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Imaging Findings of Retroperitoneal Ganglioneuroma[1]

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Purpose: To characterize the typical radiologic appearance of ganglioneuromas of the adrenal gland and extra-adrenal retroperitoneum.

Materials and Methods: The findings of diagnostic imaging studies (CT, $n=5$; ultrasound, $n=1$) involving six patients aged 19 - 58 years with pathologically proven ganglioneuroma were retrospectively analyzed by three radiologist in terms of the lesions' size, shape, margin, location, CT attenuation (unenhanced/contrast-enhanced), necrosis, calcification, relationship with adjacent vessels, and US echogenicity.

Results: The maximum diameter of the six tumors ranged from 10 to 14 (mean, 11.3) cm, and the margin was well-defined in all cases. The homogeneous or slightly heterogeneous attenuation demonstrated at unenhanced CT was less than that of muscle. Dense nodular calcification was present in one case. At contrast-enhanced CT, enhancement was poor ($n=5$), mild and septum-like, or delayed, heterogeneous and focal ($n=3$), or involved subtle foci ($n=1$). In no case was there evidence of necrosis or hemorrhage. Local invasion was absent, but adjacent vascular encasement ($n=2$) or displacement ($n=2$) occurred. Ultrasonic examination demonstrated low echogenicity and mild heterogeneity ($n=1$).

Conclusion: A ganglioneuroma is an uncommon benign neural crest tumor which should be included in the differential diagnosis of a retroperitoneal mass which presents as a well-defined tumor, tend to encase or displace adjacent major blood vessels, and shows low attenuation at unenhanced CT and poor or septum-like focal enhancement at contrast-enhanced CT.

Index words : Ganglioneuroma
Retroperitoneal space, neoplasms
Computed tomography (CT)
Ultrasound (US)

Ganglioneuromas are rare benign neurogenic neoplasms which occur in children and adults. Neurogenic tumors arising from the autonomic nervous system include ganglioneuromas, ganglioneuroblastomas and neuroblastomas, among them ganglioneuromas demonstrate the most mature form (1). Ganglioneuromas are commonly located in the posterior mediastinum and retroperitoneum (2, 3), are slow growing, and are usually hormonally inactive. Patients are therefore, often asymptomatic even though the mass is large (3, 4). The aim of this study is to characterize the typical radiologic appearance of retroperitoneal ganglioneuroma.

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Materials and Methods

Between 1996 and 2001, we encountered six patients [M:F = 2:4; age, 19 - 58 (mean, 40) years] with ganglioneuroma of the adrenal gland and extra-adrenal retroperitoneum. We retrospectively reviewed the CT images relating to five of the six, and the US images relating to one; the presence of the tumors was pathologically confirmed by surgical resection ($n = 5$) or needle biopsy ($n = 1$). Two patients presented with abdominal pain, but the mass was found incidentally in the remaining four. No patient was found to have hypertension or endocrine disorders. Two neoplasms originated from the retroperitoneum, without involvement of the adrenal gland, but in the remaining four cases, the origin of the mass, either adrenal or extra-adrenal, was uncertain.

Because the patients in this study were seen at several hospitals over a five-year period, imaging studies involved the use of various equipment and techniques. Three radiologists analyzed the imaging features of tumor in terms of their size, shape, margin, location, CT attenuation (unenhanced/contrast-enhanced), necrosis, calcification, relationship with adjacent vessels, and US echogenicity.

Results

The clinical features and imaging findings of the six ganglioneuromas are summarized in Table 1. CT and US depicted a well-defined mass in all six patients. The tumor was located in the right retroperitoneum in three cases, in the left retroperitoneum in one, in the right extra-adrenal retroperitoneum in one, and in the left extra-adrenal retroperitoneum in one. The maximum diameter of the six tumors ranged from 10 to 14 (mean, 11.3) cm; they were oval in shape, while the other three showed some lobulation.

The homogeneous or mildly heterogeneous attenuation observed in five of our six patients at unenhanced CT was less than that of muscle. Dense nodular calcification was noted in one case (Fig. 1A). Contrast-enhanced CT demonstrated poor enhancement in all cases (Fig. 1B), and in three of five lesions, mild septum-like or ill-defined focal delayed enhancement was observed (Fig. 1C). In one tumor, subtle enhancing foci were visible. In none was there evidence of necrosis or hemorrhage, and no local invasion was noted; adjacent vascular encasement ($n = 2$) or displacement ($n = 2$) had, however, occurred (Figs. 2A and 2B). Although the tumors showed an expansile growth pattern, with encasement of surrounding vessels, vascular compromise did not occur.

