A case of a rare adrenocortical carcinoma mimicking neuroblastoma

Kavitha Rozario Fiona Ryan Taffy Makaya
Oxford Children's Hospital, Oxford, UK

History

A 10 month old girl presented with a bluish lump on the left side of her abdomen. She had a hard ballotable abdominal mass and small hard mobile subcutaneous nodules over her abdomen, left scapular and left paraspinal area at the level of T4 to T5. There were no signs of virilization.

Initial investigations and management

Ultrasound	calcified mass (9cm by 6cm by 9 cm) arising from the left adrenal gland. Calcified lesions in her liver and lungs suggesting metastasis.
MRI	adrenal mass and intraspinal deposits with some cord compression.
MIBG scan	Negative
Urine	negative for vanillylmandelic acid (VMA)

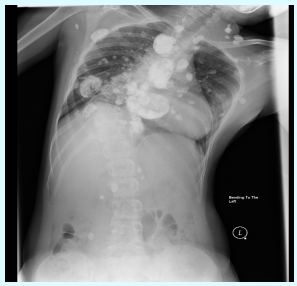
This was initially thought to be neurobloastoma and treated with 2 cycles of chemotherapy with Carboplatin and Etoposide.

Repeat MRI done 6 weeks post chemotherapy, showed a slight increase in size of the mass. She developed further skin nodules on her back which were biopsied, the results of which were inconclusive.

She had a further cycle of chemotherapy. (VIDE protocol)

Further investigations and management

Urine steroid profile - low steroid concentration. Serum androgens suggested that this adrenal mass was a non-steroid secreting tumour. Initial ACTH < 10ng/L, cortisol 636 nmol/L – these were taken when child was on steroids.
A Synacthen test (done 6 months into treatment) showed adrenal insufficiency with low cortisol (peak of 157 nmol/L and high baseline ACTH of 133 ng/l). Our patient was on Mitotane, an adrenolytic agent at this time.



X-ray showing multiple metastatic lesions.

Immunohistological analysis of a further skin biopsy was positive for vimentine, cytokertin and synaptophysin which was indicative of metastatic adrenocortical carcinoma. Genetics showed no amplification of MYCN gene, which is usually associated with neuroblastoma.

She was started on Hydrocortisone. She underwent bilateral adrenalectomy and T5 to T8 laminoplasty and decompression to relieve the cord compression caused by the intra spinal metastatic lesion. The prognosis at this stage was very guarded.

Presently:
• Our patient has residual metastases in the spine – which have remained stable over the years.
• She continues on high dose hydrocortisone (16mg/m^2/day) to keep her ACTH levels undetectable. She continues on maintenance Fludrocortisone.
• She is now nine years post treatment and is doing well. She is leading an active life, with no evidence of disease progression.

Discussion

Adrenocortical carcinomas (ACC) are very rare accounting for 0.2% of childhood malignancies. They affect girls more than boys (2:1). The peak incidence is 3.5 years of age.
Upto 50% of children with sporadic ACT have germline p53 mutations and may be associated with Li-Fraumeni and Beckwith-Wiedemann syndromes. Over 90% present with symptoms and signs of virilisation.

Staging in childhood ACC involves the size, weight, and amount of resection of the tumor.
Stage I is a tumor < 5 cm, weighing < 200 g, with complete resection;
stage II — > 5 cm, > 200 g, with complete resection;
stage III — local spread to lymph nodes, kidney, inferior vena cava or incomplete resection
stage IV — distant metastases to either lung or liver or to both.

Surgery remains the cornerstone of treatment in stages I – III of ACC. Despite surgery, recurrence rate can be as high as 80%. The role of chemotherapy in the management of childhood ACT has not been established although occasional tumors are responsive to mitotane or cisplatin-containing regimens.

References
1. Ng L, Libertino JM. Adrenocortical carcinoma: Diagnosis, evaluation and treatment. J Urol. 2003;169:5–11.
2. Michalkiewcz E, Sandrini B, Figueiredo B, Miranda EC, Caran E, Oliveira-Filho AG, et al. Clinical and outcome characteristics of children with adrenocortical tumors: A report from the international pediatric adrenocortical tumor registry. J Clin Oncol. 2004;22:838–45.
3. Cagle PT, Hough AJ, Pysher TJ, Johnson EH, Kirkland RT, Holcombe JH, et al. Comparison of adrenal cortical tumors in children and adults. Cancer. 1986;57:2235–7.
4. Teinturier C, Pauchard MS, Brugieres L, Landai P, Chaussain JL, Bougnere PF. Clinical and prognostic aspects of adrenocortical neoplasms in childhood. Med Pediatr Oncol. 1999;32:106–11.
5. Schteingart DE, Doherty GM, Gauger PG, Giordano TJ, Hammer GD, Korobkin M, et al. Management of patients with adrenal cancer: Recommendations of an international consensus conference. Endocr Relat Cancer. 2005;12:667–80.