increase in pressure probably was of recent origin but despite normal cardiac dimensions left ventricular function was grossly impaired with extreme hypokinaesia of the left ventricular wall. The E/A ratio was below one in both of them, suggesting reduced left ventricular compliance.

A remarkable finding in our cases was the isolated increase in plasma noradrenaline in the absence of a phaeochromocytoma. Although we cannot entirely exclude the possibility that this was secondary to the pulmonary oedema (especially not in patient 2), we think such a mechanism to be less likely because one would have expected a comparable rise in adrenaline and activation of renin. The question arises, therefore, whether acute oversecretion of noradrenaline was the primary event that has induced contraction of the coronary arteries with subsequent ischaemia and hypokinaesia of the heart (stunning). In other words, left ventricular dysfunction during acute episodes of hypertension may be due to a direct effect of catecholamines on the heart rather than to pressure overload as is also observed in patients with phaeochromocytoma.5,6

In conclusion, the present findings demonstrate that sometimes severe myocardial hypokinaesia (stunning) may accompany severe hypertension in the absence of overt coronary disease. Perhaps excess secretion of noradrenaline is involved in the pathophysiology of this phenomenon.

References


A feminizing adrenocortical carcinoma presenting with gynaecomastia

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Summary: A case is presented in which gynaecomastia was the sole initial presenting symptom of a feminizing adrenocortical carcinoma. This rare pathological lesion is discussed.

Introduction

Adrenocortical neoplasms are rare. They may be classified into benign and malignant, and functioning and non-functioning types. All may produce local signs and symptoms, such as loin pain, a mass, or even haematuria if the kidney is invaded.1 Malignant tumours of the adrenal cortex are considered to have a very poor prognosis. The functioning types may manifest themselves through their hormonal effects; indeed, these may be the only presenting features in some cases. A case is presented in which gynaecomastia was the
sole presenting complaint. Gynaecomastia is a common disorder, with a prevalence of up to 36% in 'normal' males.2

Patients complaining of benign breast enlargement are commonly referred to plastic surgeons; this case is discussed in this context.

Case report

A previously fit and healthy 34 year old male was referred to the Department of Plastic and Reconstructive Surgery with a 6 month history of bilateral gynaecomastia. He found the condition distressing. There were no other abnormal features in his history and physical examination was otherwise normal. A clinical endocrinological assessment and serum testosterone (normal) had been carried out. He was placed on the waiting list for excision of the redundant breast tissue. Six months later (while still on the waiting list) he was referred to the Department of Urology with a history of right loin pain which had been present for 3 months. A preliminary intravenous urogram had been carried out and was reported as normal. He underwent ultrasound of the right renal area, and this investigation disclosed the adrenal mass.

The patient was submitted for investigation (see Table I and at operation (through a right 9th rib thoraco-abdominal incision), a 750 g adrenal tumour was found. It was adherent to the undersurface of the liver, and tumour thrombosis extended through the adrenal vein into the inferior vena cava. The lesion was excised, along with a cuff of inferior vena cava and the patient made an uneventful recovery. Histology confirmed an adrenocortical carcinoma, necrotic in parts, with marked pleomorphism and numerous mitotic figures. Postoperatively his biochemistry returned to normal (although he required a short course of bromocriptine to reduce his serum prolactin to normal). Despite this, his gynaecomastia did not regress and he underwent excision 6 months after his adrenalectomy.

### Table I Preoperative endocrine values

<table>
<thead>
<tr>
<th>Serum</th>
<th>Preoperative values</th>
</tr>
</thead>
<tbody>
<tr>
<td>Oestradiol</td>
<td>410 pmol/l (40–85)</td>
</tr>
<tr>
<td>Prolactin</td>
<td>590 mU/l (&lt;400)</td>
</tr>
<tr>
<td>Testosterone</td>
<td>3.5 nmol/l (9–38)</td>
</tr>
<tr>
<td>FSH</td>
<td>0.5 U/l (0.5–50)</td>
</tr>
<tr>
<td>LH</td>
<td>2.9 U/l (3–12)</td>
</tr>
<tr>
<td>Urinary</td>
<td></td>
</tr>
<tr>
<td>Pregnantriol</td>
<td>27 μmol/24 hours (2–7)</td>
</tr>
<tr>
<td>Androsterone</td>
<td>2.2 μmol/24 hours (3.6–19.6)</td>
</tr>
<tr>
<td>17-Oxogenic steroids</td>
<td>66 μmol/24 hours (15–41.5)</td>
</tr>
<tr>
<td>Free cortisol</td>
<td>1,421 nmol/24 hours (50–250)</td>
</tr>
</tbody>
</table>

Normal ranges in brackets.

