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Case Report

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Giant Retroperitoneal Ganglioneuroma Diagnosed by Endoscopic Ultrasound: A Case Report

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1. Abstract

Ganglioneuromas are benign tumors which originate from the neural crest. This tumor affects mainly young patients.

We report the case of a big retroperitoneal ganglioneuroma found incidentally. The patient was a young female that had minor abdominal pain for two years. The diagnoses of a large retroperitoneal mass was firstly evoked by abdomial ultrasonograghy and CT. The endoscopic ultrasonography with guided fine needle biopsy allowed the diagnosis of ganglioneuroma.

Despite the slow evolution and benign character, the size of these tumors and their compressive effect on adjacent organs require treatment. The surgical excision is the chosen treatment and it offers an excellent prognosis.

2. Key Clinical Message

Young women having abdominal pain for 2 years caused by a large retroperitoneal mass pushing the adjacent organs. An endoscopic ultrasound guided fine needle aspiration was performed. The histopathological findings concluded to Ganglioneuroma.

3. Introduction

The ganglioneuroma is a benign tumor that affects mainly children and young adults. It is a rare tumor which is often located in the retroperitoneal region. It belongs to the group of neurogenic tumors that develop on the sympathetic ganglion chains. This group also includes ganglioneuroblastomas and neuroblastomas. Usually, ganglioneuromas are discovered as incidentalomas since they are often asymptomatic. The diagnostic is essencially histological and the treatment is generally based on surgery.

We report the observation of a giant retroperitoneal ganglioneuroma and discuss the diagnostic, therapeutic and evolutive aspects of this tumor.

4. Case Report

A 33 years old female, with a medical history of double mitral and aortic valve replacement by mechanical prosthesis on rheumatic valve disease, consulted for abdominal pain. The onset of the symptomatology was 2 years ago with the appearance of epigastric pain with dorsal irradiation, of moderate intensity without other associated signs. On admission, the patient was in good general health, stable, apyretic. Abdominal examination found a firm, well-limited, painless mass in the epigastric region, mobile in relation to the superficial plane and fixed in the deep plane. Abdominal ultrasonography objectified a mass of about 110 mm, projecting to the epigastric region and right hypochondrium, well limited, heterogeneous, marginally enhanced by dynamic computed tomography. The CT showed a retroperitoneal mass of 118x93x120mm lateralized to the right, encapsulated, with sharp contours, tissue density, showing very low enhancement after contrast injection and enclosing thin septa. This mass pushes the pancreas and the duodenum forward, fills the subhepatic space and comes in intimate contact with the liver and the gallbladder and the right kidney. It comes into intimate contact with numerous arterial vascular structures (Aorta, right renal artery, celiac trunk, superior mesenteric artery) and venous structures (superior mesenteric vein, portal vein, inferior vena cava) which remain permeable, not invaded. The biological check-up did not reveal any abnormality. We completed with an endoscopic ultrasound which revealed a retropancreatic mass, well limited, heterogeneous, about 80

mm long, invading neither the vascular structures nor the adjacent organs. (Figure 1) No adenopathy or intraperitoneal fluid were noted. An endoscopic ultrasound guided fine needle biopsy was performed (Figure 2). The histopathological findings concluded to Ganglioneuroma as it revealed a tumor proliferation consisting of a schwannian stroma composed of bundles of spindle cells with elongated nuclei without atypia and with scattered mature lymph node cells.

The patient is scheduled for surgery to remove the tumor.

