CASE REPORT

Splenic myoid angioendothelioma mimicking metastatic disease in an 8-year-old with Stage IV Wilms' tumour

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SUMMARY

Myoid angioendothelioma are rare and benign vascular tumours of the spleen. Radiographic evaluation and diagnosis is often challenging and subjecting tissue samples to immunohistochemical analysis is often required to make a definitive diagnosis. Myoid angioendotheliomas can be managed with open or laparoscopic splenectomy with minimal risk of recurrent disease. Herein, we present a case of a myoid angioendothelioma in a patient with stage IV Wilms' tumour.

BACKGROUND

First described by Kraus and Dehner in 1999 as part of a series of four cases, myoid angioendothelioma (MA) are rare and benign non-haematopoietic vascular neoplasms of the spleen. These histologically constitute a mixture of vascular endothelium and stromal components with myofibroblastic differentiation.¹ The mesenchymal embryologic origin of the spleen allows it to harbour such mesenchymally derived tumours. Vascular tumours of the spleen, although infrequent, constitute the majority of non-haematopoietic proliferations within the organ. They are subclassified into benign, borderline or malignant, based on cytological and morphological features.¹ Common vascular neoplasms of the spleen comprise haemangiomas, hamartomas, littoral cell angioma, angiosarcoma, lymphangioma and haemangioendothelioma.² The relative infrequency of these lesions can often lead to difficulty in differentiation between similar vascular lesions. Herein, we present a case of a patient with stage IV Wilms' tumour and MA mimicking metastatic disease.

CASE PRESENTATION

An 8-year-old boy was diagnosed with stage IV Wilms' tumour of the right kidney with metastatic disease to the lung 4 years prior to presenting with a splenic mass. He had undergone a right nephrectomy and received chemoradiation with dactinomycin, vincristine and doxorubicin as part of therapy aimed at treating his Wilms' tumour. Three years after his initial diagnosis, tumour surveillance revealed a new intra-abdominal disease with lymph node involvement. He subsequently underwent an exploratory laparotomy with lymphadenectomy and small bowel resection. Postoperatively, he was treated with a

chemotherapeutic regimen consisting of ifosfamide, carboplatin and etoposide.

A routine surveillance positron emission tomography/CT demonstrated increased fluorodeoxyglucose uptake in the spleen which was concerning for recurrent Wilms' tumour. (figure 1) An abdominal ultrasound confirmed a heterogeneous and hypoechoic splenic mass measuring 3.1×2.5×2.1 cm. He, therefore, was scheduled for a diagnostic laparoscopy. A laparoscopic splenectomy was performed without complications.

The laparoscopic splenectomy was performed in the usual fashion with four trocars. The lesion was identified in the upper pole of the spleen using laparoscopic ultrasonography and there were no other lesions identified. Notably, the lower pole of the spleen was adherent to the transverse upper abdominal incisions from a prior nephrectomy. The gastrosplenic, lienocolic, lienorenal and lienophrenic ligaments were divided with a Ligasure (Medtronic, Minneapolis, Minnesota, USA) energy device. The hilum of the spleen was divided with an endo-stapling device. The spleen was placed in an Endobag (Medtronic). In order to prevent tumour spill, the spleen was removed intact (without morcellation) by opening the left aspect of the transverse abdominal incision removing the spleen within the bag in addition to the attached peritoneum and sent for pathological evaluation.

Pathological examination of the spleen revealed a well-circumscribed nodule measuring 2.7 cm (figure 2). Microscopic examination (figure 3) showed a well-encapsulated neoplasm consisting of proliferations of epithelioid and spindle cells arranged in a vague fascicular pattern. The epithelioid cells were large, polygonal, with abundant eosinophilic cytoplasm. Numerous, small, attenuated, slit-like vascular spaces were present along with an accompanying chronic lymphoplasmacytic inflammatory infiltrate. There was no significant cellular pleomorphism or increased mitotic activity. The myoid nature of the epithelioid cells was demonstrated by immunostaining with smooth muscle actin (SMA). CD34 highlighted the sieve-like vascular pattern. Immunohistochemically, the tumour was positive for CD34, CD31 and factor VIII and negative for CD8 and CD21. These findings were consistent with a MA.

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Figure 1 Contrast CT of the abdomen and pelvis demonstrating a hypodense splenic lesion measuring 2.6×2×2 cm (arrow).

OUTCOME AND FOLLOW-UP

The patient had an uneventful postoperative course and was discharged home on the second postoperative day. He subsequently completed his chemotherapeutic regimen of ifosfamide, carboplatin, and etoposide. He did not have recurrence of his MA at 1-year follow-up.

