

Extradural Spinal Cord and Intraabdominal Ganglioneuroblastoma

G Kanakaraj*, I Zamzuri*, J Abdullah*, M Mohd Ghazali*, Z Mahamood**, A Shafiee***, M Madhavan****, M T Rahman****

*Department of Neurosciences, **Department of Surgery, ***Department of Radiology, ****Department of Pathology, School of Medical Sciences, Universiti Sains Malaysia, 16150 Kubang Kerian, Kelantan

Summary

We report a rare case of a giant extradural and intraabdominal ganglioneuroblastoma in a young Malay girl who presented to a paediatrician initially at 5 days of life with a palpable abdominal mass. Unfortunately, the parents refused any form of surgical intervention until the child was 3 years old. She subsequently underwent vascular embolisation followed by the removal of this large tumour both via the abdomen and through a laminectomy approach and subsequently refused chemotherapy. The c-myc amplification in this patient was absent, and there were no chromosomal aberrations. During the 2 year follow-up the patient remained well and ambulatory with no tumour recurrence.

Key Words: Giant ganglioneuroblastoma, Treatment

Introduction

The term ganglioneuroblastoma was introduced in 1915 to describe a transitional tumour of sympathetic cell origin containing both malignant neuroblastomatous and benign ganglioneuromatous elements. Ganglioneuroblastomas are a subgroup of neuroblastomas, a neural crest cell lineage tumour that is an intermediate tumour between neuroblastoma and gangliogliomas. We report a rare case of spinal cord ganglioneuroblastoma with intraabdominal extension in a 3 year old Malay girl and discuss the radiological and pathological features.

History and Clinical Assessment

A 3 year-old Malay girl first presented to a paediatrician in a neighbouring government hospital at 5 days of life, with persistent irritability and crying. She was noted to have a palpable lower abdominal mass with no neurological abnormalities. The initial investigations revealed raised levels of alpha fetoprotein and urinary

vanillyl mandelic acid(VMA) and the presence of a pelvic presacral mass on ultrasound of the abdomen. The parents refused any surgical intervention. The child was well until the age of 1 month, where she was diagnosed to have obstructive uropathy with abnormal electrolytes, palpable bladder and hydronephrosis. The CT abdomen showed an obvious mass in the pelvic region. She became hypertensive with her systolic blood pressure above the 95th centile, she was treated with nifedipine and intermittent bladder catheterization was taught to the parents. The parents were still not keen for surgery. Following that admission, she was hospitalized on numerous occasions with intermittent diarrhoea. At the age of 3 years, the parents finally agreed for operation after noting that the mass had increased in size and extended to the right gluteal region. Clinically, the mass was palpable per abdomen, and was a firm mass arising from the pelvis. It was non tender and it was not associated with jaundice, anaemia, lymphadenopathy and organomegaly.

This article was accepted: 10 July 2005

Corresponding Author: Jafri Malin Abdullah, Department of Neurosciences, School of Medical Sciences, Universiti Sains Malaysia, Health Campus, 16150 Kubang Kerian, Kelantan,

CASE REPORT

Another mass was also palpable at the right gluteal region. This was also firm, non tender and not associated with overlying skin changes. The neurological assessment including the spinal curvature was normal. There was no family history of phakomatoses or cancer. The child did not have skin manifestations of neuroectodermal disorders.

Magnetic Resonance Imaging (MRI) of the abdomen, pelvic and lumbosacral regions disclosed a well defined large homogenous lobulated mass with the main bulk in the pelvis, isointense to muscle on T1 and hyperintense on T2 and not suppressed on STIR with marked enhancement post contrast. It measured 10.9cm x 10.7cm x 10.1cm.

In the pelvis, the mass displaced and compressed the pelvic structures. The bladder was displaced anterosuperiorly, the rectum was compressed and displaced superiorly. The mass extended into the spinal canal through the anterior sacral foramina and filled up the spinal canal from the coccyx until the conus medullaris at the level of L3. It also extended posteriorly via the posterior sacral foramina to involve the erector spinae and right gluteal region muscles. The mass projected out from the pelvic region to the right gluteal region via the greater and lesser sciatic notches (Fig. 1). Both sciatic nerves were normal and clear demarcation was still seen. Prior to surgery, the feeding vessels of the mass were embolised to reduce intraoperative haemorrhage.

