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Wilms' tumour with spinal cord involvement

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ABSTRACT

Spinal cord involvement of Wilms' tumour is rare. A 14-year-old girl presented with an abdominal mass, paraplegia and loss of bladder and bowel control. Radiological investigations confirmed the presence of a large intraabdominal mass with infiltration into the spinal canal with impingement of nerve roots and the spinal cord. Histopathological evaluation demonstrated a nephroblastoma. It was decided to commence prompt neoadjuvant chemotherapy to render the tumour amenable to surgical resection. The patient unfortunately demised before receiving her first dose. Early diagnosis and timeous initiation of treatment is critical in limiting morbidity and mortality associated with malignant spinal cord compression.

1. Introduction

Wilms' tumour (WT) is the most common malignant renal tumour in children accounting for 6% of all neoplasms. Approximately 12% of cases are metastatic at presentation, with the lungs, lymph nodes, liver, and bone being the most common sites of metastases. Spinal cord involvement of WT is exceedingly rare, and we report a rare case and review the relevant literature.

2. Case presentation

A 14-year-old girl with no prior medical history was brought into our unit reporting a 6-month history of abdominal discomfort and paraplegia. She had first noticed a fullness in the abdomen six months ago, progressively worsening. In addition, she reported weakness of her lower limbs which had progressed over the last month to complete paraplegia with loss of bladder and bowel control. She appeared chronically ill, wasted, pale, and severely malnourished on physical examination. A large, firm, painless mass was palpable in the left upper quadrant and the left flank. She had flaccid paraplegia with 0/5 power in both legs and absent tendon reflexes. Contrast-enhanced CT of the abdomen and pelvis (Fig. 1) revealed a large cystic and partially solid mass (21 x 14.5 × 14 cm) occupying the left upper quadrant and left flank region, extending into the pelvis caudally and crossing the midline. No recognizable normal left renal tissue could be distinguished from this lesion (Fig. 1). In addition, multiple pulmonary metastases were identified. MRI (Fig. 2) revealed significant infiltration of the tumour into the spinal canal with significant impingement of numerous exiting nerve roots and impingement of the spinal cord from T9-L1. An urgent percutaneous biopsy of the lesion was performed on admission.

Histopathological examination represented a renal neoplasm comprising papillary structures covered by primitive, simple columnar epithelium (Fig. 3A). The tumoural cells contained scant apical eosinophilic cytoplasm and oval hyperchromatic nuclei with increased nuclear-to-cytoplasmic ratios (Fig. 3B). Nucleoli were inconspicuous. Some of the tissue cores had focal atrophic native renal parenchyma. No blastema or stromal elements were demonstrated. Anaplasia was not seen. Immunohistochemistry revealed strong and diffuse labelling with cytokeratin 8/18, PAX8 and WT-1 (Fig. 3C) and was negative for EMA, TFE3, HMB-45 and calretinin. The overall features were compatible with an epithelial component of a nephroblastoma.

A neurosurgery consult was of the opinion that the complete loss of cord function was likely irreversible, and surgical intervention was deferred. Further investigation by the social worker and community health

Abbreviations: WT, Wilms' tumour; CT, Computed tomography; MRI, Magnetic resonance imaging; PAX8, Paired box gene 8; WT-1, Wilms' tumour-1; EMA, Epithelial membrane antigen; HMB-45, Human melanin black-45; MDT, Multidisciplinary team; NAC, Neoadjuvant chemotherapy; SIOP, Société Internationale d'Oncologie Pédiatrique; COG, Children Oncology Group.

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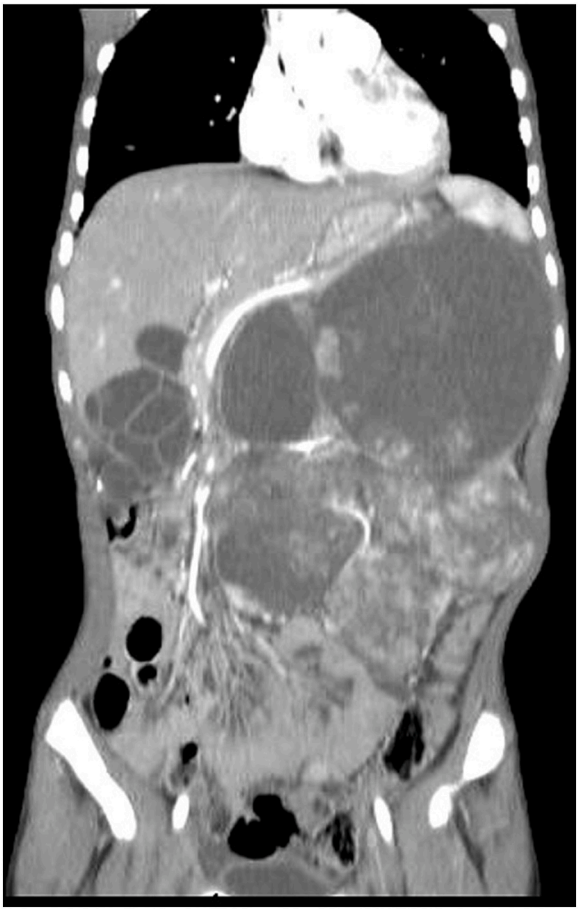

