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Imaging of retroperitoneal ganglioneuroma

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Abstract The aim of this study was to describe the radiological appearance of retroperitoneal ganglioneuroma. We retrospectively reviewed seven cases of histologically proven retroperitoneal ganglioneuroma. Ultrasound and enhanced CT were obtained in all cases, and MRI in three cases. The masses were well-circumscribed, ranged in size from 5 × 3×3 to 10 × 6×4 cm. In three cases close relationships between the tumor mass and major blood vessels were noted, resulting in vessel displacement or surrounding, but without compression or occlusion. On ultrasound examination the tumor showed a heterogeneous solid echostructure. Non-enhanced CT showed homogeneous or mildly heterogeneous low attenuation, and a punctate calcification was seen in one case. Contrast uptake was absent ($n = 1$) or delayed ($n = 6$). Progressive but incomplete enhancement was observed in three cases. On MRI, T2-weighted images showed a high signal intensity. Dynamic studies depicted the same enhancement pattern as described on CT. Ganglioneuroma is a rare tumor which should nevertheless be included in differential diagnosis of retroperitoneal masses when presenting as a well-delimited tumor with possible tendency to surround or displace major blood vessels, low density on non-enhanced CT, and delayed progressive enhancement on CT and MRI.

Key words Ganglioneuroma · Retroperitoneal neoplasms · CT · MRI

Introduction

Ganglioneuroma is a rare benign neoplasm that originates from sympathic ganglia. It belongs to the group of neurogenic tumors, including also ganglioneuroblastoma and neuroblastoma. Even if rare, the diagnosis of ganglioneuroma has to be taken into consideration in the differential diagnosis of retroperitoneal tumors. The purpose of this retrospective study was to determine the radiological appearance of retroperitoneal ganglioneuroma, with particular attention to CT and MRI findings.

Materials and methods

We retrospectively reviewed seven cases of retroperitoneal ganglioneuroma which were diagnosed at our institution from August 1987 to May 1997. The mean age was 39 years (age range 12–66 years). Six of the patients were female. In five cases the tumor was discovered incidentally by US performed for unrelated reasons. The two other patients presented with back pain and cruralgia, respectively. No patient was found to have hypertension or endocrine disorders.

Diagnosis was obtained by complete surgical resection in all patients.

Ultrasound and enhanced CT were obtained in all cases, and MRI in 3 cases. Because of the 10-year course of the study, examinations were obtained from various equipment. For this reason we detail only the parameters of the two most recent MR studies, which were performed on a equipment (Impact 1 T, Siemens, Er-

Table 1 The CT and MR imaging findings

Patient no.	Gender	Age (years)	Tumor location	Tumor size (cm)	CT findings			MRI findings		
					Attenuation on nonenhanced CT (HU)	Enhancement pattern[a]	Tendency to surround vessels	T1WI signal	T2WI signal	Enhancement pattern[a]
1	F	56	Medial	6 × 4.5 × 3	15	2	Yes			
2	F	28	Lateral (adrenal involvement)	10 × 6 × 4	32	3	Yes			
3	H	61	Lateral	5 × 3 × 3	35	3	No	Homogeneous < liver	Moderately > liver	
4	F	12	Medial	6 × 4.5 × 4	19	2	No			
5	F	66	Lateral	7.5 × 5 × 4	35	1	No			
6	F	20	Adrenal gland	8 × 6 × 5	18	2	No	Homogeneous < liver	Moderately > liver	2
7	F	29	Lateral	8 × 7 × 5.5	38	3	Yes	Homogeneous < liver	Moderately > liver	3

[a] Type 1: no enhancement; type 2: early delineation of fine internal septa and progressively increasing enhancement of majority of the tumor; type 3: progressive but incomplete enhancement of tumor, without internal septa

langen, Germany) allowing dynamic evaluation of enhancement (every minute during the first 5 min and then every 3 min up to 20 min). The T1-weighted gradient-echo sequence (TR/TE/flip angle: 122/4.8/ ms/75 °) allowed acquisition of seven contiguous slices during one 15-s breathhold. T2 weighted images were also obtained (TR/TE: 2200/80–120 ms).