Under general anaesthesia, the child was laid supine, cleaned and draped for abdominal laparotomy. The transverse lower abdominal skin incision (Pfennestiel) with rectus abdominis cutting was made. The vertical extension was found to be necessary to have a good exposure. The abdomen was opened in layers and the peritoneum was entered. The findings were that the urinary bladder was pushed anterosuperiorly, the

uterus and ovaries lay just behind the urinary bladder and on the tumour. The tumour was well encapsulated and firm in consistency. It was mobilized and freed from adjacent organs. The tumour feeding vessels were ligated and as the tumour filled the whole pelvis, it was difficult to remove en-bloc as the entry point was via the left sciatic notch. The residual was diathermised and removed by piece meal. The wound was closed in layers. The child was positioned prone with the abdomen freed and the hips and chest well supported. The vertical skin incision was made from the L3 level and curving to the right incorporating the right gluteal mass till the level of S4. The dissection proceeded further till the tumoural plane was noted at the right gluteal region. A piece meal tumour removal was done and all macroscopic tumour was removed with preservation of the right sciatic nerve. The dissection proceeded medially until the intrapelvic parts were noted via the greater and lesser sciatic notches. The intrathecal part was removed by posterior sacrectomy of S1-5 and laminectomy of L5. The tumour was extradural in location but an area of dura defect was noted. Inspection intradurally revealed no tumour and thus the defect was plugged and sutured with fascia and enforcement with fibrin glue was done. The wound was closed in layers after insertion of a subarachnoid lumbar drain.

Histopathological examination revealed characteristics features of ganglioneuroblastoma. The tumour was composed predominantly of ganglioneuromatous stroma which was strongly reactive for S100 protein with occasional well defined nests (Fig. 2). These nests were composed of a mixture of neuroblastic cells and maturing ganglions in a neurofibrillary background. No mitosis or areas of necrosis was present. The parents refused chemotherapy and a 2 year follow up revealed that the patient was well and ambulatory with no recurrent growth.

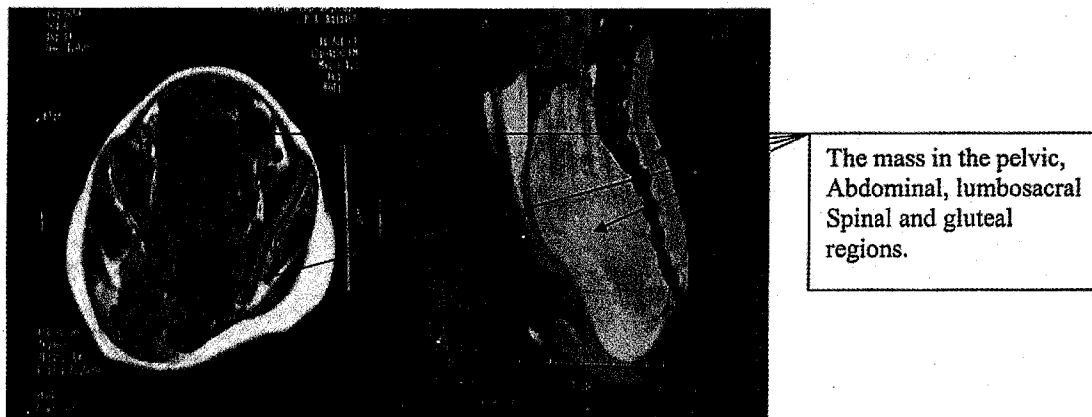


Fig. 1: The T1 and T2 weighted MRI images: The mass appeared isointense on T1 and hyperintense on T2. It occupied the pelvic, abdominal, lumbosacral spinal and gluteal regions.

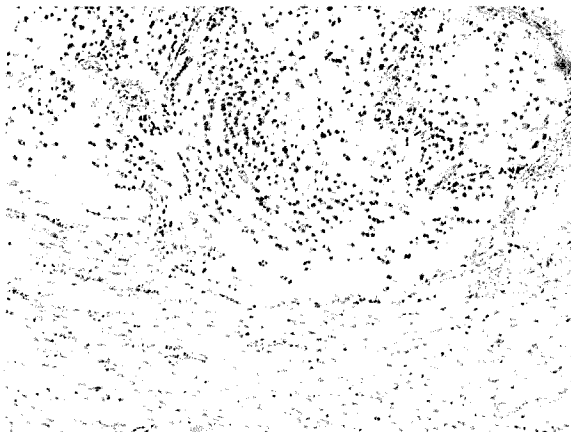


Fig. 2: Shows Schwannian stroma in the lower half of the field and a nodular collection in the upper field.

