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A CASE OF RETROPERITONEAL GANGLIONEUROMA

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We describe a rare case of a primary retroperitoneal ganglioneuroma that was found accidentally during preoperative examinations conducted due to a suspicion of ovarian cancer. Computerized tomography was not able to differentiate the retroperitoneal mass from an adrenal tumor. We diagnosed the retroperitoneal tumor by means of selective adrenal angiography.

Key words: Ganglioneuroma, Retroperitoneal tumor

INTRODUCTION

Ganglioneuroma is a benign neoplasm composed of nerve fibers and mature ganglion cells. The posterior mediastinum is the most common site of origin, but this tumor may also occur in the retroperitoneum, pelvis and adrenal gland¹⁾. In Japan, primary retroperitoneal ganglioneuroma is rare in occurrence.

CASE REPORT

A 43-year-old woman was referred to our office for examination of the urinary tract. She was subjected to medical examinations for a possible ovarian cancer. A drip infusion pyelography (DIP) was carried out, and slight inferior displacement of the right kidney was demonstrated (Fig. 1). Computerized tomography (CT) revealed a small mass separable from the right kidney (Fig. 2). We suspected an adrenal tumor or a retroperitoneal mass. An endocrinological data were within the normal ranges.

Aortography and selective renal angiography did not show any tumor-feeding arteries nor tumor stain at the late phase. Selective adrenal angiography revealed that the right adrenal gland was displaced laterally and appeared to be characteristically crescent-shaped (Fig. 3). Later a retroperitoneal mass was diagnosed.

The operation revealed a retroperitoneal mass and an intact adrenal gland. The

resected specimen was a 7×5×3 cm solid mass in the retroperitoneal space (Fig. 4). The mass abutted on the right renal upper pole, and was resected completely. We found no evidence of invasion into adjacent structures. The microscopic pathology was as described below. The tumor was composed of mature ganglion cells which were large ovoid or pyriform in shape, and had one or two ovoid nuclei in an extrinsic position. In some parts of the tumor, many ganglion cells aggregated and proliferated in a neoplastic fashion (Fig. 5). We diagnosed this as a ganglioneuroma of the retroperitoneum.

DISCUSSION

In this patient, CT was not able to differentiate the retroperitoneal mass from an adrenal tumor. The normal right adrenal gland is triangular in shape when visualized by selective adrenal angiography²⁾, but demonstrated characteristic opacification in this case. It seems to be useful to perform selective adrenal angiography in this type of situation.

Ganglioneuroma originating retroperitoneally grow slowly, attain a large size and cause symptoms of compression and mechanical displacement. Primary retroperitoneal ganglioneuroma is a rare disease, and the tumor itself does not cause any symptoms except for producing homovanillic and vanillylmandelic acids. It is difficult to detect these tumors when small.

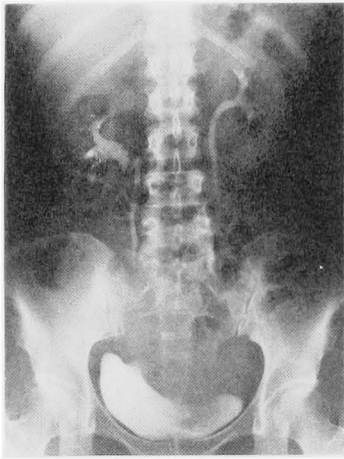


Fig. 1. DIP shows slight displacement of the right kidney inferiorly.

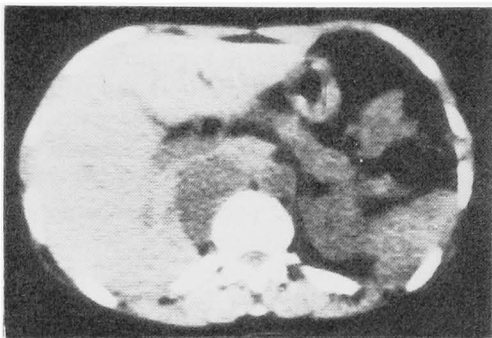


Fig. 2. CT reveals a low density mass on the right side of vertebral bone.

