Angiomyxoid tumour in the renal peripelvic tissues with features of aggressive angiomyxoma

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Abstract
A case of angiomyxoid tumour in the renal peripelvic soft tissue of a 45 year old woman is reported. The encapsulated tumour, measuring 12 x 9 x 6 cm, was solid and firm. The cut surface revealed a yellowish-white, gelatinous, and glistening tumour. Histologically, the tumour was composed of loosely textured spindle and stellate cells with prominent blood vessels of variable calibre. The tumour cells stained strongly for vimentin. These findings strongly resemble those associated with aggressive angiomyxoma in the vulva and perineum.

Keywords: Renal peripelvis, mesenchymal tumour, angiomyxoma, aggressive angiomyxoma.

Mesenchymal tumours of the renal peripelvic tissue are uncommon. Here, we report a myxoid tumour with a prominent vascular component, histologically and immuno-histochemically indistinguishable from aggressive angiomyxoma in the female vulva and perineum.1-3

Case report
A 45 year old Japanese woman presented with a painless mass in the right upper abdomen. A computed tomography scan revealed a tumour in the right renal pelvis, measuring 12 cm in diameter, with a central low density area. A clinical diagnosis of renal cell carcinoma was made because of the unclear border between the tumour and the kidney, and the patient received a right nephrectomy. The renal pelvis and ureter were not affected by the tumour and none of the regional lymph nodes were enlarged. The tumour had not recurred 21 months following surgical excision.

Pathology
On gross examination, the tumour was located in the peripelvic soft tissue of the right renal pelvis. When depressed, it was easily detached from the right kidney and renal pelvis. The encapsulated tumour, measuring 12 x 9 x 6 cm, was solid and uniformly firm without lobulation. The cut surface revealed a yellowish-white, gelatinous, and glistening tumour with foci of petechial haemorrhage. No necrotic or cystic areas were present.

Histologically, the tumour was composed of loosely textured mesenchymal spindle and stellate cells with a prominent vascular component. The tumour cells had ovoid nuclei with finely dispersed chromatin and small indistinct nucleoli (fig 1). Foci of increased cellularity were present. A myxoid change was prominent in the stroma, with dispersed delicate wavy collagen fibres. The matrix was strongly positive on staining with colloidal iron and very weakly positive with alcian blue at pH 2.5. Prominent blood vessels, ranging from medium sized arteries and veins to capillaries, were distributed haphazardly throughout the tumour tissue. The arteries

![Figure 1 Several spindle tumour cells with ill-defined cytoplasm are scattered in a myxoid matrix (haematoxylin and eosin).](image-url)
frequently displayed medial muscular hypertrophy; some vessels showed hyaline thickening of the walls and some were associated with extravasation. These vessels had elastic lamellae, revealed when stained with elastica van Gieson stain (fig 2). Small amounts of mature adipose tissue were encased in the tumoral tissue, but no neoplastic components of smooth muscle or adipose tissue were identifiable.

An immunohistochemical study was performed on formalin fixed, paraffin wax embedded sections using the labelled streptavidin biotin technique. Primary antibodies against vimentin, actin, α-smooth muscle actin, desmin, factor VIII related antigen, S-100 protein, neuron specific enolase, and α-1-antichymotrypsin (Dakopatts, Copenhagen, Denmark) were used.

The mesenchymal spindle and stellate cells stained strongly for vimentin, weakly for actin and α-smooth muscle actin, and negatively for desmin. Smooth muscle cells of the vessels stained positively for actin, α-smooth muscle actin, and desmin. No immunoreactivity was observed on staining for S-100 protein, neuron specific enolase, or α-1-antichymotrypsin. The endothelial cells of vessels displayed immunoreactivity for factor VIII related antigen.

Discussion
The tumour in the present case was composed of a mixture of myxoid and vascular elements, highly reminiscent of aggressive angiomyxoma and angiomyofibroblastoma. The mesenchymal spindle and stellate cells were strongly positive on staining for vimentin and, to a lesser extent, for actin and α-smooth muscle actin.

Aggressive angiomyxoma is characterised by the presence of prominent myxoid and vascular components and most frequently occurs in the female vulva, pelvic floor, and perineum. It shows local infiltrative growth and frequently recurs. The tumour cells are regarded as myofibroblastic or fibroblastic in nature because of their ultrastructural and immunohistochemical features. Aggressive angiomyxomas also have been reported in the uterine cervix and in the male scrotum, inguinal region, spermatic cord, and pelvis.

Angiomyofibroblastoma is a benign mesenchymal tumour of the vulva, mimicking aggressive angiomyxoma. It is well circumscribed and its cells show immunoreactivity for both vimentin and desmin.

Not withstanding the clear border of the lesion, the histological and immunohistochemical findings in this case more closely resembled those of aggressive angiomyxoma than those of angiomyofibroblastoma. The tumour was not defined as "aggressive" because it seemed to have no potential for local recurrence.

Angiomyxomas have also been reported in the umbilical cord and skin, but these tumours differed from the present case and from aggressive angiomyxoma.

The differential diagnosis includes angiomylipoma and myxoid tumours such as myxoma, nerve sheath myxoma, myxoid lipoma, myxoid liposarcoma, and myxoid malignant fibrous histioctyoma. Angiomyolipoma, consisting of a mixture of thick-walled blood vessels, smooth muscle and adipose tissue, frequently occurs in the kidney. Although a few cases of extrarenal perinephric angiomyolipoma have been reported in the literature, the case reported here did not have an adipose element and its vessels had elastic lamellae. Its microscopic distinctions from other myxoid tumours have been summarised by Steeper and Rosai.

References