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Castleman's disease -- a rare disease of lymph nodes

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month with WBC count of 4.0x10^9/L and Platelet count of 538x10^9/L. Digital gangrene did not progress beyond the affected areas at the time of admission.

Discussion

Clonal thrombocytosis was excluded as a cause of thrombocytosis by the bone marrow examination in this patient. Besides infection of gangrenous digits, no other site of infection was identified and as she had received antibiotics prior to being admitted in the hospital, no infective organism could therefore be isolated. However with the institution of antibiotics, hydration and antplatelet therapy, the patient improved systemically and the digital gangrene stabilized without local infection and with progressive delineation of viable and gangrenous parts of digits.

Although the exact mechanism for reactive thrombocytosis is unknown, it may result from persistent overproduction of one or more thrombopoietic factors that act on megakaryocytes or their precursors in addition to principal factor thrombopoietin (TPO) which is expressed primarily in liver but is also found in bone marrow, spleen and kidney. This would indicate an endocrine as well as paracrine role of TPO action in regulating thrombopoiesis.

It may be due to the overproduction of pro-inflammatory cytokines, such as interleukins IL-1, IL-6, IL-11 and TNF that occurs in chronic inflammatory, infective and malignant states. The presence of elevated levels of IL-1, IL-6, C-reactive protein, granulocyte colony-stimulating factor (G-CSF), and granulocyte-macrophage colony-stimulating factor (GM-CSF) in individuals with this condition suggests that these cytokines may be involved in reactive thrombocytosis states. IL-6 is mainly produced by monocytes, but it can also be produced by lymphocytes, endothelial cells and fibroblasts. Infusion of IL-6 have shown an increase in platelet numbers. More than 80% of platelets with reactive thrombocytosis have raised IL-6 levels.

References


Case Report

Castleman's disease of the duodenum

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Abstract

Castleman's disease is a rare lymphoproliferative disorder of uncertain origin. Just two cases of Castleman's disease of the gastrointestinal tract have been reported. These were found in the stomach. However, as far as we know, Castleman's disease of the duodenum has not been reported. This is the first report of hyaline vascular subtype of Castleman's disease at the duodenum.

Introduction

Castleman's disease, or angiofollicular lymphoid hyperplasia, or angiomatosus lymphoid hamartoma is a rare lymphoproliferative disorder of uncertain origin, which was first described in 1956. The pathologic characteristic of this disease is hyperplasia of the lymph follicle, multiple blood vessel penetration, and infiltration of plasma cells in the interfollicular area. In 1972, Keller et al. named this disease as Castleman's disease and divided into two types: hyaline vascular (HV) type, and plasma cell (PC) type, and this classification is still used.

To our knowledge, only two cases of Castleman's disease of the gastrointestinal tract have been reported and both were in the stomach and no case of duodenal disease has been reported. We report a rare form of this disease with
a literature review.

Case Report

A 49-year-old woman presented with vague abdominal discomfort and nausea for four years. Physical examination was unremarkable, laboratory data and X-rays were within the normal limits. Abdominal ultrasound showed a well-defined, 4x5 cm mass at the tail of the pancreas which was verified in the CT scan lying in the 4th segment of the duodenum (Figure-1).

![Figure-1: Oral and IV contrasted CT scan. A solid mass with homogeneous contrast enhancement at the duodenal wall.](image)

Upper gastrointestinal system endoscopy was normal as only 2nd part of duodenum could be seen with the scope.

The patient therefore underwent laparotomy. A 4.2 x 4 x 5.5 cm tumour was found in the 4th segment of the duodenum (Figure-2) but no intra-abdominal lymphadenopathy or any visceral abnormality was seen. The tumour was dissected from duodenum and duodenum repaired by one layer single sutures. Postoperative recovery was uneventful and patient was discharged on 7th postoperative day.

![Figure-2: A- Picture of the mass before resection. B- Picture of duodenum after resection of the mass, just before the serosal repair.](image)

Histopathological examination of the specimen showed small distinct vascular follicles surrounded by palisading layers of small lymphocytes with radially arranged capillaries consistent with the diagnosis of HV variant of Castleman’s disease.

Six months after surgery the patient is fine with no relapse.

