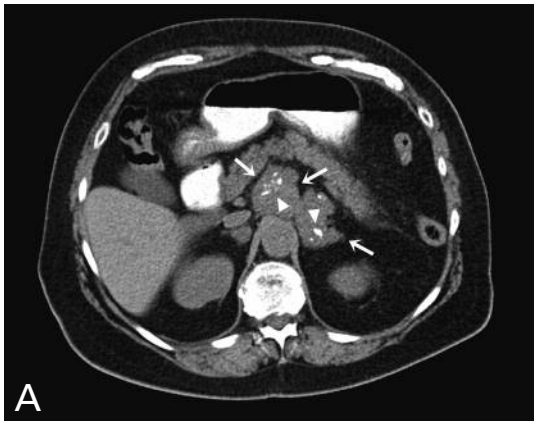
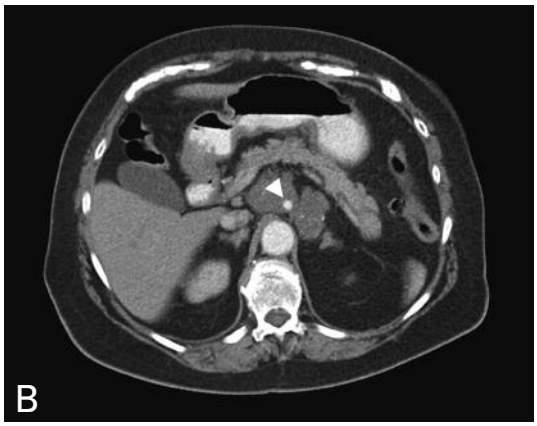
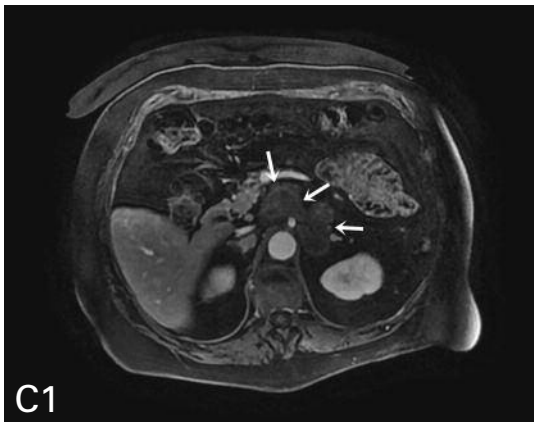
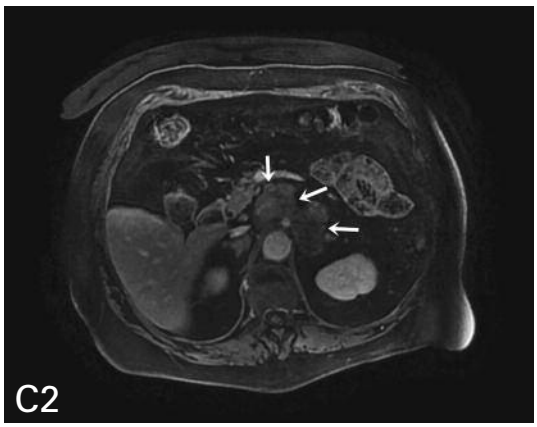
JBR–BTR, 2004, 87: 272.

IMAGES IN CLINICAL RADIOLOGY









Extra-adrenal retroperitoneal ganglioneuroma

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A 74-year-old woman presented to the hospital complaining of fatigue and atypical epigastric discomfort. Physical examination revealed epigastric tenderness. She had no relevant clinical history. Laboratory findings were within normal limits. A CT scan of the abdomen was performed and showed a large well-defined, lobulated hypodense mass in the retroperitonium (Fig. A, arrows), containing small punctate calcifications (Fig. A, arrowheads). After intravenous contrast administration the mass showed slight and inhomogeneous enhancement. The mass surrounded the celiac trunc (Fig. B, arrowhead) and superior mesenteric artery, without evidence of luminal narrowing. On MRI, the lesion was homogeneous and hypointense relatively to the liver on T1, and heterogeneous and hyperintense on T2. Axial contrast-enhanced dynamic T1-weighted MR images obtained 40 seconds (Fig. C1, arrows) and 100 seconds (Fig. C2, arrows) after contrast material injection showed gradual but heterogeneous enhancement of the mass. The radiological differential diagnosis included lymphadenopathy and neurogenic tumor. No malignancy was found elsewhere in the body. CT guided percutaneous biopsy of the mass was performed and pathology revealed an extra-adrenal ganglioneuroma. Encasement of the vascular structures made radical excision impossible. Follow-up by imaging after 6 months was recommended.

Comment

Ganglioneuroma is a rare, benign neurogenic tumor, composed of mature Schwann cells, ganglion cells and nerve fibers, arising typically along the paravertebral sympathic plexus and occasionally from the adrenal medulla. The two most common sites are the retroperitonium and posterior mediastinum, followed by the cervical region and pelvis. The tumor has been most often described in adolescents and young adults, but all ages can be affected. Clinical presentation is often asymptomatic. Most abdominal ganglioneuromas are hormone silent and detected incidentally by abdominal imaging studies. The prognosis is excellent and recurrence is rare after surgical resection.

CT and MRI can be used to develop a differential diagnosis. Retroperitoneal or adrenal ganglioneuromas appear as a well-circumscribed oval or lobulated mass. CT images show a homogeneous, hypo-attenuating mass containing discrete and punctate calcifications in 20% of the cases. Typically, slight or inhomogeneously enhancement is reported on delayed scans. On MR imaging, the lesions are relatively homogenous with low signal intensity on T1 WI and intermediate to high signal intensity on T2 WI. The tumor signal intensity on T2 WI is influenced by the portion of myxoid stroma to cellular components and collagen fibers. Dynamic contrast-enhanced T1-WI shows lack of early enhancement, and gradually increasing, inhomogeneous enhancement. Curvilinear bands of low SI on T2 WI, giving the tumor a whorled appearance, are characteristic. The imaging appearance, including the anatomic location, shape and internal architecture support the correct diagnosis of this tumor but only biopsy can reveal the diagnosis of a ganglioneuroma.

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