Results

The imaging findings are summarized in Table 1. The tumor was located medially in two cases and laterally in five cases. The adrenal gland was involved in two cases. One of these two tumors was limited to the adrenal gland. The masses were well-circumscribed in all patients, presenting an oval shape in six cases and a lobulated shape in one case. They ranged in size from 5 × 3 × 3 to 10 × 6 × 4 cm. In three cases close relationships with major blood vessels resulted in displacement or surrounding, but without compression or occlusion.

On ultrasound examination the tumor was of slightly heterogeneous echostructure, without posterior acoustic enhancement (Fig. 1). Non-enhanced CT showed homogeneous or mildly heterogeneous low attenuation, ranging from 0 to 20 HU in three cases and from 20 to 40 HU in four cases (Figs. 2, 3). A punctate calcification was noted in one case. After intravenous injection of contrast medium, one tumor did not enhance. Contrast uptake was in the other cases initially low, resulting in three cases in the delineation of thin intratumoral septa.

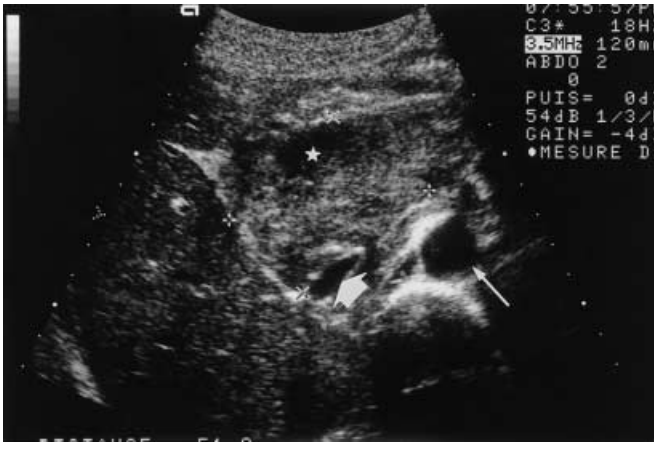


Fig. 1 Sonographic appearance of an extra-adrenal mass (patient 7). Transversal sonogram shows a heterogeneous solid mass, close relationship with inferior vena cava *(thick arrow)* and portal vein *(star)*. *Thin arrow* indicates aorta

Delayed scans showed in six cases a progressive but incomplete enhancement of the tumor, without any centripetal pattern.

On T1-weighted images the mass showed an homogeneous signal intensity lower than that of liver; on T2-weighted images the tumor showed a heterogeneous high signal, markedly more intense than that of the liver in one case, and slightly in two cases (Figs. 3, 4). Dynamic studies, available in two of three cases, depicted

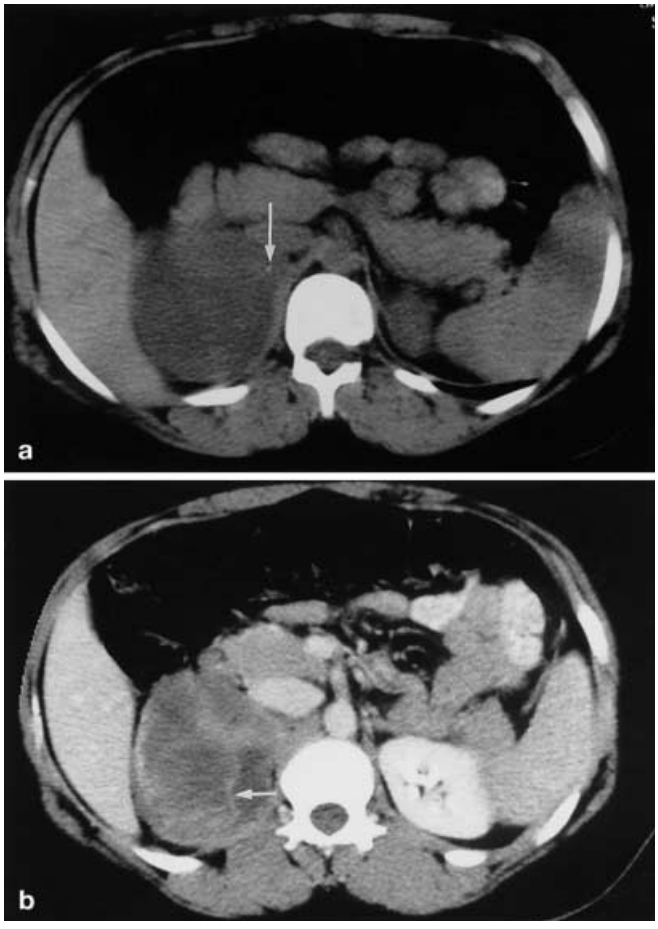


Fig. 2 a, b The CT features of an adrenal ganglioneuroma (patient 6). **a** Unenhanced CT shows a homogeneous hypodense (18 HU) mass with a punctate calcification *(arrow)*. **b** After injection, early enhancement of fine intratumoral septa is observed *(arrow)*

same enhancement pattern as described on CT scan: One case showed early enhancement of fine internal septa, and two cases showed progressive incomplete filling.