Discussion

Castleman’s disease is characterized by cancerous growth in the lymph node tissue throughout the body. Most often the mediastinal lymph nodes are affected. There are two main types of Castleman’s disease i.e. HV type and PC type, HV type accounts for approximately 90% of the cases. Most patients with HV type are asymptomatic and have a solitary lesion. Treatment is surgical resection of the primary lesion and the recurrence rate is low when the resection is complete. PC type frequently occurs in multicentric fashion and is associated with symptoms, like fever, splenomegaly, and leukocytosis. Some cases of PC type transform to malignant lymphoma and Kaposi’s sarcoma (KS). Lymphoma and KS occur during the course of multicentric Castleman’s disease in 18 and 13 percent of cases, respectively.5,6

Though definitive diagnosis necessitates histological analysis, radiological features may be helpful. HV lesions often demonstrate fine calcification that is evident even on plain X-rays, but our patient had a normal roentgenogram. On CT or magnetic resonance imaging, lymphoid lesions, which are involved, typically demonstrate homogeneous contrast enhancement, as was seen in our case and this distinguishes Castleman’s disease from other masses such as thymomas or lymphomas, which generally show no enhancement on CT scan.7

The most common site of involvement of hyaline vascular-type Castleman’s disease is the mediastinum,2 but any lymph node site can be involved8 and there are reports of this disease in the porta hepatis and hepatoduodenal
ligament. Complete surgical excision is virtually curative in all cases reported so far however, local recurrence has been observed after subtotal or partial resection.

References

Case Report

Adult Unilateral Supraglottitis as a consequence of Acute Submandibular Sialadenitis

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Abstract

Acute Submandibular sialadenitis (ASS) may spread to sublingual and submandibular spaces. On rare occasions it can spread along the floor of the mouth, to the base of the tongue and to the pre-epiglottic space, valleculae and epiglottis, leading to supraglottitis. Adult supraglottitis is an acute inflammation of the supraglottic structures. Unilateral supraglottitis in an adult as a consequence of ASS is a unique complication as described in this case report. Sore throat with severe odynophagia and tachycardia were the early warning signs. Early suspicion, prompt airway management, antibiotics, proper rehydration and analgesics are the mainstay of treatment.

Introduction

Adult Supraglottitis is an uncommon but potentially life threatening inflammation of the supraglottic region.1 Rarely infection from the sublingual and submandibular spaces may spread along the floor of the mouth, to the base of the tongue and inferiorly to the pre-epiglottic space, valleculae and epiglottis leading to supraglottitis.2 ASS leading to unilateral supraglottitis in an adult is a unique complication. This life threatening condition may result in complete upper airway obstruction and sudden death. Early suspicion and proper evaluation is mandatory to prevent emergency airway crisis. Emergency physicians should be aware of the varied ways in which adult supraglottitis can present and always maintain a high index of suspicion.

Case Report

A 25 year old Egyptian man presented to our Emergency Department with severe sore throat, dysphagia and odynophagia and swelling in the submandibular region for the past 3 days. He had been taking treatment but there was no relief. His symptoms worsened with increased severe dysphagia for liquids and mild respiratory discomfort. The patient was admitted with an impression of acute tonsillolaryngitis. He complained of mild respiratory distress, mild trismus and severe odynophagia. On examination there was a tense right sided submandibular swelling with cellulitis extending down to the thyroid cartilage. He was mildly febrile and had a pulse rate of 100/min. BP was normal with respiratory rate of 25/min. His oxygen saturation was 98% on air. Examination of the throat was difficult because of trismus. A tender swelling on the right posterior side of the tongue was noted. The patient was managed in a high-dependency unit. An urgent x-ray lateral soft tissue neck was done. X-ray revealed thickening of the epiglottis and widening of the vallecular space. (Fig: 1) With a primary diagnosis of supraglottitis with submandibular lymphadenitis, the patient was aggressively treated with Injection Ceftriaxone, Metrogyl, hydrocortisone, analgesics and intravenous fluids. A standing order for immediate intubation in the event of respiratory distress was given.

Blood tests showed leucocytosis with normal haemoglobin. Blood chemistry was normal. Throat swab was negative. A CT scan done on the 2nd day after