Fig. 1. Contrast-enhanced CT scan showing a large cystic and partially solid mass occupying the left upper quadrant and left flank region, extending into the pelvis caudally and crossing the midline.

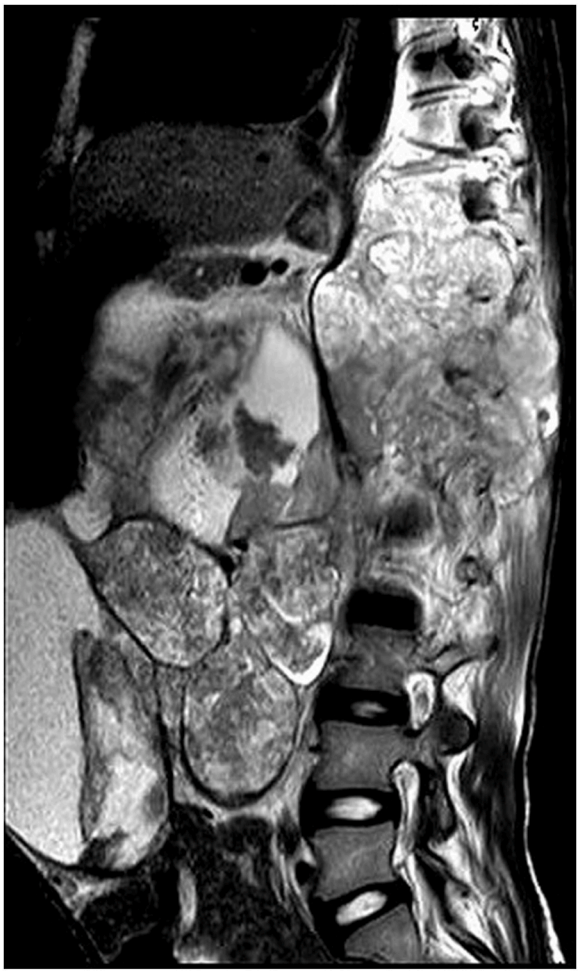

Fig. 2. MRI of the spine showing significant infiltration of the tumour into the spinal canal.

care representative revealed that the child comes from impoverished and difficult circumstances. The mother, the primary caregiver, was an unemployed, elderly alcoholic. A diagnosis of stage IV WT was made. It was decided at the multidisciplinary team (MDT) meeting to initiate prompt neoadjuvant chemotherapy (NAC) with vincristine, actinomycin-d and doxorubicin in the hope of making the tumour more amenable to surgical resection. Unfortunately, the patient demised soon after this MDT meeting before the initiation of NAC. The mother declined a postmortem examination to confirm the cause of death.

3. Discussion

Spinal cord involvement in childhood malignancies ranges from 2.7 to 4%. Neuroblastomas, soft tissue sarcomas, osteogenic and Ewing sarcomas are the most common malignancies responsible. In WT, spinal cord involvement is infrequent. The exact mechanism accounting for spinal cord involvement is unclear. Direct extension of the tumour through the vertebral foramina and subsequent spinal cord compression was the most likely explanation in our case. Other plausible mechanisms of spinal cord involvement in WT include haematogenous dissemination through the collateral circulation of the paravertebral venous plexus, lymphatic spread through the vertebral foramen, and extension along the perineurium of the spinal nerves or skeletal metastasis to the vertebral body.[1,2] Back pain, lower limb weakness, sensory loss, sphincter, and autonomic dysfunction are the most frequently reported symptoms associated with spinal cord compression.[3] These clinical features must be promptly recognized and treatment initiated as soon as possible to allow any chance of neurological recovery.

In its typical appearance, WT is triphasic and consists of variable proportions of blastemal, stromal, and epithelial cells. Our case only showed epithelial elements. The percutaneous biopsy sample may not have been entirely representative, or the tumour could have represented a monophasic (epithelial predominant) WT – a rare histological variant. Examination of the excised tumour would have given more representative sections to distinguish between the two and assist with prognostication using the Société Internationale d'Oncologie Pédiatrique (SIOP) and Children Oncology Group (COG) schemas.