Surgical resection was easily performed in four cases but was more difficult in three cases because of the close vascular relationships. Complete resection, however, was always possible.

At pathologic examination the tumors appeared homogeneous, with no evidence of necrosis nor hemorrhage. No immature components or associated tumors were encountered.

Discussion

Ganglioneuroma is a benign tumor that belongs to the group of the neurogenic tumors, including also ganglioneuroblastoma and neuroblastoma. Neuroblastoma is a neoplasm of high malignancy, ganglioneuroblastoma of an intermediate degree of malignancy. Ganglioneuromas are benign neoplasms, but 25% of them have proved to contain poorly differentiated components such as ganglioneuroblastoma, neuroblastoma, or pheochromocytoma [1, 2, 3]. Retroperitoneum is the first or the second most common location of ganglioneuroma (32–52%). The posterior mediastinum accounts for 39–43% of the cases, neck and pelvis for 8–9% [4, 5]. Ganglioneuromas represent 0.7–1.6% of all primary retroperitoneal tumors [6]. All ages are encountered, with a predominance in children and young adults (42–60%) [2, 5, 6]. Nevertheless, the adrenal location is encountered in patients older than 40 years [7, 8]. Female patients are more frequently seen than male (gender ratio: 0.72 to 0.77) [9, 10]. In our series, six of seven patients were women.

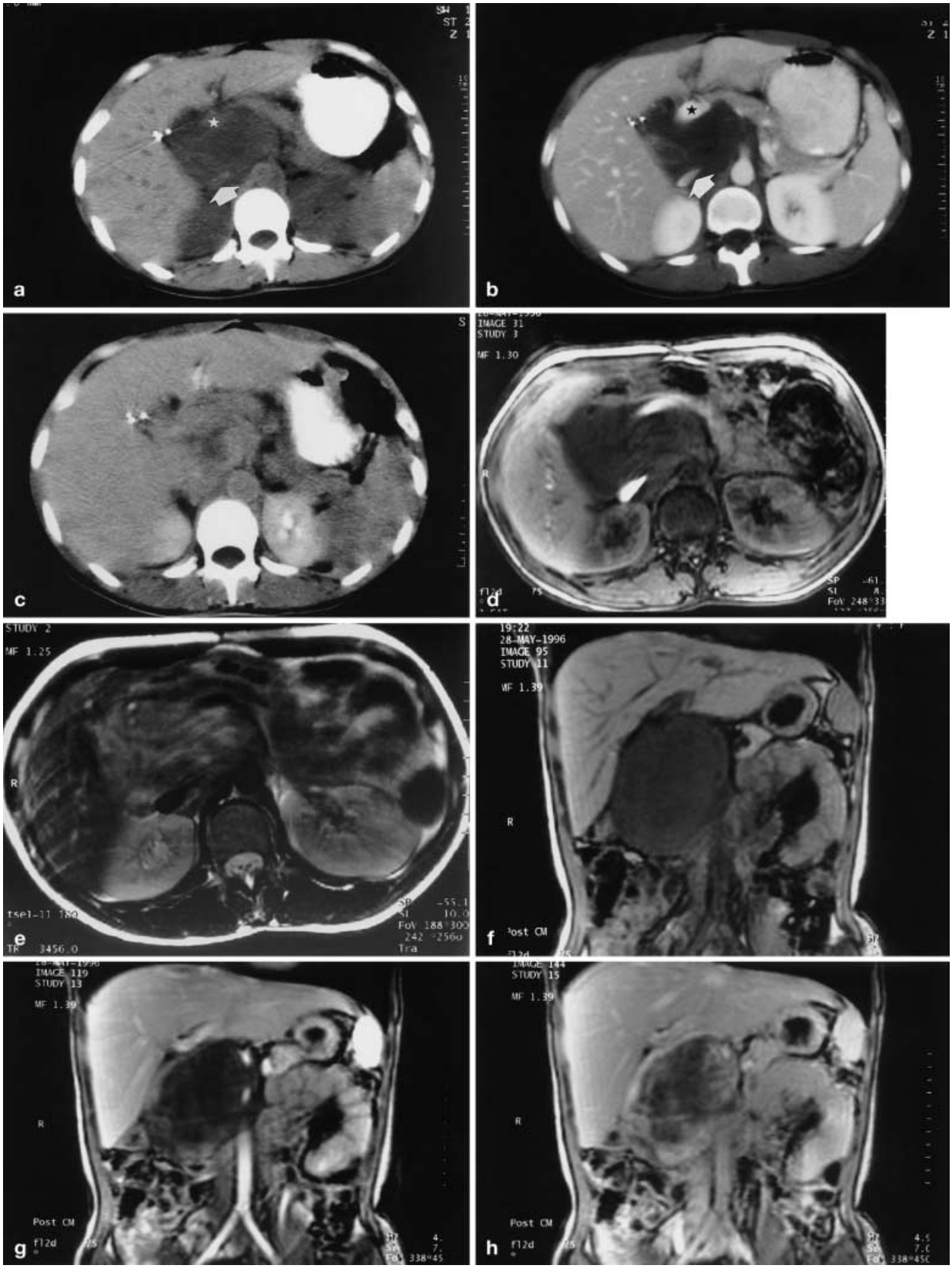
Ganglioneuromas are often asymptomatic, even if they reach large volumes; otherwise, abdominal pain or palpation of an abdominal mass are the most frequent clinical features. Hormonally active forms have been reported, the secretion of catecholamines, vasoactive intestinal polypeptide, or androgene hormones explaining symptoms such as hypertension, diarrhea, or virilization [5, 6].

