosis is necessary before treatment is started. Excisional biopsy is usually sufficient to evaluate an isolated peripheral node. A mediastinal mass can be evaluated by thoracotomy or mediastinoscopy, or thoracentesis (if there is an associated pleural effusion). An open biopsy is usually necessary for abdominal masses. Diagnostic imaging studies include CT of the primary site, chest, abdomen, and pelvis; bone scan; and (in some settings) gallium-67 scan. CT is the standard method of imaging the thorax and abdomen and the head and neck; CT is the single most useful staging investigation [4,5].

In the present case, extension of the tumour to the nervous system and kidneys and massive involvement of bones and soft tissue was due to late presentation of this patient.

**Differential Diagnosis List:** B-cell non-Hodgkin lymphoma

**Final Diagnosis:** B-cell non-Hodgkin lymphoma

**References:**


Description: Contrast-enhanced CT showing tumour mass in the left infratemporal fossa extending into the left maxillary sinus, spreading to the nasal and oral cavities (red arrows) with destructive lesions involving the maxilla and hard palate. Origin:
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Figure 2

Description: Contrast-enhanced CT showing enlargement and abnormal enhancement of the lateral rectus muscles resulting in bilateral proptosis. Also an enhancing soft tissue mass is seen in the middle cranial fossa parasellar and near the great wings of sphenoid bone. Origin:
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Description: Contrast-enhanced CT showing a large nodal mass on the left side of the neck, displacing vessels and compressing the internal jugular vein. Origin:
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Figure 4

Description: Multiple non-enhancing lesions are seen in the enhanced and enlarged kidneys. Origin:
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Description: Enlarged paracaval lymph nodes. A nodal mass is noted under the head of the pancreas (red arrow). Origin:
Description: Enlarged paravertebral soft tissues and lytic destructive lesions involving the vertebral bodies Th6-Th8, Th11-Th12, S1-S2 and the right ischium and pubis bones are seen. There is a large epidural soft tissue mass on the level S1-S2, deviating the thecal sac to the right and involving the nerve roots. Also infiltration of the right obturator internus muscle is seen. Origin:
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