Pararenal malignant melanotic nerve sheath tumour: a rare tumour in an unfamiliar location

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DESCRIPTION

We present the case of a man in his 70s without any significant medical history who was being investigated for acute epididymo-orchitis and was incidentally found to have a large pararenal mass. Of note, our patient did not have any history of cutaneous melanoma. MRI revealed a retroperitoneal complex cystic lesion anterior to the right kidney (figure 1D) which contained dependent complex material reflecting blood products (arrow). On precontrast T1 sequences, nodular high signal along the anterior aspect of this lesion (figure 1A, arrow) demonstrated enhancement of postgadolinium injection (figure 1B, arrow), indicating the presence of melanin. Enhancement of the capsule of this lesion was also be appreciated (figure 1B,C). This large, complex cystic lesion was closely related but separate, appearing to the adjacent renal parenchyma with mass effect and distortion of the adjacent anterior renal parenchyma.

Following an uncomplicated radical right nephrectomy and histopathological analysis, this lesion was identified as a pararenal malignant melanotic nerve sheath tumour (MMNST). Figure 2 demonstrates the gross specimen with the large, melanin pigmented brown pararenal tumour (star) and the adjacent normal right renal parenchyma and perirenal fat. The cavity and cystic compartments of the tumour contained fresh haemorrhage as was seen as dependent complex material on MRI (figure 1D). On the histology slide beneath magnified at ×40, focal melanin (arrowhead) can be appreciated within a spindle cell area correlating with the regions of high T1 signal and enhancement seen on MRI.

MMNST was previously categorised as melanotic schwannoma by the WHO prior to its most recent classification update on soft tissue and bone tumours. This reclassification reflects a better appreciation of this rare tumour’s malignant potential which is higher than previously thought. Previously, melanotic schwannomas were thought to have been generally benign entities with only occasional cases of a more aggressive course. A recent, large clinicopathological, immunohistochemical and gene expression profiling study of 40 cases found these tumours to have a local recurrence and distant metastases rates of 35% and 42%, respectively. In this series, 8 of the 11 patients with metastases developed within 48 months of diagnosis. These tumours are often associated with Carney complex (an autosomal dominant tumour predisposition syndrome with skin pigmentary abnormalities, myxomas and endocrine tumours),

Figure 1  MR images of a large retroperitoneal mass. (A) Axial precontrast T1-weighted MRI image demonstrating high T1 signal (arrowhead) which demonstrates enhancement post administration of gadolinium (B, arrowhead) representing melanin. This cystic mass also demonstrated dependent complex material internally representing blood products (D, arrowhead) and enhancement of the thickened and irregular wall (C) on the coronal postcontrast T1-weighted imaging.

Figure 2  Gross specimen at time of resection demonstrating the pigmented cystic lesion (star) separate from the adjacent renal parenchyma and histology slide at ×40 magnification identifying focal melanin (arrowhead) within a spindle area.
although clinical course cannot be predicted by morphological features.

Given the limited number of published cases of this tumour, data on treatment and follow-up are limited. Therefore, close patient follow-up in a multidisciplinary team setting with regular radiological imaging is important in identifying local and metastatic recurrence. Finally, the most common location for MMNST is a paravertebral location; however, these tumours can arise anywhere within the retroperitoneum as in our case, and maintaining a broad differential which includes neurogenic tumours is important for atypical lesions of the retroperitoneum.

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Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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REFERENCES

Learning points
- Melanotic schwannomas have been recently reclassified as malignant melanotic nerve sheath tumours to reflect better understanding of the significant malignant potential of this rare tumour, which was previously thought to be benign.
- Maintaining a broad differential in the diagnosis of lesions of the retroperitoneum, including nerve-based soft tissue tumours, is important when imaging features are not definitive.
- These rare tumours require close multidisciplinary follow-up with regular radiological imaging an important component.