Retroperitoneal ganglioneuromas are well-defined tumors with oval, crescent, or lobulated shape [2, 5]. One particular feature, present in three of our seven cases, has also been reported [3, 5], consisting of a tendency of the tumor to surround major blood vessels, resulting in absent or mild compromise of the lumen. It has to be noted that, in our series, this pattern never led to an incomplete surgical resection of the mass, since the wall of the vessels was not infiltrated by the tumor.

Sonographic appearance is not specific, the mass showing an heterogeneous solid structure. Ultrasound can be helpful in localizing the origin of the mass and in visualizing the relationship to the vessels.

Computed tomography allows precise anatomical description of the tumor: its relationship to the vessels are better depicted with helical acquisition. It is the most sensitive method in detecting calcifications which are present in 2.4–40% of cases [2, 6]. Low, non-enhanced densities are common. Enhancement usually is poor. Early enhancement of linear septa, as we observed in three cases, has not yet been reported. An interesting

Fig. 3 a–h The CT appearance of an extra-adrenal mass (patient 7). **a** Solid hypodense (38 HU) tumor with **b** poor early contrast uptake and **c** late heterogeneous enhancement. Surrounding of inferior vena cava *(arrow)* and portal vein *(star)* are clearly shown (surgical clips are related to a previous cholecystectomy). **d** Tumor shows a homogeneous hyposignal on T1-weighted images, and **e** signal intensity is moderate on T2-weighted images. Dynamic study (**f** before gadolinium injection, **g** 40 s after injection, **h** 5 min after injection) confirms the progressive contrast uptake