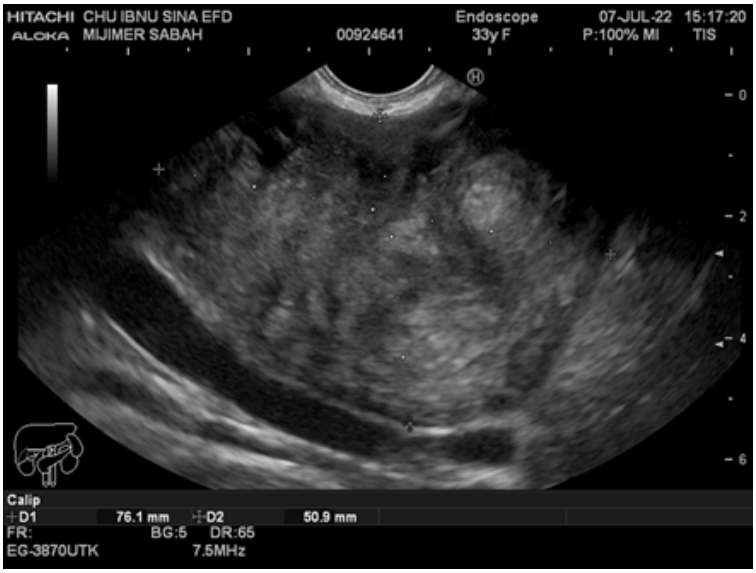

Figure 1: Endoscopic ultrasound image and sizing of the tumor

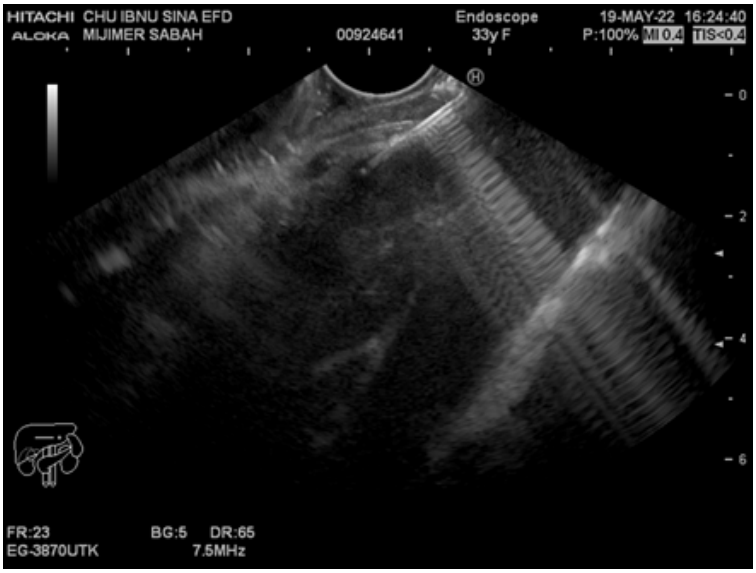

Figure 2: Endoscopic ultrasound fine needle biopsy of the tumor

5. Discussion

Ganglioneuroma is a rare benign tumor which has a neural crest origin. It arise from the sympathetic nervous system and belongs to neuroblastoma group. [1] It is composed of mature gangliocytes and a stroma containing nerve cells, in contrast to neuroblastoma and ganglioneuroblastoma, which are composed of more immature gangliocytes with a greater progression potential [1,2]. Ganglioneuroma's frequency is estimated at seven per million individuals [3]. It accounts for 69% of the sympathetic nervous system's tumors and 0.7 to 1.6% of primary retroperitoneal tumors [4, 5]. Ganglioneuroma affects mainly children and young adults [6-9]. Females are more often touched with a sex ratio of 0.72-0.77 [4, 10, 11]. Our case confirm this repartition as it was a young female.

GN can arise in any sympathetic tissue. Therefore it can have differents localisations : cervical, mediastinal, retroperitoneal or pelvic localization. The retroperitoneum is the most frequent localization of these tumors (32 to 52%), followed by the mediastinal localization (39 to 43%) [9, 11-13].

The mode of revelation is often fortuitous, as in the case of our patient. These tumors evolve quietly and are most often discovered accidentally. GN cases can be defined as incidentalomas. It is usually asymptomatic until it grows to a large size. Then, abdominal pain, palpation of an abdominal mass, or compression of neighboring organs or an alteration of general condition lead to the diagnosis [9]. Indeed, our patient was asymptomatic until the tumor reached a size that caused abdominal pain. Some ganglioneuromas have secretory properties that may lead to diarrhea, hypertension or signs of virilization [4, 11, 15-17].

