A case report of an angiomyxoma in the liver

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ABSTRACT

Angiomyxomas are uncommon myxoid tumours arising most commonly from the pelvis. A 46-year-old woman with a history of polycystic kidney disease presented asymptotically for surveillance ultrasonography; changes were noted in the size and morphology of her liver cysts. Subsequent radiological assessment displayed features suspicious of malignancy and a right hemihepatectomy was performed with curative intent. Pathological examination of the resected specimen found histology consistent with an angiomyxoma arising primarily from the liver parenchyma. Follow-up review of the patient has been uneventful with annual imaging showing no evidence of recurrence. Angiomyxomas do not characteristically invade other tissues. However, any liver lesion displaying suspicious features of malignancy should be resected in the absence of disseminated disease.

KEYWORDS

Liver – Angiomyxoma – Liver cyst – Polycystic kidney disease – Liver tumour

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Myxomas are benign tumours thought to arise from multipotent mesenchymal cells. They are the most common primary cardiac tumour (usually a solitary tumour in the left atrium)¹ and have seldom been described elsewhere in the body. Cardiac myxomas are typically resected owing to the risk of thromboembolism and sudden cardiac death.¹ The histogenesis and embryological development of myxomas is poorly understood. However, it has been suggested that intracardiac mesenchymal cells from the epicardium and endocardium are precursor cells.² Angiomyxomas (further divided into superficial and aggressive) are a rare subtype of myxoid tumours, found almost exclusively in the soft tissues of the pelvis and perineum.³

A report of nine cases of aggressive angiomyxoma in the pelvis and perineum described homogeneity with regard to the histological appearances displayed in each case.⁵ The primary tumours contained small spindle cells within a myxoid background; uniform nuclei with dense chromatin were observed in the tumours. To our knowledge, only one other case of angiomyxoma in the liver has been described in the literature.⁴

Case history

The case described is that of an asymptomatic 46-year-old woman, treated previously with radical radiotherapy at 15 years of age for an ovarian dysgerminoma, with a history of polycystic kidney disease and a few isolated liver cysts. The cysts in the kidney were under regular surveillance with transabdominal ultrasonography performed every two years. In March 2012 the ultrasonography showed changes

in the size and morphology of a right-sided liver cyst, with features suspicious of malignant transformation. Initial differential diagnoses included seeding into a hepatic cyst from the original ovarian tumour, a simple liver cyst complicated by haemorrhage and a biliary cystadenocarcinoma. A full set of blood tests including tumour markers showed no abnormality.

Computed tomography (CT) of the thorax, abdomen and pelvis, and subsequent magnetic resonance imaging (MRI) of the liver were performed. The CT liver showed a 12cm x 9cm complex hypoattenuating mass in the right lobe of the liver (Fig 1) with no further sites of disease in the body. On magnetic resonance imaging (MRI), the mass was of high T2 and low T1 signal intensity (Fig 2) but had a complex appearance with progressive enhancement of an extensive network of internal septations (Figs 3–5). Marked capsular enhancement was observed during the equilibrium phase of contrast imaging. Although the delayed enhancement of the mass suggested a fibrous component, a potential malignant lesion could not be excluded.

Subsequently, a right hemihepatectomy was performed with no intraoperative complications. Postoperative recovery was uneventful and the patient was discharged home on day 7.

Histopathology results were consistent with the right lobe weighing 938g. There was a normal gallbladder and a large, unilocular, mucin filled cyst with well circumscribed walls measuring 130mm x 60mm. Microscopic examination revealed a loose myxoid stroma with both well and poorly formed blood vessels (Fig 6). There was no atypia or malignancy observed and the background liver showed features

Figure 1 Axial computed tomography during the portal venous phase showing a large hypoattenuating mass in the right lobe of the liver, which appears predominantly cystic but contains internal septations. Note the simple cysts in the left lobe of the liver and left kidney

of mild chronic inflammation. The cyst was fibrotic and hyalinised. Overall, the features were that of an angiomyxoid lesion. The specimen displayed a cystic portion lined with a single layer of low cuboidal epithelium in keeping with a simple cyst. The differential diagnoses of the lesion (based on microscopic appearance prior to immunohistochemistry results) were a gastrointestinal stromal tumour, a smooth muscle tumour and a myxofibrosarcoma.