Discussion

Histologically, ganglioneuroblastoma is composed of nests of neuroblastic cells in the ganglioneuromatous stroma. It is less aggressive than neuroblastomas but it may metastasise to the liver, bone, orbit and intracranial compartment.

The initial age at diagnosis tends to be lower than for the other primitive tumours of childhood with approximately 83% of all cases occurring in the first 6 years¹. The major sites are the paravertebral sympathetic chains in the posterior mediastinum and retroperitoneum and the adrenal gland. Collectively, the adrenal gland and abdominal sympathetic ganglia are the sites of origin for 63-76% of tumours, especially

CASE REPORT

in the fetal, congenital and early infantile cases². In our case the sympathetic ganglia on the left side was most probably the site of origin. Symptoms and signs of tumour development are related to the specific anatomical region. Ureteral and bladder obstruction is due to the lower abdominal mass, diarrhoea possibly to oversecretion of vasointestinal peptide hormone (VIP) and hypertension because of elevation in serum catecholamines determined clinically by measuring the urine level for its metabolite, the vanillyl mandelic acid (VMA). No neurological deficits occurred, because the extradural component was not large enough to compress the cauda equina and did not extend high enough to compress the spinal cord as a dumb bell parapspinal lesion.

The ganglioneuroblastoma is a delicately encapsulated lobulated mass, the margins can be poorly defined and infiltration into adjacent tissue is common. The tissue is typically pink grey with areas of focal haemorrhage or necrosis. The most primitive component of the peripheral neuroblastic tumours are small and round to slightly elongated cells with scant, ill defined cytoplasm without apparent cytoarchitectural differentiation and a high nucleocytoplasmic ratio. Prognosis of this tumour is related to the stage of the disease. Localised tumours have a more favorable outcome than tumours with locoregional infiltration and disseminated disease. Other important prognostic factors are patterns of histopathology or grade and changes at the genomic level, such as amplification of N-myc oncogene, ploidy of the tumour and chromosomal aberrations. An absence of N-myc amplification, absence of chromosomal aberration and aneuploidy are associated with favorable prognosis. In this patient there was absence of c-myc amplification and chromosomal aberration.

This patient had INSS stage 2A. There were no palpable or detected lymph nodes noted on clinical examination, imaging and intraoperatively. The bone scan to detect metastasis revealed no abnormality. Treatment for patients categorized as low risk consists most commonly of surgery alone but in our case surgery without chemotherapy was given due to parents' refusal. Chemotherapy is reserved for patients who are symptomatic, such as from spinal cord compression or, with respiratory compromise secondary to hepatic infiltration. The chemotherapy consists of carboplatin, cyclophosphamide, doxorubicin, and etoposide. The cumulative dose of each agent is kept low in order to minimize permanent injury from the chemotherapy regimen.

Radiation therapy is reserved for patients with symptomatic life- or organ-threatening tumor that does not respond rapidly enough to chemotherapy, or for intermediate-risk patients whose tumor has responded incompletely to both chemotherapy and attempted resection and also has unfavorable biologic characteristics. Radiation therapy to the primary site is recommended for high-risk patients even in cases of complete resection³.

Conclusions

Large ganglioneuroblastoma are rarely reported in Asia and as in this case required complex management of both neurosurgery and oncology specialities. The patient's good Karnofsky status is most probably due to the absence of c-myc amplification which when present may indicate poor prognosis.

-
1. Kubota M, Suita S, Tajiri T et al. Analysis of the prognostic factors relating to better clinical outcome in ganglioneuroblastoma. *J Paed Surg* 2000; 35(1): 92-5.
 2. Joshi VV, Cantor AB, Altshuler G et al. Conventional Versus Modified morphologic criteria for Ganglioneuroblastoma. A review of cases from the Paediatric Oncology Group. *Arch of Path & Lab Med* 1996; 120(9): 859-65.
 3. Brodeur GM, Pritchard J, Berthold F et al. Revisions of the International criteria for neuroblastoma diagnosis, staging and response to treatment. *J Clin Oncol* 1993; 11(8): 1466-77.