When the mass was discovered, MRI or CT was usually performed to define the size, location, composition of the mass and its relationship with adjacent structures; specifically, this last aspect is important for the surgical approach [1, 18]. On ultrasound, it can be a heterogeneous or homogeneous mass of solid echostructure. On CT, ganglioneuromas appear as oval to round tumors, of low density and well circumscribed [4, 19]. Contrast enhancement is usually mild and heterogeneous, and fine tumor calcifications are often seen [4, 10, 11, 19]. Ganglioneuromas usually show a T1 hyposignal and a T2 hypersignal on MRI [4, 9-11, 19]. Contrast enhancement after Gadolinium injection is nonspecific, ranging from no contrast enhancement to weak inhomogeneous uptake or sometimes even very strong enhacement [6,11].

The main differential diagnoses of ganglioneuroma are ganglioneuroblastoma and neuroblastoma [10]. But imaging allows us to differentiate between these these different diagnoses. Other differential diagnoses are numerous when faced with a large tissue retroperitoneal lesion developing between normal structures [6,13]. But considerieng semiologic nuances and the context of discovery helps narrow the range of diagnosis. Thus, when faced with a solid, homogeneous retroperitoneal mass in an asymptomatic patient, some of the plausible diagnoses are or nonsecretory paraganglioma if the tumor is hypervascularized, and ganglioneuroma in case of a paucivascular lesion [6, 13]. Other diagnoses must be evoked. Among the benign tumors, teratoma and lipoma have a fatty contingent, schwannoma often has a cystic component, desmoid tumor is poorly limited, cystic lymphangioma is fluid, and retroperitoneal seminoma is accompanied by an altered general condition. Malignant lesions (primary, secondary, lymphomatous) are multiple, often necrotic, and invade the surrounding structures [6, 13]. Nevertheless, it remains difficult to discriminate a benign tumor like ganglioneuroma from other kind

of lesions: without a histopathological exam, it's hard to achieve a definitive diagnosis. The diagnosis of ganglioneuroma is anatomopathological [3, 10, 11]. A biopsy or an aspiration cytology can be very usefull but it depends on the site and the technical possibilities. The reports of ganglioneuroma diagnosed by endoscopic ultrasound guided fine needle biopsy are rare [22].

When an EUS biopsy is performed, a diagnosis of retroperitoneal ganglioneuroma may be established, preventing invasive surgery and its accompanying risks in favor of routine follow-up imaging as long as the tumor remains small and patient asymptomatic or poorly symptomatic [24].

Nevertheless, the final diagnosis will only be made after a histological study of the surgical specimen. Indeed, although the preoperative biopsy allows the diagnosis, a complete analysis of the excisional specimen is still necessary because of the possibility of neuroblastoma contingents but also of pheochromocytoma within the ganglioneuroma [6, 20].

The treatment remains surgical and consists in the excision of the tumor [1, 4, 19]. This procedure is all the more difficult as the tumor is large and has intimate contacts with the adjacent structures, especially the large vessels (IVC and aorta) [12]. Treatment should be performed early not only to confirm the nature of the mass, but also to prevent enlargement and compression of adjacent structures. The approach is usually a transperitoneal laparotomy, mainly for large masse's as in our patient [13]. The laparoscopic approach remains possible and even preferred for small well-defined retroperitoneal masses without intimate contact with the large vessels. The evolution of these tumors is slow, but volume increase is the rule in the absence of treatment. The survival rate with this tumor remains good despite incomplete tumor resection. Adjuvant chemotherapy or radiotherapy is not indicated due to the benign nature of the disease [1, 21]. Complications are mainly mechanical. Local recurrence is exceptional, but the possibility of malignant transformation into a ganglioneuroblastoma is possible, hence the interest of surveillance [1, 6, 19].

6. Conclusion

Ganglioneuromas are neurogenic tumors, rare, of essentially retroperitoneal development from sympathetic ganglion cells. They generally evolve in a low-noise manner, which explains their discovery at a stage of voluminous tumor. Imaging, in particular CT and MRI is very useful, as it allows to specify the relationship of the of the tumor with the neighboring organs which condition the complete resection of these tumors. This complete resection is the only guarantee of a good evolution, thus avoiding recurrence and degeneration.

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