Although several guidelines may be used to manage WT, our unit adopts the SIOP guidelines, and NAC was preferred to render the tumour more operable. Due to the complete loss of cord function, our neurosurgery colleagues did not deem her a candidate for surgical decompression. Unfortunately, the patient demised before the commencement of NAC.

Black children of sub-Saharan African descent consistently show the highest incidence of WT globally at 11 cases per million. Due to contemporary advances in medicine and a multimodal treatment approach, including surgery, multiple-drug chemotherapy and radiotherapy, as well as the availability of standardized treatment guidelines from large multidisciplinary cooperative cancer groups, namely the COG and SIOP, the 5-year survival for patients with WT in the developing world is now more than 90% and is hailed as one of the greatest success stories in modern oncology. This is in stark contrast to the overall survival at 5-years in sub-Saharan African nations, reported as low as 25%.[4] Unfortunately, our patient presented very late in the disease process due to multiple social and economic issues.

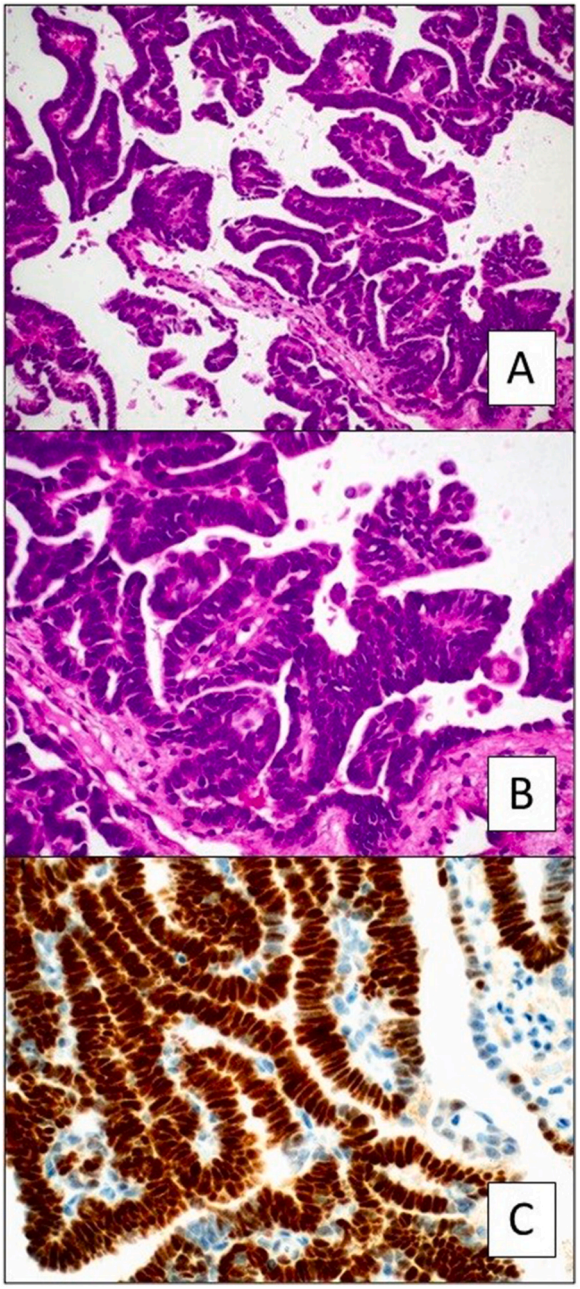

Fig. 3. (A) Papillary structures with fibrovascular cores are seen (Haematoxylin and Eosin, 200x), (B) High-power magnification demonstrates a simple epithelial lining comprising cells with high nuclear-to-cytoplasmic ratios and nuclear hyperchromasia (Haematoxylin and Eosin, 400x), (C) WT-1 immunohistochemistry is strongly and diffusely positive in the neoplastic cells (400x).

While one must acknowledge social, structural and cultural barriers responsible for this dismal overall survival in developing countries, a recent review by Apple and Lovvorn in 2020 suggested there may also be an underlying biological and molecular basis that may account for this discrepancy.[5] More research needs to be done to understand Wilms' tumorigenesis in our setting.

4. Conclusion

Malignant spinal cord compression is associated with a poor prognosis and may result in permanent paralysis, sensory loss and sphincter dysfunction. Therefore, early diagnosis and timeous initiation of treatment protocols, including high dose corticosteroids, chemotherapy and surgical resection, are critical in limiting functional morbidity and mortality.

Conflicting interests

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Ethical approval

No ethical approval is required by the institution for the publication of individual case reports.

Informed consent

Written informed consent was obtained from the patients mother for the anonymised information and the accompanying images to be published in this article.

Contributor ship

JJ and AA reviewed the literature and drafted the manuscript. All authors issued final approval for the version to be submitted for publication.

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