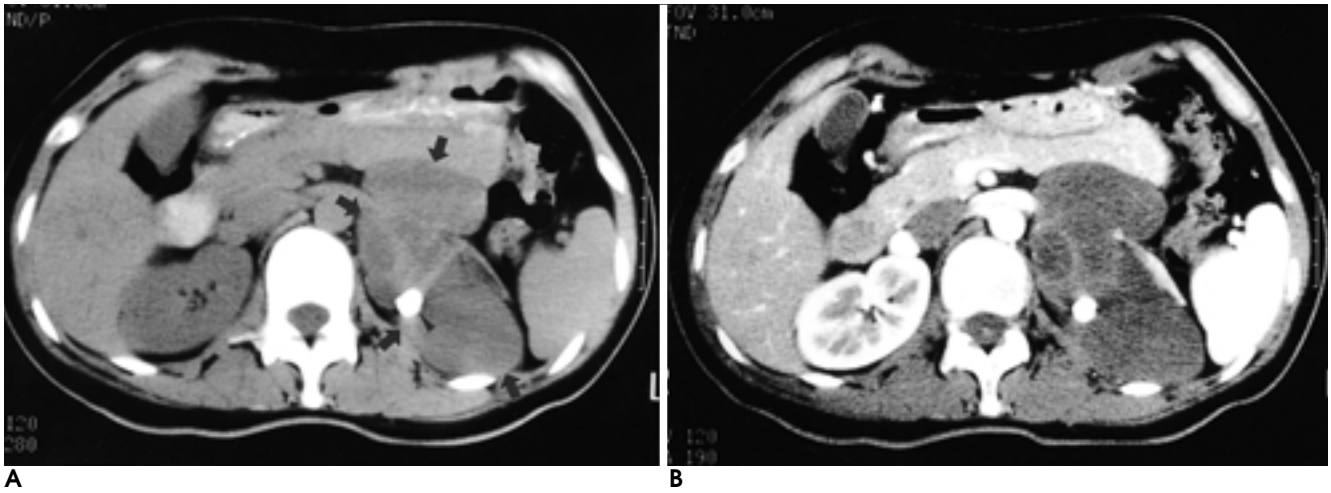
Ultrasonic examination was performed in one patient prior to surgery, and low echogenicity with mild heterogeneity was demonstrated. Ultrasound showed that this mass had smooth, well-defined margins (Fig. 3).

The histological and immunohistochemical analysis of all surgical or biopsy specimens revealed benign ganglioneuroma, and there was no immature component.

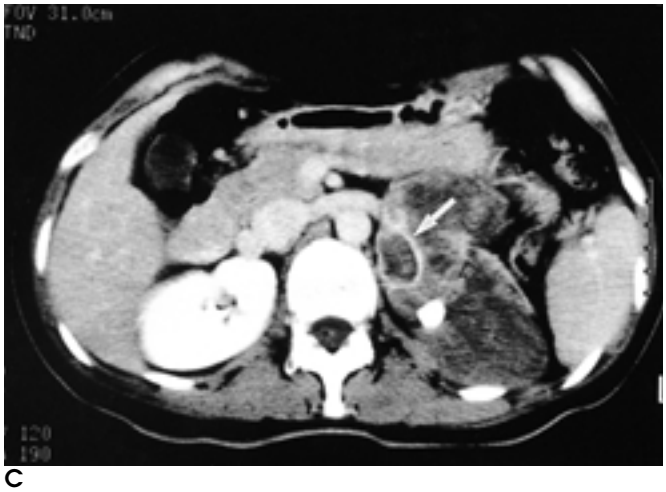
Table 1. Imaging Findings in Six Patients with Adrenal and Extra-Adrenal Ganglioneuroma

Patient No./ Age (y)/ Sex	Size (cm)	Shape	Margin	Location	CT attenuation (Unenhanced/ Contrast-enhanced)	US echogenicity	Necrosis	Calcification	Adjacent vessels
1/58/M	13× 8	Ovoid	Well-defined	Right retroperitoneum	Homogeneous low/ poor	-	-	-	Encasement (IVC, RV)
2/25/F	11×10	Lobulated	Well-defined	Right retroperitoneum	Slight low/poor (S)	-	-	-	Encasement (IVC, RV, Portal vein)
3/19/F	10×8	Ovoid	Well-defined	Right retroperitoneum	-	Heterogeneous hypoechoic	-	-	-
4/33/F	14×10	Lobulated	Well-defined	Left retroperitoneum	Homogeneous low/ poor (S)	-	-	Nodular	-
5/51/M	10×7	Lobulated	Well-defined	Left extra-adrenal retroperitoneum	Homogeneous low/ poor (S)	-	-	-	Displacement (IVC, RA)
6/52/F	10×8	Ovoid	Well -defined	Right extra-adrenal retroperitoneum	Low/ subtle enhancing foci	-	-	-	Displacement (IVC)

