Phaeochromocytoma of the urinary bladder localized by selective venous sampling and computed tomography

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Summary
Clinical and biochemical evidence of a catecholamine-secreting tumour was documented in a 19-year-old female presenting with malignant hypertension. The tumour was successfully localized to the pelvis by means of plasma noradrenaline assay from multiple sites, after previous attempts at localization had failed. Subsequently, computed tomographic (CT) scan allowed correct pre-operative localization to the urinary bladder.

KEY WORDS: phaeochromocytoma, bladder, venous sampling, CT scan.

Introduction
The majority of phaeochromocytomas arise from the adrenal medulla and, including those in ectopic locations, more than 95% are intra-abdominal. Extra-adrenal phaeochromocytomas are difficult to localize pre-operatively (Engelman, 1977). A patient with an extra-adrenal phaeochromocytoma is reported, where successful localization to the bladder was achieved by computed tomographic (CT) scan subsequent to the results of plasma noradrenaline assay in samples taken from multiple sites (Moss, Greenbaum and Sever, 1980; Huw Jones et al., 1979). Previous attempts at localization had failed.

Case report
A 19-year-old female presented with bitemporal headaches, and was found to have a blood pressure of 290/180 mmHg and bilateral papilloedema on fundoscopy. Because of restlessness and agitation, postural hypotension could not be assessed. After normotension had been achieved with oral prazosin, she admitted to paroxysms of headache, palpitations and diaphoresis occurring at infrequent intervals, with no obvious precipitating factors. Urinalysis was entirely normal. Of significance was the fact that 4 years previously, she presented with 'pre-eclamptic toxemia'. After control of her hypertension, she delivered a healthy infant vaginally but was lost to follow-up.

Blood pressure was controlled on α-blockade, with postural hypotension being a troublesome feature. β-blockade was introduced because of persistent sinus tachycardia. A clinical diagnosis of a phaeochromocytoma was confirmed biochemically, with urinary vanillyl-mandelic acid, total metanephrines and total catecholamines being persistently and markedly elevated.

Attempts at pre-operative localization of the tumour by means of abdominal computed tomography (CT) and selective arteriography failed to reveal any tumour. The cystographic phase of the arteriogram was unsuccessful in documenting any tumour. Selective venographic sampling with assay of plasma noradrenaline (Cat-A-Kit, Upjohn Diagnostics, Michigan, U.S.A.) from various sites was performed. Plasma noradrenaline concentrations in peripheral plasma was grossly elevated at 20 ng/ml (upper limit of normal 0·8 ng/ml). Highest plasma noradrenaline concentration was found in blood from the left common iliac vein, suggesting origin from left side of the pelvis (Fig. 1). Repeat CT scan of the pelvis revealed a 6·3×4·3 cm mass arising from the right dome of the bladder (Fig. 2) which was subsequently confirmed on ultrasonography.

Under general anaesthesia, cystoscopy was performed, but no mass lesion or ulceration of the bladder mucosa was observed. However, a prominence of sub-mucosal vessels was observed overlying...
Vesical phaeochromocytomas usually present with symptoms of catecholamine-excess related to micturition (Flanigan et al., 1980). Hypertension was noted in 60% of cases with bladder phaeochromocytoma, haematuria in 59% and micturitional attacks in 47% (Ochi et al., 1981). The case reported lacked any symptoms of catecholamine excess related to micturition, haematuria was not documented and of note is that, 4 years before this current problem, 'pregnancy-related hypertension' did not in any way precipitate a hypertensive crisis during vaginal delivery of a healthy infant.

Anatomically, bladder phaeochromocytomas are most commonly located in the trigone (25%) or dome (24%) and are visible at cystoscopy in 84% of reported cases (Ochi et al., 1981). The case reported lacked the usual cystoscopic findings of bladder mass or ulceration of the bladder mucosa.

Localization of the phaeochromocytoma was initially attempted by computed tomography, as this technique is non-invasive and has been reported as being successful in 91% of cases studied (Stewart et al., 1978). The adrenals were normal in size and shape and no intra-abdominal masses were present. At the time, limited cuts of the pelvis at 30 mm intervals were obtained without vesical contrast, and were reported as normal. Selective venographic sampling for plasma noradrenaline localized the tumour to the left pelvis. Repeat CT scan of the pelvis was then successful in localizing the mass to the right dome of the bladder. This emphasizes the importance of meticulous attention to detail when performing these procedures, in order not to miss mass lesions in relatively unusual localities.

The reason for the localization of the tumour to the left side of the pelvis by venographic sampling, despite right-sided location, was evident at surgery. The vascular supply was from a prominent leash of vessels coursing across the dome of the bladder towards the left internal iliac system.

Although the literature has mention of bladder phaeochromocytomas being localized by selective venographic sampling for noradrenaline, in none of these reports was this the initial investigation, the tumour initially being suspected to be in the bladder because the patients volunteered symptoms related to micturition. The tumours were initially localized to the bladder by either intravenous urography or angiography, and subsequently selective venous sampling was done as confirmatory evidence (Khan et al., 1982; Timmis, Brown and Allison, 1981).

To the best of our knowledge, this is the first documented case of a bladder phaeochromocytoma being initially localized by selective venographic sampling, and subsequently by CT scan and ultrasound (Modlin et al., 1979; Ochi et al., 1981; Flanigan et al., 1980; Das and Lowe, 1980; Scott and Eversole,

Discussion

Phaeochromocytomas of the bladder are rare tumours, accounting for less than 0.06% of all bladder neoplasms (Laestma and Price, 1971) and, to date, only about 80 cases have been published (Meyer, Sane and Drake, 1979; Ochi et al., 1981).
1960) before surgical intervention, and emphasizes the value of selective venographic sampling for localization of phaeochromocytomas when non-invasive techniques fail.

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References


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