Retroperitoneal schwannoma: a common tumour in an uncommon site

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We describe a female adult patient who presented with acute retention of urine and vague abdominal discomfort. A provisional diagnosis of ovarian tumour was made after cross-sectional imaging. At laparotomy a very large retroperitoneal mass was biopsied and found to be a schwannoma after pathological examination. The clinical, radiological, and pathological features of this disease are discussed in this report.

Introduction

Schwannomas are nerve sheath tumours, usually found in the head, neck and on the flexor surfaces of the extremities. They are believed to be benign tumours, with few cases of malignant transformation reported. Schwannomas are rarely found in the retroperitoneal cavity. There are few reports describing the radiological features of this disorder, and even fewer local reports describing this disorder. Pathologically, it is a well-encapsulated lesion demonstrating specific Antoni A/B areas. Characteristic immunochemical features are a positive S-100 and a negative CD-34. In this report, we aimed to illustrate the clinical and radiological features of this common disease in an uncommon location.

Case report

A 48-year-old woman presented to our emergency department with a first episode of acute retention of urine and vague abdominal discomfort. A physical examination revealed an 8-cm firm mass in her pelvic region. Biochemical investigations revealed a normal white cell count and normal levels of the tumour marker, CA125.

An urgent computed tomographic (CT) scan of her abdomen and pelvis showed a 10 cm x 10 cm x 9.3 cm well-defined, contrast-enhancing pelvic mass displacing a normal, smooth uterus anteriorly. The mass had neither calcification nor any fatty component and the fat planes between the adjacent structures were preserved. Moreover, the right ovary appeared to be normal while the left ovary was inconspicuous (Fig 1). Thus, a provisional diagnosis of left ovarian tumour was made and the gynaecological team was consulted.

Management options were discussed at an inter-departmental X-ray meeting and it was decided that surgery was the best choice. The patient then underwent an operation but no solid ovarian tumour was found. Instead, a huge, matted retroperitoneal tumour with a significant mass effect on the ureters and uterus was found. Due to the technical problems anticipated with removal of the mass, an open biopsy was performed and a specimen sent for tissue diagnosis. This was reported as benign schwannoma so the mass was left in situ. The diagnosis was later confirmed after pathological examination of the specimen.

After thorough discussions between consultants from different disciplines, it was decided that surgical removal of the mass was the best course of treatment but the patient refused further surgery. She was given symptomatic relief and eventually became asymptomatic.

Follow-up CT scans (Fig 2) showed static progress with pressure erosion of the sacrum mimicking malignant bony destruction.

Discussion

Schwannomas (neurilemmomas) are nerve sheath tumours that usually affect the head, neck and the flexor surfaces of the extremities. It is rare to find a schwannoma in the retroperitoneal cavity. They usually affect adult patients aged 20 to 50 years and males are more frequently affected than females.

Radiology can be used to make the diagnosis, to assess the extent of the lesion, to look for features of malignant transformation, to monitor treatment, and to guide a tissue biopsy.

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Schwannomas have true capsules composed of epineurium. The tumour mass is characteristically eccentric with respect to the affected nerve. Retroperitoneal schwannomas show cystic degeneration in up to 60% of cases while calcification is seen in 23% of cases only.[1]

Although target and fascicular signs are characteristic radiological features of schwannoma, these are not frequently seen in retroperitoneal schwannomas. The 'target sign' is seen on T2-weighted CT images as a hypo-intense signal in the centre and hyper-intense signal in the periphery. This is caused by fibrous components in the centre surrounded by myxomatous elements in the periphery.[2,3] The 'fascicular sign' is the appearance of the fascicular bundles in neurogenic tumours. Another feature worth mentioning is the destruction of adjacent osseous structures, something widely reported in the literature and also seen in our patient, who developed sacral erosion. The exact pathogenesis is not yet known but some authors consider pressure erosion a possible aetiology.