a
b
c
IMAGE 31
STUDY 3
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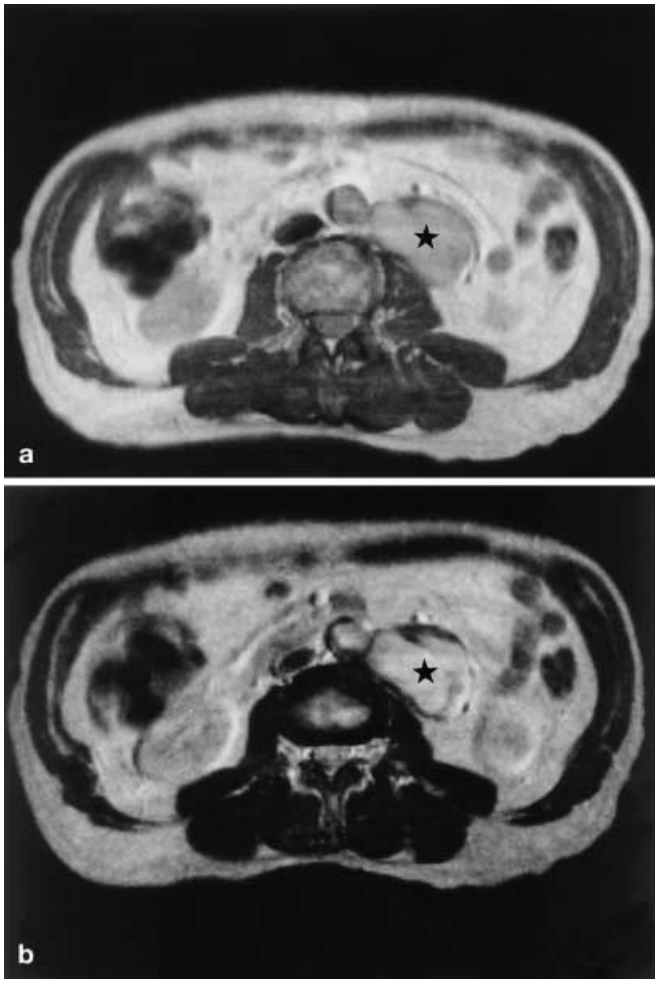

Fig. 4a, b A 61-year-old man (patient 3) T2-weighted images (TR = 2400 ms, TE = 80 and 140 ms, respectively) shows a well-delimited latero-aortic tumor presenting an increased signal intensity *(H)*

pattern was present in six of our cases, characterized by a delayed heterogeneous contrast uptake, resulting in an incomplete filling of the tumor without any centripetal characteristic. This pattern has already been reported by Ichikawa et al. [3] who attributed this type of enhancement to the presence of abundant myxoid matrices, explaining the progressive accumulation of contrast medium in the extracellular space.

This pathologic characteristic may also explain, according to Ichikawa et al. [3], the MRI appearance of ganglioneuroma, which consists of a low signal intensity on T1-weighted images and inhomogeneous but marked high intensity on T2-weighted images. We have no explanation for the relatively moderate hypersignal of two of the three ganglioneuromas that we could explore with MRI, although abundant myxoid matrices were also noted at pathological examination. Nevertheless, dynamic studies (available in two cases) were in concordance with the description by Ichikawa et al. [3] of a gradually increasing enhancement.

The seven ganglioneuromas of our retrospective study shared the characteristic of not showing any atypical component such as a less-differentiated neurogenic tumor (neuroblastoma or ganglioneuroblastoma) or a pheochromocytoma. Several cases of such composite tumors have been described in the literature, with the presence of a different component conferring to the ganglioneuroma a higher degree of heterogenicity and reducing the accuracy of lesion characterization [2, 5].

Ganglioneuroblastoma and neuroblastoma have a higher degree of cellularity and a reduced extracellular space compared with ganglioneuroma, resulting in a higher unenhanced density on CT and a relatively lower hypersignal on T2-weighted images [2]. Nevertheless, the importance of overlap between the imaging features of these three types of tumors prevents any attempt to distinguish them preoperatively [5, 6]. Ichikawa et al. [3] considers the aspect of calcifications to be helpful, since they can be discrete and punctate in ganglioneuroma, rather amorphous and coarse in malignant neurogenic tumors.

Surrounding of major vessels is also observed in both ganglioneuromas and more undifferentiated neurogenic tumors, but occlusion is usually related to malignant forms [10].

Percutaneous biopsy may orientate the diagnosis, but the frequent association with undifferentiated components limits its utility.

Distinction between ganglioneuroma and pheochromocytoma can be difficult, particularly in case of catecholamine secretion. According to Ichikaw et al. [3], T2-weighted images are not helpful in differentiating these two tumors since both have marked hypersignal, but administration of contrast material is contributive, with an intense early enhancement of pheochromocytoma in contrast to the delayed progressive enhancement of ganglioneuroma.

In conclusion, ganglioneuroma is a rare retroperitoneal tumor that should nevertheless be considered in the case of a well-delimited tumor with possible tendency to surround major blood vessels and presenting low density on nonenhanced CT and delayed progressive enhancement on CT or MRI.

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