CASE REPORT

Association of angiomyolipoma and oncocytooma of the kidney: a case report and review of the literature

K Pillay, J Lazarus, H C Wainwright

Aim: The association between renal carcinoma and angiomyolipoma is rare. Only 14 cases have been reported in the literature. The purpose of this paper is to present an additional case and review the literature on this association.

Patient and methods: A healthy 42 year old woman was found to have a left flank mass incidentally when she presented for a Papanicolaou smear. The computerised tomography scan revealed a left lower pole renal mass consistent with a renal cell carcinoma. A nephrectomy was performed and the patient recovered uneventfully. The nephrectomy specimen was processed routinely. In addition to haematoxylin and eosin staining, immunohistochemistry for CAM 5.2, vimentin, CD34, antismooth muscle actin, and HMB45 was carried out. Transmission electron microscopy was also performed.

Results: Macroscopically, the lower pole of the kidney contained a well circumscribed, non-encapsulated, tan coloured tumour with a large area of central haemorrhage measuring 10.5 cm. In addition, there was a 0.4 cm poorly circumscribed unencapsulated yellow nodule adjacent to the tumour. Microscopically, the larger tumour showed characteristic features of an oncocytooma. Numerous mitochondria were seen on electron microscopy. The smaller yellow nodule was an angiomyolipoma.

Conclusions: This paper presents an additional case of oncocytooma associated with angiomyolipoma. Of the 15 cases described in the literature, three were associated with the tuberous sclerosis complex, all from a single study. In tuberous sclerosis, angiomyolipomas are more commonly associated with renal cell carcinoma. If angiomyolipomas are found incidentally in nephrectomy specimens together with other tumours, it is important to exclude tuberous sclerosis retrospectively.

The association between renal oncocytooma and angiomyolipoma is rare. To the best of our knowledge, only 15 cases have been reported in the literature (including our present case).1-7 In the case described by Schneck et al the oncocytooma was present in a partial left nephrectomy specimen. The right nephrectomy specimen showed a transitional cell carcinoma involving the pelvis and calyceal system, with an incidental cortical angiomyolipoma.1 Jimenez et al reported five cases, and one of these patients had multiple oncocytoomas (oncocytosis).7 Oncocytoomas have also been associated with cortical adenomas and renal cell carcinoma, whereas angiomyolipomas have been associated with renal cell carcinomas, a papillary adenoma, and a metanephric adenoma.2,7,8

FIGURE 1 Computed tomography scan of the abdomen showing a large left renal neoplasm.
of the kidney, there was a well circumscribed, non-encapsulated, tan coloured tumour, with a large central area of central haemorrhage, measuring 10.5 cm in diameter. There was no evidence of necrosis, perirenal fat involvement, or involvement of the renal vein. In addition, there was a 0.4 cm poorly circumscribed unencapsulated yellow nodule adjacent to the tumour (fig 2).

Microscopic findings
Sections of the kidney showed a well circumscribed tumour composed of cells with abundant granular eosinophilic cytoplasm growing in a tubular manner (fig 3). The nuclei were small, round, and regular with a centrally placed nucleolus. Focal nuclear pleomorphism was noted, although there were no mitotic figures present. There was no evidence of a papillary architecture, areas of clear cell carcinoma, or sarcomatoid or spindle cell areas. There were areas of stromal hyalinisation, myxoid change, and haemorrhage, but there was no evidence of necrosis. The tumour cells were positive with CAM 5.2 and negative for vimentin.

The adjacent renal parenchyma showed a non-encapsulated, irregular tumour displaying mature adipose tissue, smooth muscle, and blood vessels (fig 4). The last two components were highlighted by the antismooth muscle actin and CD34 immunostains, respectively. The smooth muscle component showed some nuclear pleomorphism and hyperchromasia. The features were those of a renal angiomyolipoma. The HMB45 stain was negative in this case.

Electron microscopy
The cytoplasm of the cells in the larger tumour was filled with mitochondria (fig 5). The diagnosis of an oncocytoma was made.

LITERATURE REVIEW
Table 1 lists the available details of the other published cases.

DISCUSSION
Renal oncocytomas, first described by Zippel in 1942, are benign, relatively uncommon neoplasms accounting for 3–5% of renal parenchymal tumours. Most of these tumours are
Renal angiomyolipomas are uncommon neoplasms accounting for less than 1% of surgically removed tumours. However, they are the most common mesenchymal tumour of the kidney and are characterised by the presence of a variable mixture of mature adipose tissue, blood vessels, and smooth muscle. The perivascular epithelioid cell is thought to be the cell of origin of angiomyolipomas. Although renal angiomyolipomas, like other tumours derived from the perivascular epithelioid cell, are characteristically positive for HMB-45, a cell of origin is the intercalated cell of the cortical portion of the collecting tubule.

Surgical excision/nephrectomy remains the treatment of choice because there is no definite preoperative method of identifying these tumours.

Renal angiomyolipomas are uncommon neoplasms accounting for less than 1% of surgically removed tumours. However, they are the most common mesenchymal tumour of the kidney and are characterised by the presence of a variable mixture of mature adipose tissue, blood vessels, and smooth muscle. The perivascular epithelioid cell is thought to be the cell of origin of angiomyolipomas. Although renal angiomyolipomas, like other tumours derived from the perivascular epithelioid cell, are characteristically positive for HMB-45, a small proportion of these tumours do not stain with this antibody.

Renal angiomyolipomas are uncommonly associated with tuberous sclerosis, which is an autosomal dominant disorder characterised by seizures, mental retardation, skin lesions, and hamartomatous lesions in many organs. Rarely, angiomyolipomas can be associated with von Hippel-Lindau disease, von Recklinghausen syndrome, and autosomal dominant polycystic kidney disease.

However, most angiomyolipomas occur sporadically. In tuberous sclerosis, angiomyolipomas are found predominantly in women in the third and fourth decades of life and they tend to be asymptomatic, small, multifocal, and bilateral. In sporadic cases, they are seen in 40 to 70 year old women and are usually larger, symptomatic, single, and unilateral.

In a series of 36 cases of concurrent angiomyolipoma and renal cell neoplasia reported by Jimenez et al, the median size of angiomyolipomas was 0.5 cm in the sporadic cases and 3 cm in those patients with tuberous sclerosis. In both clinical settings, angiomyolipomas were more commonly the incident finding. The mean ages of the patients with sporadic and tuberous sclerosis tumours were 59 and 53 years, respectively.

Although, the presence of fat in angiomyolipomas may aid in the radiological diagnosis, these tumours may still mimic a renal cell carcinoma, particularly if the fat content is low or if the fat is obscured by blood.

Take home messages

- This is only the 15th case of oncocytoma associated with angiomyolipoma to be reported.
- Although our case did not occur in the setting of the tuberous sclerosis complex, three of the previously reported cases have done so.
- In tuberous sclerosis, angiomyolipomas are more commonly associated with renal cell carcinoma.
- If angiomyolipomas are found incidentally in nephrectomy specimens removed because of the presence of other tumours it is important to exclude tuberous sclerosis retrospectively.

Table 1 Age, sex, and association with tuberous sclerosis of the 15 patients with angiomyolipoma in association with oncocytoma

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NK, not known.

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REFERENCES