DISCUSSION

MA are extremely rare tumours of the spleen with an indolent clinical course. Morphologically, MAs arise from the red pulp of the spleen and are characterised by a composite of vascular spaces and stromal cells with myoid features.³ MA was first described in 1999 by Kraus *et al*, as a haemangioendothelioma-like tumour with myoid features.^{1,3} To the best of our knowledge, only five reports exist in medical literature describing this rare neoplasm in seven different patients. Consistent with the current case, a majority of these were discovered incidentally. Patient ages ranged from 3 to 51 years. Five of these diagnoses were associated with a concurrent neoplasm (table 1). Although MA has been described in a patient with Wilms previously,³ ours

Figure 2 Gross splenic specimen with a grayish-pink capsule and a 2.5 cm indurated nodule (arrow). Cross-sections of the nodule revealed a well-circumscribed mass measuring 2.7×2.1×2.0 cm. The suture attached indicates adhesion to the peritoneal wall.

Figure 3 (A) Tumour (right) well demarcated from adjacent splenic parenchyma (left) (20×). (B) Cellular areas separated by incomplete bands of fibrosis (100×). (C) Plump epithelioid tumour cells with visible nucleoli and eosinophilic cytoplasm along with small vascular channels (400×). (D) Chronic lymphoplasmacytic inflammatory infiltrate (400×). (E) Strong and diffuse immunoreactivity for smooth muscle actin in the epithelioid cells (400×). (F) Sieve-like vascular pattern highlighted by immunostaining with CD34 (400×).

remains the first report of such a lesion developing at such an advanced stage of the disease

Due to the relatively high concurrence with other neoplasm, it is often easy to confuse MAs for metastatic disease.⁴ Differential diagnoses of this lesion include primary vascular tumours of the spleen like angiosarcoma, hamartoma, haemangioma, haemangioendothelioma and haemangiopericytoma.⁵ Metastatic disease, however, is a more frequent occurrence within the spleen and should remain high on the list of differentials while evaluation. Radiologically vascular splenic neoplasms demonstrate differences in contrast enhancement patterns on CT or MRI. A delayed enhancement pattern on dynamic-contrast MR although not characteristic has been reported for these tumours. However, differentiation utilising imaging is often difficult due to frequently overlapping radiological features.^{2,4}

Kraus *et al* reported no clinical recurrence among the three reported patients between 12 and 30 months. No evidence of recurrent disease was found for the subsequent reports at their respective follow-ups.² This is consistent with the patient reported herein who had no evidence of recurrent disease on follow-up in a year's time.

Histological and immunophenotypic features of MA are used to differentiate these neoplasms from classic hamartomas, haemangiomas and previously characterised haemangioendotheliomas of the spleen. They likely represent proliferations of myoid elements native to the spleen.⁵ Vascular immunophenotypic features of MA are CD34+, CD31+, FVIII+ and CD8-. Stromal cells are believed to be differentiated from native myoid cells and stain SMA+, desmin+, S100± and CD8-.^{1,3,5}

Table 1 A brief comparison of cases of splenic myoid angioendothelioma reported in medical literature

Authors	Year	Age (years)	Sex	Diagnosis made	Associated diagnosis	Tumour size (cm)	Follow-up
Kraus <i>et al</i>	1999						
Case 1		3	F	Incidentally	Beckwith-Wiedemann Syndrome	2.5	No recurrence at 16 months
Case 2		7	M	Abdominal pain	None	4.0	No recurrence at 30 months
Case 3		43	F	Incidentally	Cystic mucinous pancreatic neoplasm	1.9	No recurrence at 12 months
Karim <i>et al</i>	2004	51	M	Incidentally	Abnormal liver function	3.5	No recurrence at 12 months
Chan <i>et al</i>	2005	4	M	Incidentally	Wilm's tumour	2.0	No recurrence at 3 months
Jang <i>et al</i>	2013	41	F	Incidentally	Rectal cancer	2.2	No recurrence at 17 months
Geramizadeh <i>et al</i>	2017	36	F	Abdominal pain	None	3.0	No recurrence at 3 months
Present case	2017	8	M	Incidentally	Wilm's tumour	2.7	No recurrence at 12 months

Although MA is a morphologically distinct lesion, its histological spectrum, biological behaviour and relationship to other vascular tumours have not been completely characterised to date.

A laparoscopic approach was utilised to perform the splenectomy in this case. This approach has not been previously described for MAs in the few reported cases. Minimally invasive splenectomy is associated with a shorter length of stay, superior cosmesis and fewer complications.⁶ It is also, however, is associated with increased operating time and blood loss when compared with an open approach.^{6–10} In one study, laparoscopic splenectomy had an increased hospital cost and no difference in blood loss when compared with the open approach in children.⁹ Therefore, laparoscopic splenectomy is a viable procedure of choice for management of benign and malignant haematological disorders, splenic cyst and splenomegaly in adults and children.^{8–10} Further cases of MA are required to determine a biological, pathological and histological association between MA and Wilms' tumour.

Learning points

- Myoid angioendotheliomas are benign tumours of the spleen that are of mesenchymal origin.
- These have a relatively high concurrence rate with other neoplasms.
- Imaging is typically not reliable to make a diagnosis and tissue samples, subject to immunohistochemical analysis, are often required.
- Laparoscopic approaches to splenectomy appear to be effective in management.

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