

LETTERS TO THE EDITOR

Androgen secreting adrenocortical carcinomas

EDITOR,—As oncologists we read Wolthers *et al*'s retrospective experience of androgen secreting adrenal cortical tumours with interest.¹ This report is very important because it represents a substantial proportion of the UK's experience of this rare tumour of childhood. It also presents an opportunity to raise a number of important issues.

The main scientific content of this paper focuses on an attempt to correlate malignant tumour behaviour (local invasion and metastasis) with an analysis of histological characteristics. Like others, Wolthers *et al* were unable to identify any such correlation. There is, however, agreement that larger tumours (> 5 cm, 200 g, 200 cm³) are associated with a greater frequency of expression of malignant characteristics. To believe that small tumours are benign and big ones malignant is a misunderstanding of cancer, and to make progress we need to understand better intrinsic tumour biology. We therefore recommend that tumour samples from each of these rare cases are appropriately stored for future biological analysis (United Kingdom Children's Study Group wide protocol for collecting and banking tumour specimens for biological studies (98 BS 05); The UKCCSG, University of Leicester, 22–28 Princess Road West, Leicester LE1 6TP, UK).

We do not know the optimal treatment for these tumours following surgery.² As they are rare and have a substantial mortality, we propose a multicentre collaboration between endocrinologists and oncologists to address this issue. Such a collaboration would apply a multidisciplinary methodology to evaluate approaches to treatment through cancer registration, standardised clinical management, clinical trials of treatment and national/international audit of long term endocrinological and oncological outcome. This approach will prospectively recruit tumours nationally over a shorter time period and will have greater statistical power. It will involve specialists familiar with the needs of the child with cancer and with established links to those interested in researching factors that might predict tumour invasion and metastasis. We have initiated the first steps towards this goal through the officers of the UK Children's Cancer Study Group and the Royal College of Paediatrics and Child Health Endocrine Group.

Finally, it is now recognised that adrenal cortical tumours are frequently associated

with genetic causes of cancer predisposition, which may be the first manifestation of a familial cancer predisposition syndrome.³ Genetic investigation and counselling of the family are therefore an important part of patient management.

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- 2 Bonacci R, Gigliotti A, Baudin E, *et al*. Cytotoxic therapy with etoposide and cisplatin in advanced adrenocortical carcinoma. *Br J Cancer* 1998;**78**:546–9.
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Fatal chickenpox: negative electron microscopy of vesicular samples may be misleading

EDITOR,—A 12 year old boy was referred with a three day history of severe abdominal and chest pain radiating to the back. He had a vesiculo-haemorrhagic rash affecting his trunk and face for two days. He had been taking prednisolone (2 mg/kg/day) and diuretics since being diagnosed with rheumatic carditis four weeks previously. Two days before admission chickenpox was considered as a diagnosis in the referring hospital, but a vesicle scrape taken for electron microscopy was negative.

On admission ischaemic bowel was suspected, and a laparotomy and oesophagogastroduodenoscopy were performed. Apart from a haemorrhagic oesophagitis and a moderately engorged liver, no abnormalities were found. The next day his condition deteriorated with shock, hepatitis, and coagulopathy. A repeat vesicle scrape was again negative on electron microscopy, but immunofluorescence demonstrated varicella zoster virus (VZV); serum VZV IgG was not detected. Despite treatment with intravenous aciclovir and foscarnet along with full intensive care unit support, he developed multiorgan failure and died three days later.

Chickenpox specific, and possibly lifesaving,¹ antiviral treatment was delayed by two days in this case as a result of negative electron microscopy of a vesicle scrape. This

is not a sensitive technique, only detecting virus at a concentration of 10⁶ to 10⁷/ml.² In a review of paired samples sent to our laboratory over the past two years, nine of 33 samples positive for VZV by immunofluorescence gave false negative results by electron microscopy. Severe abdominal or back pain, frequently preceding the vesicular rash, appears to be a common feature of complicated varicella in immunocompromised patients,³ and aciclovir may be beneficial even at the time of visceral dissemination.^{1,4} Treatment of these children should thus start without delay, and the results of examination of vesicular material, particularly where electron microscopy alone is available, must be interpreted with caution.

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- 2 Doane FW, Anderson N. Methods for preparing specimens for electron microscopy. In: *Electron microscopy in diagnostic virology*. Cambridge: Cambridge University Press 1987:14–31.
- 3 Morgan ER, Smalley LA. Varicella in immunocompromised children. Incidence of abdominal pain and organ involvement. *Am J Dis Child* 1983;**137**:883–5.
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CORRECTION

Kearns Sayre syndrome initially presenting as hypomelanosis of Ito. *Arch Dis Child* 1999;**81**:280.

The authors for this letter should have been: Talia Kakourou, Anastasia Garoufi, Nikolaidou Polyxeni, Evmorfia Dafni, Migdaleni Tsamouri, A Papadimitriou, T Karpathios and not as published.

The error is regretted.