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Case Study

SOLVING THE PUZZLE: A CASE REPORT OF CASTLEMAN'S DISEASE

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ABSTRACT

Castleman's disease (CD) is a rare entity that requires a high level of suspicion. The diagnosis of our patient was a challenge. The diagnosis of CD was missed in the first reading of mesenteric lymph nodes. If physicians couldn't approach final diagnosis after extensive investigations, it is worth to review patient's laboratory, radiological and histopathology results retrospectively to be sure that the diagnosis is not missed.

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INTRODUCTION

69 years old male had gastric lymphoma and treated by chemotherapy and radiotherapy 12 years ago. He was in remission and referred to our hospital because of one year history of multiple mouth ulcers at the side of tongue and inner side of lips (0.5x0.5 cm), anorexia and weight loss (27 Kg in 12 months). He had one month history of epigastric pain colicky, intermittent, no specific relieving or aggravating factors. Fever up to 40 C associated with night sweating. No gastroenterology, rheumatology, respiratory or neurology symptoms. He had history of raw milk ingestion and he was ex-smoker for 30 years ago. His physical examination showed multiple mouth ulcers and mild tenderness at epigastric area. His laboratory investigations showed WBC11.06, hemoglobin 123 gm/l, platelets 421. Erythrocyte sedimentation rate (ESR) 100, CRP 59.3. Renal, hepatic, bone profiles, Thyroid stimulating hormone (TSH), and Chest x ray were normal. Blood, urine and sputum cultures were negative. The investigation for Brucella, malaria, human immunodeficiency virus (HIV), hepatitis C and B virus (HCV &HBV), antinuclear antibody (ANA) test, *Epstein*-Barr *virus* (EBV), Cytomegalovirus (CMV), peripheral blood smear, and purified protein derivative (PPD) skin test were negative. Mouth ulcer biopsy showed aphthous ulcers and no malignancy. The ulcer improved after the dermatologist started him on local steroids with colchicine and dapsone.

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Computed tomography (CT) chest, abdomen and pelvis findings were multiple small mesenteric lymphadenopathies. Upper endoscopy finding was diffuse erythematous mucosa all over the stomach. The colonoscopy finding was multiple right superficial ulcerations and deep ulcer at ileocecal valve. Biopsies from stomach were negative for malignancy. Biopsies from the colon showed chronic inflammatory cells and no TB, or CMV, or malignancy. Capsule endoscopy had been passed successfully showed multiple small ulcers along the small intestine. Surgical biopsy from mesenteric lymph nodes reported as reactive lymphoid follicular hyperplasia and no malignancy or TB. The case was discussed again with the pathologist, who took another look on the same previous biopsies without doing any extra tests and diagnosed was CD of plasma cell type. The oncologist recommended starting rituximab with prednisone 20 mg daily. Clinically the patient became afebrile, no more oral ulcers and both ESR &CRP significantly decreased. He was discharged home in stable condition.

DISCUSSION

CD is a group of rare lymphoproliferative disorders sharing characteristic clinical and histological features .Interest in CD has grown in recent years following an association with HIV infection and herpers simplex virus 8 (Waterston, 2004 and Dupin, 2000), CD can be either localized or multicentric . Localized mesenteric CD is rare (El Demellawy, 2009 and Al-Amri, 2010). The localized form is more common (Newlon,

2007). Eight cases of CD have been diagnosed over the last 15 years at a university hospital in Saudi Arabia (Waterston, 2004). The affected age group ranges from adolescence into the seventh decade (Mohanna, 2006). The pathogenesis is unknown, but the bulk of evidence points toward faulty immune regulation, resulting in excessive B-lymphocyte and plasma-cell proliferation in lymphatic tissue (Al-Amri, 2010). The most common location for this lesion is in the chest, although it can be seen in the pelvis, retroperitonium, axilla, or in the neck (TEF, 2007). Patients present with a systemic illness that manifests as disseminated lymph nodes, constitutional symptoms (e.g. fatigue, fever, weight loss, and sweats), autoimmune abnormalities, recurrent infections, and laboratory abnormalities (e.g. anemia, hypoalbuminemia, hypergammaglobulinemia, and an increased ESR. The oral ulcers in CD has been associated with a very high incidence of autoimmune phenomena such paraneoplastic pemphigus, systemic lupus erythematosus and Sjögren's syndrome (Hsiao, 2001). There is evidence that the tumor may be present for very long periods of time before the autoimmune disease develops. Some speculate that deregulated cytokine production, especially increased production of interleukin (IL)-6, may be an important trigger for the development of autoimmunity in CD (Biondi, 1989). Histologically, three forms of the disease have been described; the hyaline-vascular type usually found in the mediastinum (about 90% of cases), the plasma cell type which involves extra-thoracic sites and a rare mixed type (Regal, 2010). Approximately 90% of cases of the localized form are of the hyaline-vascular type. Patients present with a solitary mass, and their disease typically follows a benign course (Newlon, 2007). Most cases of the multicentric form are of the plasma cell type. The hyaline vascular variant is characterized by extensive capillary proliferation and a lymphocyte-predominant infiltrate surrounding small germinal centers. Hyalinized fibrous tissue surrounding the proliferating capillaries is typically present (Barrie, 1996). The plasma cell variant has sheets of mature plasma cells within the interfollicular tissues surrounding larger germinal centers and has significantly less vascularity (Barrie, 1996). The localized form may originate from either of these variants, but the multicentric version is almost exclusively derived from the plasma cell variant. There are some benign and malignant conditions, including lymphoma and thymoma, which may appear histologically similar to CD. Immunohistologic and immunologic gene rearrangement studies of the specimens can be useful in solidifying the diagnosis. Identifying an immunophenotypically varied population of B lymphocytes with polyclonal surface and cytoplasmic immunoglobulin markers helps to confirm the diagnosis of CD and differentiate it from lymphoma (Herbelin, 1998).

It is recommended to screen for malignancies in patients with CD to exclude malignant lymphoma or Kaposi sarcoma that can occur as long-term complications of the disease (TEF, 2007). The multicentric form is aggressive and often culminates in death secondary to infectious complications or malignancy (e.g., lymphoma, Kaposi's sarcoma, or follicular dendritic cell sarcoma); malignancies have been reported to arise in as many as 32% of patients with multicentric CD (Newlon, 2007). The diagnosis of CD is established by biopsy and treatment is based on published case reports only, as there are no randomized trials of therapy (Waterston, 2004). Localized CD may be considered a disease with benign prognosis after radical surgery, with low rate of recurrence. In multicentric CD prognosis may be poor depending on number and site of lesions. Patients with the systemic form who were treated with chemotherapy and rituximab had complete remission (Al-Amri, 2010). In conclusion, if physicians couldn't approach final diagnosis after extensive investigations. It is worth to review patient's laboratory, radiological and histopathology results retrospectively to be sure that the diagnosis is not missed. Localized mesenteric CD is rare and has to be considered in the differential diagnosis of mesenteric tumors.

Conflict of interest

The authors have no conflicts of interest and no financial relationships with any organizations that might have an interest in the submitted work.

Consent

written informed consent was obtained from the patient for the publication of this case report.

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