A Case of Advanced Unicentric Retroperitoneal Castleman's Disease, Associated With Psoriasis

Mohammad Ali Mohagheghi[1], Ramesh Omranipur[1], Fereshteh Ensani[2], Alireza Ghannadan[2], Shahriar Shahriaran[3], Farhad Samiee[4], and Zahra Sedighi[5]

[1] *Department of Surgical Oncology, Cancer Institute, Imam Khomeini Hospital Center, Tehran University of Medical Sciences, Tehran, Iran*

[2] *Department of Pathology, Cancer Institute, Imam Khomeini Hospital Center, Tehran University of Medical Sciences, Tehran, Iran*

[3] *Department of Radiology, Cancer Institute, Imam Khomeini Hospital Center, Tehran University of Medical Sciences, Tehran, Iran*

[4] *Radiation Oncology, Cancer Institute, Imam Khomeini Hospital Center, Tehran University of Medical Sciences, Tehran, Iran*

[5] *Cancer Research Center, Cancer Institute, Imam Khomeini Hospital Center, Tehran University of Medical Sciences, Tehran, Iran*

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Abstract- We present here a 32-year-old male with advanced lately diagnosed, right sided retroperitoneal mass, which had been already treated due to progressive muco-cutaneous lesions clinically consistent with psoriasis, during recent four years. The advanced retroperitoneal mass resected surgically and reported as hyaline-vascular castleman disease with a dense focus of coarse calcification, on histopathology. Association of psoriasis and castlman disease is discussed in this case report.

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Introduction

Castleman disease (CD) is an angio-follicular lymph node hyperplasia, occurring along lymphoid chain. It presents as a localized or a systemic disease, most commonly in mediastinum, neck, axilla and pelvis (1).
CD with three histologic subtypes: hyaline vascular, plasma cell and mixed, and two clinical types: multifocal (multicentric) or localized (unicentric), is considered as a rare lymphoproliferative disorder. Retroperitoneal location of unicentric CD is extremely rare.

Case Report

A 32-year-old male patient was referred to Cancer Institute from department of Urology with diagnosis of huge right side retroperitoneal mass, for further diagnosis and treatment. As a known case of Psoriasis, he was treated by several oral and topical medications, mainly prednisolone, hydroxicholoroquine and methotrexate, by dermatologists. During routine fallow up, microscopic hematuria incidentally was discovered. Sonography of abdomen and pelvis revealed a 124×70 mm. Retroperitoneal mass, on right side of abdomen with close proximity to the kidney, liver, colon and pancreas and mass effect, but no invasion on inferior vena cava (IVC). CT scan on 17/3/1394 demonstrated a 140×80 mm. Heterogenous mass with relatively defined borders, and central dense calcification. It was extended from lower liver surface superiorly to pelvic area inferiorly. It had close relation to right kidney, liver, pancreas, duodenum, and colon, but did not invade to them. IVC was compressed by the mass, but was patent and right renal vein had been encircled by it (Figure 1). One previous surgical attempt at another center failed to resect the mass. The case was discussed on Cancer Institute Tumor Board and recommended to wide surgical resection.

With a midline incision abdominal cavity was approached. A huge, adherent and highly vascularized solid mass, occupying entire right abdominal cavity from inferior surface of the liver to pelvis was revealed. The mass which was located retroperitoneally, had compressed inferior vena cava, kidney, liver, pancreas and colon, and had close relation to duodenum, abdominal aorta and right side psoas muscle. Non of the above mentioned areas had been invaded by the tumor. The right renal vein had been encased and partially occluded by tumor. The mass was resected after difficult

Corresponding Author: M. A. Mohagheghi
Department of Surgical Oncology, Cancer Institute, Imam Khomeini Hospital Center, Tehran University of Medical Sciences, Tehran, Iran
Tel: +98 21 66581638, Fax: +98 21 66581542, E-mail address: mamohagheghi@gmail.com

dissection, meticulous vascular ligation and hemostasis, ligation and resection of the affected part of righr renal vein and a margin of contagios soft tissue as possible. The kidney preserved, its blood circulation has already been supplied via adrenal vessles. There was no evidence of any other lymphadenopathy or involved center elsewhere. The total blood loss estimated 550 ml and blood transfusion was not needed. Operation time was 3 h 20 min. Postoperative course was uneventful, and patient discharged after 3 days.

Histopathological and immunohistochemical findings were consistent with hyaline vascular Castleman disease (Figures 2-4). The treatment was followed by a course of radiotherapy, which was completed at 14/7/94. The patient was fallowed and 5 months thereafter, he was in good condition with no evidence of active disease. A sonography of retroperitoneal space showed a 35×23 mm. cyst, with thick membrane and clear contents, between lower pole of right kidney and inferior vena cava, which had been decreased comparing the previous study (organized hematoma).