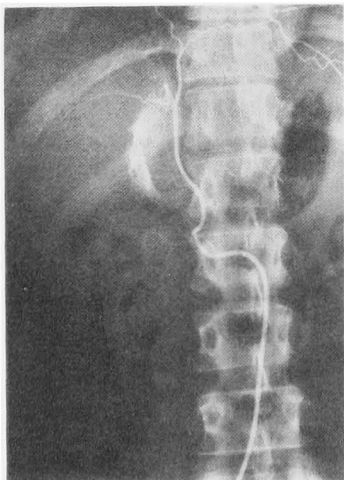


Fig. 3. Selective adrenal angiography demonstrates laterally-displaced right adrenal gland, which seems to be crescent-shaped.

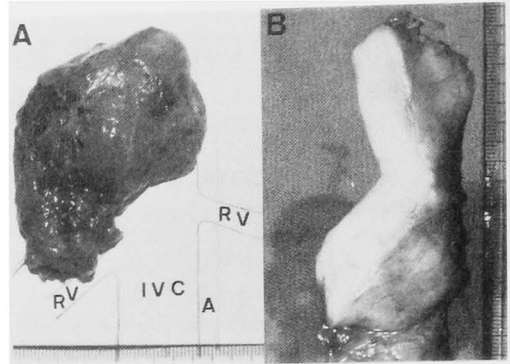


Fig. 4-A. Excised tumor appears as a well-encapsulated solid mass.
 B. Cut surface reveals the white and homogenous appearance of a ganglioneuroma.

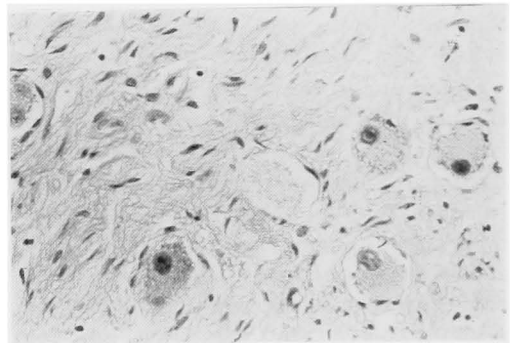


Fig. 5. Microscopic view of ganglioneuroma. The tumor was mainly composed of mature ganglion cells, but some of the ganglion cells have aggregated and proliferated in neoplastic fashion.

Our case was accidentally detected in the course of preoperative examinations.

Stout reported 107 cases of ganglioneuroma which originated in the retroperitoneal space, and retroperitoneal origin is not necessarily rare in occurrence³⁾. In the Japanese literature, 36 cases of ganglioneuroma have been reported to originate in the retroperitoneal space.

Retroperitoneal ganglioneuroma is virtually impossible to differentiate from other retroperitoneal tumors without laparotomy.

A well-differentiated benign ganglioneuroma is composed of mature ganglion cells which are large, pyriform or irregular in shape.

Stout stated that approximately 25% of ganglioneuroma are not truly benign, but contain poorly differentiated elements and

may metastasize³⁾.

Adequate excision generally leads to cure. The patients should be kept under long-term medical observation.

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和文抄録

後腹膜神経節細胞腫の1例

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症例は、卵巣腫瘍で当院婦人科へ入院の患者で、術前排泄性腎盂造影検査を受けた。この結果右腎がやや下降していたため、CT検査を行い副腎腫瘍が疑われた。しかし選択的に副腎動脈造影を施行したところ、

正常副腎が描出され、副腎の hypovascular mass か後腹膜腔腫瘍が疑われた。上記診断のもとに、手術を行ない、後腹膜腔発生の神経節細胞腫と診断した。後腹膜腔原発の神経節細胞腫としては、われわれの症例が本邦第37例目であると考えられる。

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