Primary adrenal B cell lymphoma

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CASE REPORT

A 57-year-old man presented with a 12-hour history of abdominal pain of sudden onset. The pain was localized to the right upper quadrant, and there were no relieving or aggravating factors. There was also no associated nausea or vomiting. Clinical examination revealed tenderness and voluntary guarding in the right upper quadrant, but no signs of rebound or peritonism. His only other symptom was night sweats which had been occurring for 2 years. There was no relevant past medical or family history.

An ultrasound scan of the abdomen demonstrated a 7.5x6.6x4 cm mass, superior to and separate from the right kidney and separate from the right lobe of liver. The mass contained multiple internal echoes and had appearances consistent with an adrenal mass. The other abdominal organs including the contralateral adrenal gland appeared within normal limits.

The ultrasound findings were confirmed on abdominal computed tomography (CT) scan and followed up by urine biochemistry, chest X-ray and routine blood tests, which were all normal. Protein electrophoresis showed a diffuse increase in γ-globulins (15 g/litre; range 4–7 g/litre). A CT-guided adrenal biopsy was performed which yielded a core of mostly necrotic tissue on which the differential diagnosis of malignant lymphoma or tuberculosis was raised. The patient proceeded to right adrenalectomy, which was uncomplicated.

A spherical tumour mass 85x80x75 mm (weight 230 g), with a smooth capsule and some normal adrenal gland attached to one pole, was resected. Opening the tumour revealed mostly necrotic yellowish material containing flecks of calcification. Histological examination showed some viable tumour at the periphery of the mass destructively infiltrating residual normal adrenal gland. The neoplastic cells were arranged in solid sheets and possessed large vesicular nuclei and 1–2 nucleoli. There was a high mitotic and apoptotic rate. The neoplastic cells were immunoreactive for leucocyte common antigen and the B cell marker L26, and the appearances were thus of a diffuse high grade B cell lymphoma (*Figures 1* and *2*).

Staging procedures including bone marrow trephine biopsy and CT scanning of the chest, abdomen and pelvis did not demonstrate any evidence of disease elsewhere. The patient received chemotherapy (cyclophosphamide, mitozantrone, vincristine and prednisolone) and has maintained a complete remission for 12 months.

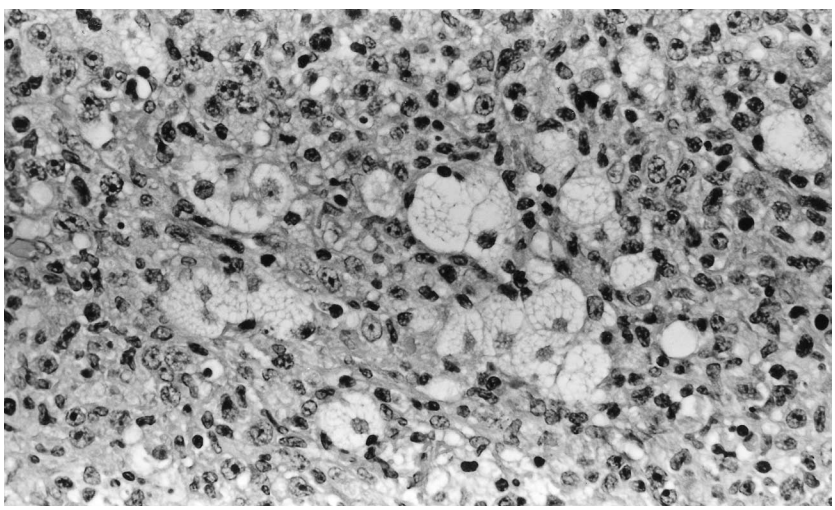

Figure 1. There is destructive infiltration of the adrenal cortex by a diffuse population of nucleolated lymphoid blasts. Some residual adrenocortical cells with abundant vacuolated cytoplasm are visible.

INTRODUCTION

Non-Hodgkin's lymphomas (NHL) more often involve extranodal sites than Hodgkin's lymphomas, and secondary involvement of the adrenal gland is relatively frequent at autopsy of patients with NHL (about 25% of cases) (Rosenberg et al, 1961). In contrast, primary localized adrenal NHL is very rare. Only 24 cases have been reported in the literature during the last 40 years (Sasagawa et al, 1995), in addition to cases where other organs were involved or who had lymphadenopathy at the time of diagnosis. We present a further case of primary B cell NHL of the adrenal gland.

DISCUSSION

This is a very unusual presentation of malignant lymphoma, and although there are rare case reports of primary adrenal malignant lymphoma, adrenal involvement is almost always the result of more widespread disease. The present case concerns a 57-year-old male with unilateral adrenal involvement. Of the 24 cases reviewed in the literature (Sasagawa et al, 1995), primary adrenal B cell NHL has a mean age of diagnosis and/or presentation of 65 years (range 39–81 years), with 75% of cases being male. In addition, of the 24 cases identified, 70% (17 cases) consisted of bilateral adrenal involvement, with the majority (76%) occurring in males. A predominance of B cell NHL was observed, 54% (13 of 24 cases), and 7 of these 13 cases were diffuse large cell NHL, as in the present case.