(S) = septum like delayed enhancement
IVC / RV/ RA/ = inferior vena cava / renal vein/ renal artery

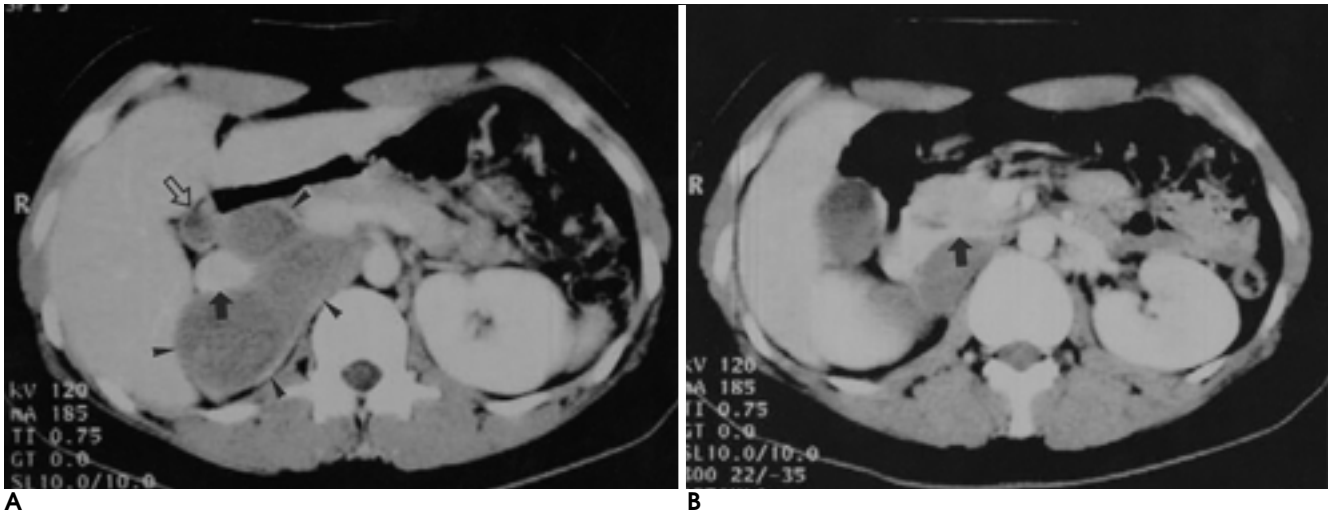


A B



C

Fig. 1. A 33-year-old woman presented with incidentally detected abdominal mass.
A. Unenhanced CT shows a mass (arrows) with homogeneous or mildly heterogeneous attenuation of less than that of muscle in the left retroperitoneum. Note a dense nodular calcification (arrowhead) in the mass.
B. Contrast-enhanced CT in early arterial phase shows poor enhancement of the mass.
C. Contrast-enhanced CT in late excretory phase shows septum-like (white arrow) and heterogeneous focal (open arrow) enhancement.



A B

Fig. 2. A 58-year-old man presented with incidentally detected abdominal mass.
A. Contrast-enhanced CT shows poor enhancing right retroperitoneal mass (arrowheads) encircling the inferior vena cava (arrow). Open arrow indicates the gall bladder.
B. Left renal vein (arrow) is stretched by the tumor. Note symmetric parenchymal enhancement of both kidneys. There is no discrepancy of both renal enhancements suggestive of no vascular compromise.

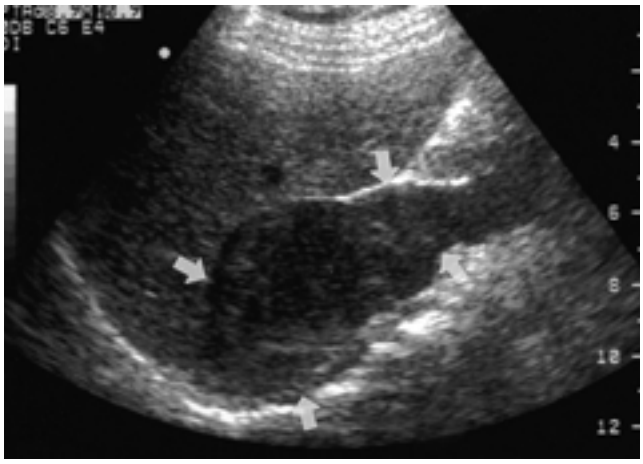


Fig. 3. A 19-year-old woman presented with abdominal discomfort.
Longitudinal transabdominal sonogram shows heterogeneous hypoechoic mass (arrows) in the right retroperitoneum. The mass has smooth, well-defined, and lobulated margin.