In the absence of typical signs, diagnosing a retroperitoneal schwannoma is difficult when using cross-sectional imaging alone. Its large size can also make it challenging for the radiologist to determine its exact location because a retroperitoneal schwannoma can displace the uterus and mimic an intra-peritoneal mass. The differential diagnosis should include retroperitoneal lymph nodes and ovarian tumours. The former can be distinguished by evidence of other lymphadenopathy and the latter can sometimes be distinguished by identifying the normal separate ovaries. Computed tomography and ultrasonography should be used to play guide tissue biopsies and identify the aggressive features of a malignant schwannoma, which account for about 1% of retroperitoneal schwannomas.[4]

Schwannomas are encapsulated tumours. Microscopically, they demonstrate Antoni A areas (densely cellular, arranged in short bundles or interlacing fascicles) and Antoni B areas (fewer cells, organised, with great myxoid component). Being positive for S-100 and negative for CD-34 are another two features supporting a correct diagnosis.[5,6]

Detecting a malignant retroperitoneal schwannoma is crucial for effective management as these carry a much poorer prognosis. Its clinical, radiological, and histological features usually clinch the diagnosis. Radiologically, a malignant schwannoma has irregular margins and infiltrates the adjacent structures. Distant metastases via peri-neural and intra-neural routes are characteristic features. Histologically, an infiltrative margin with nuclear palisading is a striking feature[7]; while clinically, pain is the single most important feature differentiating a malignant schwannoma from the

後腹腔神經鞘瘤：常見的腫瘤發生在罕見位置的病例報告

一名成年女性病發時出現尿滯留及腹痛，影像學初步診斷為卵巢腫瘤。為病人進行剖腹術發現一腹腔腫瘤，經活組織檢查證實為神經鞘瘤。本文會討論神經鞘瘤的臨床、影像學，以及病理學症狀。

FIG 1. Axial and sagittal post-contrast computed tomographic scans: a huge well-defined enhancing pelvic mass is seen displacing the urinary bladder anteriorly. The fat planes between the mass and the adjacent structures are preserved; this is a feature of non-aggressiveness. The normal right ovary is noted (arrow) and the bony erosion of the left S1 is seen (arrowhead)
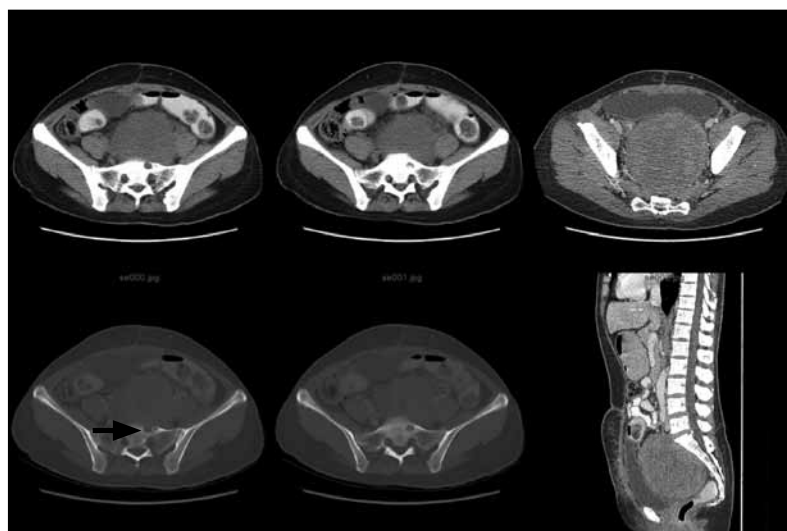

FIG 2. Follow-up axial and sagittal computed tomographic scans showing a pelvic mass and sacral destruction (arrow) with static progress. It was found to be a retroperitoneal benign schwannoma intra-operatively

benign version.

In terms of treatment, surgery is the modality of choice for retroperitoneal schwannomas. Chemotherapy and radiotherapy have only limited

roles. In malignant schwannomas, adjuvant chemotherapy or radiotherapy have marginal added benefit; some authors advocate induction chemotherapy for this condition.[8]

Conclusion

A retroperitoneal schwannoma is a rare disorder and most cases are benign. Radiology plays an important role in the management of retroperitoneal schwannoma, particularly in its diagnosis and monitoring of treatment. It is important to identify a malignant schwannoma, because it is managed differently and has a poor prognosis. The diagnosis of the malignant form is based on its different clinical, radiological, and histological features.

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