The histopathology report suggested the lesion was consistent with a cystic myxoma but owing to the rarity of such lesions (with no other case reported in the literature at that time), the slides were sent for a second opinion to another histopathology department at a

Figure 2 Axial T2 weighted magnetic resonance imaging: The liver mass is of high T2 signal intensity, reflecting the cystic nature of this lesion

Figure 3 Axial fat saturated T1 weighted magnetic resonance imaging: The right lobe of the liver mass is of low T1 signal intensity

hepatopancreatobiliary tertiary centre. This report described a mildly cellular lesion with bland stellate cells in a fibromyxoid stroma, with inflammatory infiltrate. Immunostaining was positive in some lesional cells for smooth muscle actin. These findings indicated the lesion was not a gastrointestinal stromal tumour or myxofibrosarcoma (Table 1) and were instead consistent with an angiomyxoma.

At the four-month postoperative follow-up clinic visit, the patient was well and symptom free. She then entered a follow-up programme consisting of annual contrast CT of her thorax, abdomen and pelvis alongside a hepatopancreatobiliary specialist nurse-led clinic appointment. At 60 months after surgery, she remains well and free from disease.

Figure 4 Axial fat saturated T1 weighted magnetic resonance imaging 30 seconds after administration of intravenous gadolinium contrast: The liver mass demonstrates internal septal enhancement

Figure 5 Axial fat saturated T1 weighted magnetic resonance imaging 5 minutes after administration of gadolinium contrast: The liver mass demonstrates progressive internal septal and capsular enhancement during the later phase of imaging

Figure 6 Microscopic examination showing a mildly cellular lesion with bland stellate or short spindle cells in a fibromyxoid stroma with an inflammatory infiltrate. Immunostaining is positive in some lesional cells for smooth muscle actin, and negative for CD31, CD34, CD117, DOG1, desmin, S100 protein, AE1/AE3, α -fetoprotein, MUC4, h-caldesmon, oestrogen receptor and progesterone receptor

Discussion

Angiomyxomas have rarely been discovered in the liver, with only one other such report published.⁴ Although our patient had a previous history of treated malignancy, the histological findings were not consistent with a metastatic lesion and so it seems likely that this lesion arose primarily from the liver.

Despite not having been diagnosed preoperatively, the mass had a sufficiently alarming radiological appearance to warrant surgical resection. The progressive internal septal and capsular enhancement found on MRI was consistent with the presence of an internal fibrous stroma, as found subsequently on histology. Although this can be observed with mesenchymal tumours (eg malignant fibrous histiocytomas), there is considerable overlap with other malignant tumours, making exact diagnosis difficult radiologically.⁵

Angiomyxomas have not been described as characteristically invading other tissues but the features observed on MRI that were suggestive of malignancy were the primary factor in deciding to resect the lesion. These tumours have

not been reported to undergo malignant transformation. Nevertheless, depending on their location in the liver, they could result in compression of local structures leading to symptoms of compression of the inferior vena cava. Preoperative diagnosis of an angiomyxoma in the liver is very difficult given the rarity of such lesions in this location. However, any lesion observed in the liver suspicious of malignancy should be removed in the absence of frank-disseminated disease, with postoperative pathological examination providing a firm diagnosis.

References

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Table 1 Distinguishing pathological features of an angiomyxoma compared with the differential diagnoses of the lesion based on radiological findings

	CD34	CD117	DOG1	Desmin	Smooth muscle actin	h-caldesmon
Angiomyxoma (current case)	–	–	–	+	+/-	–
Gastrointestinal stromal tumour	+	+	+	+/-	–	–
Leiomyoma	–	–	–	+	+	+
Myxofibrosarcoma	–	–	–	+	+/-	–

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