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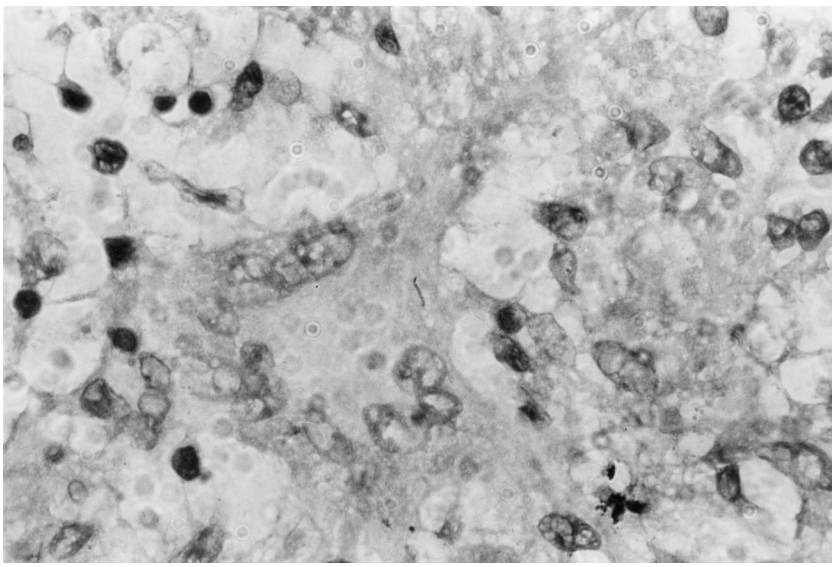

Figure 2. The lymphoma cells show positive membrane staining with the B cell marker L26.

The increased availability of computed tomography (CT), ultrasound and magnetic resonance imaging has made it possible to detect adrenal masses that were previously difficult to approach. Other causes of unilateral adrenal enlargement to consider, which are commoner than adrenal lymphoma, include adenoma, metastatic carcinoma, tuberculosis, primary adrenal carcinoma and phaeochromocytoma. Primary adrenal lymphoma has been described rarely as presenting with symptoms of adrenal insufficiency (Schnitzer et al, 1986; Carey et al, 1987). There was no evidence of adrenal insufficiency in the present case, and in general, about 90% of adrenal tissue must be destroyed before abnormal gland function can be detected (Guttman, 1930).

The diagnosis of primary adrenal NHL can be difficult to establish, and in only 10 of the cases reported previously to ours was the histological diagnosis made preoperatively with CT-guided needle biopsy (Sasagawa et al, 1995). The prognosis of primary adrenal NHLs reported in the past appears to be poor (Bauduer et al, 1992), with 8 months being the longest time from diagnosis to death. Our case remains in remission a year after his adrenalectomy. The increasing use of CT- or ultrasound-guided needle biopsy should improve the prognosis owing to prompt chemotherapy following an early histological diagnosis. **HM**

Bauduer F, Delmer A, Le Tourneau A et al (1992) Primary adrenal lymphoma. *Acta Haematol* **88:** 213–5

Carey RW, Harris N, Kliman B (1987) Addison's disease secondary to lymphomatous infiltration of the adrenal glands. Recovery of adrenocortical function after chemotherapy. *Cancer* **59:** 1087–90

Guttman PH (1930) Addison's disease: a statistical analysis of 566 cases and a study of the pathology. *Arch Pathol* **10:** 742–85

Rosenberg SA, Diamond HD, Jaslowitz B, Craver LF (1961) Lymphosarcoma: A review of 1269 cases. *Medicine* **40:** 31–84

Sasagawa I, Sadamori N, Itoyama T et al (1995) Primary adrenal lymphoma with chromosomal abnormalities. *Acta Haematol* **94:** 156–62

Schnitzer B, Smid B, Lloyd RV (1986) Primary T-cell lymphoma of the adrenal glands with adrenal insufficiency. *Hum Pathol* **17:** 634–6

In the Public's View...

Not with a bang, but with a *whimper+

Have you seen the film 'Independence Day'? It's completely unrealistic. It's not the spaceship 500 km long that's the problem, nor the daughter vessels 10 km in diameter obliterating whole cities with gouts of blue flame. I can even cope with Will Smith flying an alien fighter through the enemy defences having never flown one before, and Jeff Goldblum blocking the mother ship's force shield with a computer virus. All this I can accept, but the film is quite ridiculous: the American President's wife dies of uncontrollable internal bleeding with just one forearm drip and without having an operation of any sort. And she is able to sit up, smile, and embrace her daughter as she fades away.

We need have no fear of murderous aliens, our end will be far more mundane. The human race will die out when, frozen into immobility, its members are able to remember only PINs and access codes. With my latest banker's card came a strict little diktat telling me that I was not under any circumstances to write my PIN anywhere, even in a disguised form. This same instruction applies to my other banker's cards (3), my E-mail accounts (3), various web-sites that need access codes (4), and half the doors I pass through during a routine working day.

The human brain is extremely poor at remembering unconnected strings of characters. There is some hope if users are allowed their own passwords. Amazingly, the commonest password is 'password', and people tend to choose family names but there are all sorts of ruses to help users to remember but non-users not to find out. One suggestion is the initial letters of words in the first line of poems. It's probably a good idea to avoid nursery rhymes: any code-breaker would crack MHALL and JAJWUTH pretty quickly.

My favoured method is to take a word, double one of the letters, and then add a number and non-alphanumeric. Even the largest computer in the world would take some time to get to %cuupboard8, although the commonest way computer security is broken is when users are observed entering passwords or clever hacking programs save entered passwords in invisible files. It's for that reason that passwords should be changed every now and again, but that requires the forgetting of the old password and learning of the new. I give myself about 5 years before I reach PINword-saturation.

Access codes are even worse. They cannot be user-defined, and are forever being changed. It is possible to remember them only if they are written down. I would be interested to know how much the NHS spends on security, and whether anyone thinks it is worth it. Hospitals are such sprawling places, with such varied people roaming about, that anyone can go anywhere if they have the confidence to do so. I'd be especially interested to know how much it has cost to turn obstetric units into places that are now better at preventing clinical staff getting access in an emergency than at preventing a visitor walking off with a baby. Politically unacceptable to do otherwise, but how much does it cost to prevent the abduction of one baby? **HM**

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