Giant dumbbell-shaped schwannoma but not transforaminal: transdiaphragmatic

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Accepted 3 May 2014

SUMMARY

A 31-year-old man reported a 3-month history of right upper abdominal pain and effort dyspnoea. The laboratory data demonstrated no abnormality. On abdominal ultrasound, a gross solid lesion with smooth borders was seen behind the liver. CT scan revealed a huge tumour, 23×17×15 cm in size, it was in the right posterior mediastinum and extended to the right retroperitoneum. In sagittal plane CT images, the lesion was like a dumbbell shape. CT-guided biopsy revealed a spindle cell tumour.

BACKGROUND

First, schwannoma should be kept in mind for the differential diagnosis of a giant thoracoabdominal solid lesion. Second, the dumbbell-shaped tumour is a non-specific term for any of a number of usually benign tumours. The nerve sheath tumour may assume the shape of a dumbbell as a classic when the tumour extends outward the spinal canal through the vertebral foramen. This finding may contribute to the diagnosis. But the dumbbell shape of the nerve sheath tumours could occur anywhere except vertebral foramen as seen in our patient.

CASE PRESENTATION

A 31-year-old man reported of right upper abdominal pain and effort dyspnoea for 3 months. He had a good general condition and had no 'B symptoms'. On physical examination, the patient had tenderness during palpation of the right upper quadrant of the abdomen and there were crackles at the right base of the chest on auscultation.

The patient had no significant medical, social or family history.

INVESTIGATIONS

No significant abnormalities were found on standard blood tests.

His chest X-ray revealed homogen opacity at the inferior and middle zone of the right haemithorax.

On abdominal ultrasound, a gross solid lesion with smooth borders was detected behind the liver.

The thoracoabdominal multidetector CT (MDCT) with contrast media was performed. MDCT revealed a huge tumour, 23×17×15 cm in size, in the right posterior mediastinum extended to the right retroperitoneum. The lesion was solid, encapsulated and had diffuse mild contrast enhancement. The tumour did not contain areas of fat and calcification density (figure 1A, B).

In sagittal plane MDCT images, the lesion was like a dumbbell shape (figure 2).

Figure 1 (A) Coronal CT scan showing the right posterior mediastinal tumour that extends towards the right retroperitoneum. (B) Axial CT scan showing retroperitoneal tumour behind the liver compresses the inferior vena cava (black arrow).

DIFFERENTIAL DIAGNOSIS

A 31-year-old man presented with right upper abdominal pain and effort dyspnoea for 3 months without B symptoms. On physical examination, the patient had tenderness on the right upper quadrant of the abdomen and there were crackles at the right base of the chest on auscultation. He might have a renal stone or gallstone disease, liver parenchymal disease or lower lobe infection of the right lung. His chest X-ray revealed a homogen opacity at the inferior and middle zone of the right haemithorax and on abdominal ultrasound, a gross solid lesion was detected behind the liver. Radiological examinations revealed that the symptoms were due to the giant solid thoracoabdominal mass.

The giant solid thoracoabdominal tumour was considered as a sarcomatous tumour or a nerve sheath tumour in the differential diagnosis.

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To cite: Cuce F, Kara K, Asker M, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-203487

Figure 2 Sagittal CT scan showing a giant thoracoabdominal dumbbell-shaped tumour. The intrathoracic (black rim) and retroperitoneal (white rim) parts of the tumour are connected by a relatively narrow segment (arrow) passing through the diaphragm.

TREATMENT

The MDCT images indicated a giant sarcomatous tumour or nerve sheath tumour in the differential diagnosis. CT-guided biopsy revealed a spindle cell tumor. Surgical resection was recommended. The tumour was completely resected en bloc with a portion of adhered left lower lobe and the right hemidiaphragm. The frozen section of the pleural and the diaphragmatic margin was found to contain no tumour. The diaphragmatic defect was repaired with a graft. The final pathology demonstrated the mass consistent with schwannoma characterised by a proliferation of spindle cells.

OUTCOME AND FOLLOW-UP

The postoperative period was uneventful and the patient was discharged home without complication. At 12-month follow-up, the patient was free from recurrence.

DISCUSSION

The term of dumbbell is used for schwannoma when the intraspinal schwannoma extends through the vertebral foramen into the posterior mediastinum or retroperitoneum.^{1 2} Although dumbbell shape of tumour is not specific for nerve sheath tumour, it may give clues for primary diagnosis. In our patient, the sagittal view of the schwannoma which passed through the

diaphragm evoked the dumbbell shape similar to that known in vertebral foramen as a classic.

Giant schwannoma is rarely located in the retroperitoneum and pelvis.³ It is a slow growing tumour. As the symptoms and clinical signs are due to the compression of adjacent organs by the tumour, schwannomas could reach upto very big sizes. The differential diagnosis of a giant thoracoabdominal solid lesion includes tumours of the oesophagus, lymphoma, sarcomas besides the neurogenic tumours such as schwannoma and neurofibroma.^{4 5}

Schwannoma typically appears as a solitary, sharply demarcated and well-capsulated round mass with smooth borders on CT images.⁶ The lesion has moderate contrast enhancement and could show punctate calcifications. MRI usually demonstrates low intensity in T1-weighted images and high intensity in T2-weighted images in the tumour. The characteristics of the lesion on CT were consistent for schwannoma in our case.

Learning points

- ▶ Schwannoma should be kept in mind for the differential diagnosis of a giant solid thoracoabdominal lesion.
- ▶ The dumbbell-shaped tumour is a non-specific term for any of a number of usually benign tumours.
- ▶ The nerve sheath tumour may assume the shape of a dumbbell as a classic when the tumour extends outward the spinal canal through the vertebral foramen. This finding may contribute to the diagnosis. But dumbbell shape of the nerve sheath tumours could occur anywhere except in the vertebral foramen.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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