In addition to regular clinical examination he has had measurements of serum oestradiol and testosterone done as tumour markers. All these parameters remain normal 30 months postadrenalectomy.

Discussion

Breast development depends upon the ratio of oestradiol to testosterone. The ratio in normal males is 1:100 or greater at all times, compared to 1:10–1:100 in the premenopausal female. The ratio in this patient was 1:8.5. Herr et al.3 point out that there may be four possible causes of abnormal breast development in males: excess oestrogen production, androgen deficiency, hepatic failure and drugs. Despite his high oestrogen levels, this patient's presentation was unusual in that he did not display any of the other features associated with this abnormality, such as testicular atrophy, loss of libido or mastalgia. This factor undoubtedly contributed to the delay in diagnosis of his tumour.

Endocrinological screening (Table I) shows that the tumour was secreting an excess of oestradiol and C-21 steroids. Given the high levels of measured free cortisol in the urine it is remarkable that no Cushingoid features were noted. This may be due to cross-reactivity of glucocorticoid precursors (which such a tumour may produce) with the free cortisol detected by the assay.4 Perhaps the relatively weak glucocorticoid effects of these compounds were not sufficient to have produced a clinical effect at the time of presentation. Depression of serum testosterone by such endocrinologically active tumours is due mainly to gonadotrophic inhibition by high levels of circulating oestriadiol, as well as the direct effect of these levels on the Leydig cells.5 It is interesting to note that, although depressed when measured preoperatively, the patient's testosterone level was normal 6 months previously, when he first sought a consultation for his gynaecomastia.

Although Gabrilove et al.6 reported on 52 cases of feminizing adrenocortical carcinoma (based on a review of published literature), it is, in fact, a rare variant. In the series of Meagher et al.,7 11 of 32 adrenal tumours were malignant, and only six of these tumours were functioning. None was feminizing, and only two of the 11 occurred in males. Similarly, Bertagna and Orth8 report a series in which 22/32 carcinomas of the adrenal cortex were functional and only two of the carcinomas occurred in men. Such tumours are seen occasionally in children, but again, most are masculinizing.9

Most authors have commented on the gloomy prognosis of these lesions. MacFarlane10 quoted an average survival in the untreated case of only 2.9 months from the time of diagnosis. Henley et al.11
suggest a 5 year survival rate of 16% in cases treated surgically. Perhaps because of its rarity, prognostic factors are difficult to discern; it appears not to depend on either clinical stage or histologic grade. Adrenocortical cancer shares with renal cell carcinoma the unusual propensity to propagate tumour thrombus along major veins, as demonstrated in the current case. One might imagine that this feature should adversely affect prognosis, and our patient's 30 month recurrence-free interval is striking in this regard.

This case serves to point out that gynaecomastia may occasionally be due to hormonally active neoplasia. Other causes include trophoblastic tumours secreting human chorionic gonadotrophin (teratomas and some seminomas) and rare entities such as Leydig cell tumours11 and pelvic sarcomas.3 Perhaps all young men with gynaecomastia ought to have a steroid screen.

References


Hypercalcaemia and multiple osteolytic lesions in childhood acute lymphoblastic leukaemia

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Summary: A 12 year old boy presenting with hypercalcaemia (calcium 3.25 mmol/l) and osteopaenia with multiple osteolytic lesions was found to have acute lymphoblastic leukaemia without lymphadenopathy or organomegaly. Hypercalcaemia is a rare feature of acute leukaemia, but the patients previously described all show very similar characteristics, which were highlighted in this patient. These include age (10–20 years), severe osteolytic bone lesions, lymphoblastic leukaemia, and normal white cell count with absent or rare circulating blasts. Parathyroid hormone levels were normal in this patient, and response to induction therapy was good. This case demonstrates that acute lymphoblastic leukaemia may present in an atypical form without peripheral blasts but with hypercalcaemia and gross skeletal changes.

Introduction

Hypercalcaemia and severe osteolytic lesions are rare complications of acute lymphoblastic leukaemia (ALL), and only a few cases with these presenting features have been recorded.1–3 This low incidence contrasts with the high incidence in some other lymphoid malignancies, such as myelomatosis and adult T cell leukaemia/lymphoma (ATLL).

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