LETTERS TO THE EDITOR

LEFT VENTRICULAR MYXOMA, ADRENAL TUMOUR AND CUTANEOUS VASCULITIS - A CASE REPORT

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Dear Sir,

The case report of the presentation of left ventricular myxoma in association with an adrenal tumour and cutaneous vasculitis[1] is an interesting and educational one. While the title clearly reflects the case reported on, please allow me to clarify on three particular points made by the authors in their discussion.

(1) **The diagnosis of a benign tumour based on CT appearance**

The authors have indicated that on ultrasonography the adrenal tumour measured 4.4 cm in diameter. It would be desirable for the reader to know the dimensions of the tumour on the CT scan. The authors have suggested that based on radiology the appearance of the tumour was most likely benign, quoting the experience of Adams et al[2]. Adams et al[2] studied 98 patients with suspected adrenal disease in whom they had confirmatory diagnosis in 73. Of these 73 patients, 10 patients had a carcinoma. Although Adams et al had shown that mixed attenuation suggested malignant lesion and uniform attenuation suggested benign lesion (statistically significantly) - even amongst their own series they found 3 of the 11 lesions which were of mixed attenuation were non malignant and one out of 24 patients with uniform attenuation had a malignant lesion. More recent papers have not concurred with the concept of reliability of CT findings in the distinction between benign and malignant lesions. Fishman et al[3] in a subsequent paper on review of the CT evaluation with clinical correlation of 38 patients with primary adrenocortical carcinomas concluded that contrary to established concepts, adrenocortical carcinoma may present as a smooth, homogenous, functioning mass 6 cm or less in diameter on CT. Paivansalo et al[4] have further suggested that there exists no single criterion to help resolve the differential diagnosis between benign and malignant lesion using CT. In a more recent paper Dunnick et al[5] have suggested that adrenal carcinomas causing Cushing's syndrome generally exceed 6 cm in diameter and are heterogeneous with areas of necrosis and calcification, although about 16% of carcinomas are <6cm in diameter and at CT may resemble adenomas. Kolmannskog et al[6] in a study of 15 patients with adrenocortical carcinomas have demonstrated that the adrenal tumours in two of their patients had diameters of 4 and 6 cm respectively.

The photograph provided in Fig 1 by the authors shows the tumour to be non-homogenous in appearance. That appearance would be less in favour of a benign mass - homogeneity is generally associated with benign masses, but benign masses can haemorrhage and be heterogeneous; conversely, adrenal cortical carcinomas can be homogenous.

Thus, it would appear that there is insufficient evidence to conclude that this tumour was benign based on the CT appearance.

(2) **The diagnosis of functioning nature of the adrenal tumour**

The authors have suggested that "the functioning nature of the adrenal tumour was demonstrated by the failure to suppress plasma cortisol and the 24 hour urine cortisol levels. Looking at their data carefully however, one notes that while hypercortisolism has been documented on a single occasion by the elevated 8 am cortisol level of 1655 µmol/L and an elevated 24 hour urine free cortisol of 0.91 µmol measured after the patient have been on dexamethasone 2 mg thrice daily for 2 days, they have not clearly documented the non-suppressibility. Firstly, the dexamethasone dose was incomplete - the traditionally accepted high dose dexamethasone suppression tests proposed by Liddle[7] utilises 2 mg dexamethasone 6 hourly for 2 days. Secondly, it is difficult to interpret non-suppressibility of the urine free cortisol based on the single 24 hour urinary levels. If one were to utilise the criterion for non-suppressibility for urine free cortisol, one needs to compare how much this is suppressed compared to basal levels. The authors have indicated in their script that the ACTH levels were "normal". That would imply that, if indeed, this patient had Cushing's syndrome then it must be ACTH-dependent. If one were to attribute the hypercortisolism to the adrenal tumour, then it must be theoretically ACTH-independent ie. the ACTH levels should be very low normal or more often undetectable. Alternatively, if one finds a solitary adrenal tumour causing ACTH-dependent Cushing's syndrome, the remotely possible ACTH-secreting phaeochromocytoma has to be on the differential diagnosis[8].

Thus the authors should have provided further clarification (under discussion) on their statement that "the functioning nature of the adrenal tumour was demonstrated. . .".

(3) **The Carney's Complex**

The authors have suggested that in this patient although there were no pigmented skin lesions the clinical features would fit easily as part of the Carney's Complex. Carney himself who first described this complex had recently[9] suggested the basis of diagnosis of this syndrome. He suggests that the condition should be suspected if one of the components (myxomas, skin pigmentation, endocrine tumours and schwannomas) is present in typical fashion. Definitive diagnosis requires the presence of either two of the components in a typical fashion or one of the components in typical fashion in a patient having an affected primary relative.

One rather unusual feature of the case reported on, is the presence of the unilateral large adrenal adenoma. Of Carney's 156 patients reported up to late 1993, 33% had primary pigmented nodular adrenocortical disease (PPNAD) of the adrenal cortex. The appearance of PPNAD is very characteristic[10] on radiology with small nodules unilaterally or bilaterally easily demonstrable on modern CT scanners - the gland itself appears slightly lumpy but is not enlarged. We are not told what is the size or appearance of the contralateral uninvolved (?) adrenal gland. The described appearance of the CT findings of the unilateral adenoma of this case report does not fit into the typical appearance seen in Carney's Complex and therefore one cannot accept this (based on the data provided in the case report) as a case of Carney's Complex.

I hope the authors could provide further clarification on their interesting case.

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AUTHORS' REPLY

Dear Sir,

We read with interest the letter by Dr Rajasoorya regarding our recently published case report and would like to offer the following reply.

The size of the adrenal tumour on CT Scan was similar to that measured by the ultrasound, about 5 cm in diameter. The right adrenal gland was not well visualised on CT Scan but did not appear to be enlarged. Dr Rajasoorya had rightly commented that the heterogenous appearance of the tumour could not be reliably used to distinguish benign from malignant lesions. In the study by Fishman et al[1], only 6 out of 38 patients with primary adrenocortical carcinoma had a tumour 5 cm or less in diameter. Similarly, in each of the series by Paivansalo et al[2] and Kolmannskog et al[3], only one patient with adrenocortical carcinoma had a tumour <5 cm in diameter; the mean diameters in the series were 8 cm and 11 cm respectively. Indeed in a large study of 307 cases, Guerrero[4] found carcinomas smaller than 5 cm in only 1.6% of cases. Benign tumours are generally round structures which are smooth in contour like in this case, whilst adrenal carcinomas are commonly irregular in shape with lobulated margins[5]. Thus, as we have stated in our discussion, the tumour in our patient was likely to have been benign. Definitive diagnosis could only be made with histology which was unfortunately not available.

We agree that it would have been desirable to use the higher dose of 2 mg dexamethasone 6 hourly for 2 days in the suppression test. A pre-suppression plasma cortisol level was not available but the post-suppression level was nevertheless markedly raised. The ACTH level was below normal (17.6 ng/L, normal range 10-40 ng/L) as would be the case in some functioning adrenal tumours.

Like Carney, we were struck by the occurrence of 2 very rare conditions - adrenal tumour and cardiac myxoma - in the same patient, who in this case also had cutaneous vasculitis. We were unable to examine her family to determine if there was an affected relative. Whilst the patient did not fulfil all the criteria for a definitive diagnosis of Carney Syndrome, the presence of a similar constellation of conditions could form part of the complex, perhaps a "form fruste" type.

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