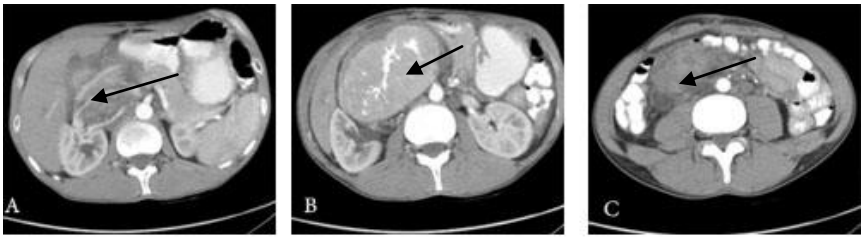
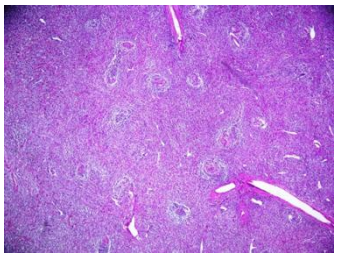


Figure 1. Contrast-enhanced computerized tomography scan shows distinctive enhancement of advanced heterogeneous retroperitoneal mass, with relatively defined borders, and close relation to the liver, kidney, pancreas (A) and central dense calcification (B), extendind downwards to the pelvis (C).



Figur 2. Hematoxylin and Eosin photomicrograph demonstrating regressing germinal centers with expansion of interfollicular areas characteristic features of hyaline-vascular Castleman's disease (magnification×10).

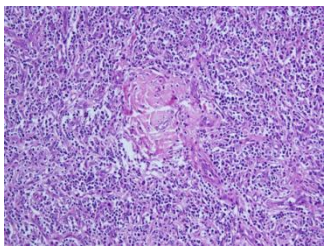


Figure 3. Hematoxylin and eosin staining photomicrographs demonstrating a germinal center with hyaline deposits(magnification×20).

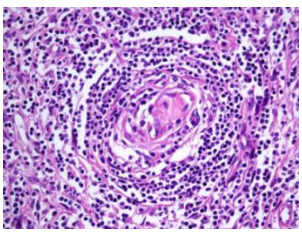


Figure 4. Hematoxylin and eosin photomicrograph demonstrating typical concentric rings of small lymphocytes forming a "lollipop" pattern (magnification×40).

Discussion

The unicentric variant of CD mostly is hyaline vascular type (>90%) (1). Hyaline vascular castleman disease usually occurs in young patients and has favourable prognosis. Histo-pathologic hallmarks of hyaline vascular are: Prominent follicles within lymphoid tissue and no sinuses with marked vascular proliferation, thick and hyalinized walls, and hyalinization of germinal centers. A capillary frequently penetrates the follicle. Concentric layering of peripheral lymphocytes resembles onion-skin and Large cells with prominent nucleoli are follicular dendritic cells (CD21+, CD35+) (2).

Radiologic hallmark of Hyaline vascular Castleman disease is mild to intensely enhancement on an enlarged solitary mesenteric or retroperitoneal mass (3).

Aa a solid retroperitoneal neoplasm, surgical resection of abnormal mass (es), whenever possible, is gold standard treatment of retroperitoneal castleman disease with excellent rate of cure. Unresectable cases with compression to adjacent organs should be treated by systemic steroids and/or radiation therapy (30-45 Gy). Radiotherapy in selected patients can assist symptom response and even cure (4).

A variety of autoimmune phenomena such as cytopenia, systemic lupus erythematosus, Sjögren's syndrome, and myasthenia gravis, has been reported with castleman disease (5). The disease has also been reported in patients with HIV infection, psoriasis, diabetes mellitus, agranulocytosus, and multiple sclerosis (6). More than 50 dermatoses, including psoriasis, have been correlated with underlying neoplasms. Cutaneous paraneopastic syndromes may before, simultaneous or after the diagnosis of neoplastic process (7).

Recently Alison V. Sears and his collegues reported castleman disease in a patient treated with prolonged anti-TNF biologic therapy for psoriasis and psoriatic arthritis. They wondered whether the emergence of CD may be linked to the prolonged anti-TNF biologic therapy (8).

The present case had been already treated for four years due to diffuse muco-cutaneous lesions clinically consistent with psoriasis. The patient's known dermatose in presented case preceded the diagnosis of CD. In fact chronic psoriasis management leaded to discovery of microscopic hematuria and finding the retroperitoneal mass at sonography. We conclude that there should be an etiologic relation, between longstanding psoriasis and different medications, including immunosuppressive drugs and Hydroxychloroquine, in our patient.

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