Introduction
Castleman disease or giant lymph node hyperplasia is a rare disorder of the lymphoid tissue most often involving the mediastinal lymph nodes. We report a case of localized retroperitoneal hyaline-vascular type of Castleman disease. A 35-year-old woman presented with mild epigastric pain with radiation to the back. A mild anemia was the only abnormal associated finding. Abdominal ultrasound and CT-scan showed a homogeneous well-demarcated mass beneath the pancreas. Excisional biopsy on laparotomy revealed a highly vascular encapsulated mass with a diameter of 6 centimeters. Histopathologic examination was diagnostic: hyaline-vascular hype Castleman disease.

Castleman disease, although rare, is one of the differential diagnoses of a retroperitoneal mass, most often discovered incidentally or due to pain and compressive symptoms. Excision is both diagnostic and curative for management of the localized form of Castleman disease.

Keywords: Castleman disease, giant lymph node hyperplasia, localized

Case report
A 35-year-old woman was admitted to hospital complaining of vague epigastric pain during the past 2 months. The pain had radiation to the back and left shoulder and improved with leaning forward. No other accompanying symptoms were present except occasional sweating. The history and family history were negative and physical examination was normal. A mild hypochromic microcytic anemia was the only abnormal finding on laboratory tests.

Abdominal ultrasound showed a well-demarcated mass 5 cm in diameter at the inferior border of the pancreatic body. Spiral abdominal CT-scan revealed a well-defined homogeneous mass, enhancing with contrast injection (Fig. 1).

On exploration, a highly vascularized encapsulated mass with a diameter of 6 cm was found in the retroperitoneum just beneath the pancre-
atic body pushing it somewhat upward.

The inferior border of the tumor had dense adhesions to the base of the transverse mesocolon. The mass was excised completely and the defect in the mesocolon was repaired (Fig. 2). Gross pathology showed an encapsulated solid mass with a creamy-red surface. On microscopic examination, numerous evenly distributed follicles with small involuted germinal centers with hyalinization were seen, surround-
ed by small concentric lymphocytes.

In the interfollicular space, numerous post-
capillary venules and lymphoid cells were seen,
proposing the diagnosis of hyaline-vascular
type Castleman disease (Fig. 3). The patient
was discharged after complete recovery.

Discussion
The localized form of Castleman disease first
described by Castleman and Towne in 1956 is a
rare disease involving the lymphoid system,
most commonly the mediastinal and cervical
lymph nodes [1].

Retroperitoneal involvement is rarely seen;
in a report of 81 cases, only one was abdominal
[2,3].

Castleman disease is seen in all age groups,
with a mean age of 43 years at diagnosis [4].

The etiology is suspected to be autoimmune
in origin. Overproduction of interleukin-6 by
the lymphoblastic lymph node is believed to
have a central role in both the localized and the
multicentric form. Two histological variants
have been described. The more frequent hya-
line-vascular type shows small hyaline-vascu-
lar follicles and interfollicular capillary prolif-
eration. The plasma cell type is characterized
by large follicles with intervening sheets of
plasma cells. This type is often associated with
systemic anemia, elevated sedimentation
rate, hypergammaglobulinemia and hypoalbu-
minemia, all normalizing after resection of the
tumor. The hyaline-vascular type is not associated
with systemic manifestations and is usually dis-
covered incidentally or due to compressive
symptoms. Only minor hematologic abnormal-
ities are reported in a few patients (3%); as in
our case with the only abnormal finding being
mild anemia [2].

“Multicentric” Castleman disease indicates a
clinicopathologic entity characterized by the
histology of Castleman disease of “mixed”
type, a predominately-lymphadenopathic presentation consistently involving peripheral nodes, manifestations of multisystem involvement and association with Kaposi sarcoma, Human Herpes Virus-8, “POEMS” syndrome and rarely AIDS [5,6].

The hyaline-vascular type lesion usually presents as a single rounded mass with a median size of 5 to 6 centimeters. Adjacent lymph nodes are sometimes enlarged, often similar in structure to the main mass [2].

On ultrasound, a uniform hypo-echoic mass similar to lymphoma can be seen. CT-scan usually shows a solid homogeneously enhanced hypo- to isodense mass. On MRI a nearly isointense to muscle in signal, intensity on T1-weighted image and heterogeneous signal characteristics within the mass on T2-weighted image can be seen [7].

Prominent vascularity of the mass and excessive bleeding is sometimes encountered, correlating with the microscopic vascular proliferation and contrast enhancement on CT-scan. This feature, as encountered in our case, can be challenging for the resection of the tumor, especially in the retroperitoneal and peripancreatic region.

The treatment of choice for localized disease is surgical excision that is both diagnostic and curative. Radiotherapy can also achieve clinical response and cure in selected patients. Multicentric castleman disease does not benefit from surgery and is best treated with combination chemotherapy [8,9].

Discussion

Castleman disease, although rare, is one of the differential diagnoses of a retroperitoneal mass. It is most often discovered incidentally or due to pain and compressive symptoms. Excision and histopathologic examination leads to the diagnosis and is simultaneously curative for the localized form of Castleman disease.

References