Discussion

Ganglioneuroma is a rare benign neoplasm arising from the sympathetic nervous system. According to reports, 60% of all patients with these tumors were aged less than 20 at the time of diagnosis, and only 5% were older than 60 (1, 3).

Ganglioneuromas occur most commonly in the posterior mediastinum (43%), followed by the retroperitoneum (32%) and neck (8%). They also occur, however, in the adrenal gland, skin, tongue, appendix, and lymph nodes (3, 5).

Even after becoming large, in most patients they are usually asymptomatic for a long time, because of hormonal inactivity. Often they are detected incidentally while a patient is undergoing abdominal imaging studies for unrelated reasons (6), and this was the case with four patients in our series. Hormonally active forms have been reported to involve the secretion of catecholamine, vasoactive intestinal polypeptide, or androgen hormones, thus explaining symptoms such as hypertension, diarrhea, or virilization (4, 7).

At CT imaging, retroperitoneal ganglioneuromas appear as well-defined, large, oval or lobulated masses which in most cases tend to encase or displace adjacent blood vessels without compromising blood flow (3, 6 - 8). Local invasion of vascular structures, including the adrenal vein, renal vein and inferior vena cava has rarely been reported, and did not occur in our cases. In contrast, vascular invasion is a feature of more than 50% of adrenal carcinomas (6).

At unenhanced CT, the homogeneous attenuation demonstrated is less than that of muscle. The tumors contain calcification in about 20% of cases (8), a feature which in childhood may help differentiate ganglioneuroma from neuroblastoma (3). In the latter, calcification is more often amorphous and of a rough pattern (3, 9), while in ganglioneuromas it is usually discrete and punctate (8, 10). In one of our patients, however, calcification was dense and nodular. At enhanced CT, varying degrees of contrast enhancement have been described; Ichikawa et al. (10), for example, reported delayed heterogeneous enhancement, and in our case, enhancement was delayed, septum-like and heterogeneous. Variations are explained by the presence in the tumors of an abundance of myxoid matrices, resulting in delayed and progressive accumulation of contrast material in extracellular space.

Ultrasound can be used to determine the presence of a ganglioneuroma, suggested by findings of homogeneous low echogenicity and a well-defined margin. Neuroblastomas and ganglioneuroblastomas frequently contain focal areas of necrosis, hemorrhage, and calcification, tissue heterogeneity which results in more echogenic interfaces than is the case for ganglioneuromas (5).

In conclusion, a ganglioneuroma is an uncommon benign neural crest tumor which should be included in the differential diagnosis of retroperitoneal masses presenting as a well-defined large tumor with a tendency to encase or displace adjacent major blood vessels.

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목적: 부신과 부신 외 후복막강에 발생한 신경절신경종의 특징적인 방사선학적인 소견을 분석하고자 하였다.
대상과 방법: 병리학적으로 증명된 신경절신경종을 갖는 19 - 58세 연령의 6명의 환자에 대한 방사선학적 소견을 후향적으로 분석하였다. 세 명의 방사선과 의사가 종양의 크기, 모양, 경계, 위치, CT 감쇠 (조영증강 전/ 조영증강 후), 괴사, 석회화, 주변 혈관과의 관계, 그리고 초음파 에코에 대하여 영상소견을 분석하였다.
결과: CT는 5명에서, 초음파는 1명에서 시행되었다. 이들 6예의 장경은 10 cm에서 14 cm (평균, 11.3 cm)으로 측정되었다. 모든 증례에서 경계는 잘 구분되었다. 비조영증강 CT 에서 근육보다 낮은 균질하거나 약간 비균질한 감쇄를 보였다. 1예에서 치밀한 소 결절성의 석회화가 보였다. 조영증강 CT에서는 거의 조영증강이 되지 않았고 (n=5), 격벽과 같거나 비균질한 초점성의 지연 조영증강 (n=3), 또는 미약한 초점성의 조영증강 부위 (n=1)를 보였다. 종양 내 괴사나 출혈의 증거를 보인 증례는 없었다. 국소적 침습은 보이지 않았으나 주변 혈관을 둘러싸거나 (n=2) 전위시키는 양상 (n=2)을 보였다. 초음파에서는 약간의 비균질한 저에코로 보였다 (n=1).
결론: 신경절신경종은 드문 양성의 신경능 기원의 종양이다. 후복막강 종양이 경계가 좋고, 주변 혈관을 둘러싸거나 전위시키는 경향을 보이고, 비조영증강 CT 에서 낮은 감쇄를 보이고, 조영증강 CT에서는 미약하거나 격벽과 같은 초점성의 조영증강을 보인다면 후복막강 종양의 감별진단에 신경절신경종이 포함되어야 한다.