The Castleman’s Disease Resembling a Spinal Sheath Tumor

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Abstract

**Objective:** Castleman’s disease is a rare lymphoproliferative disorder most often found in the mediastinum. We report a patient with Castleman’s disease originated from neuronal foramens at L5 level and extended to retroperitoneal area.

**Clinical Presentation:** A 34-year-old otherwise healthy man admitted to outpatient clinic with axial back pain. There was tenderness posteriorly at palpation over the L5-S1 level. MRI revealed a posterior epidural mass extending to retroperitoneal space.

**Intervention:** Retroperitoneal approach was performed for pathological sampling of the lesion. After report of squash preparation, retroperitoneal part of the mass extracted. Pathological examination of the lesion identified the hyaline-vascular type of Castleman’s disease. Intraspinal part of the mass showed regression after radiotherapy. The patient’s symptoms resolved.

**Conclusion:** Castleman’s disease presenting as a spinal epidural mass lesion with cord compression is very rare. Surgical treatment and radiotherapy can result in an excellent outcome.

**Keywords:** Castleman’s disease; Spinal benign tumor; Radicular pain

Introduction

Angiofollicular lymph node hyperplasia, which is known as Castleman’s disease (CD), is an uncommon and poorly described disease featured by a massive lymphoid tissue. CD’s clinical findings and pathological features were first described in 1956 by Castleman et al. [1]. The pathological condition also has been featured in various names including giant lymph node hyperplasia, lymph node hamartoma, follicular lymphoreticuloma, benign giant lymphoma, angiomatosus lymphoid hamartoma, and angiofollicular mediastinal lymph node hyperplasia. Castleman disease is distinguished three pathologic based types; hyaline-vascular (HV), plasma cell (PC) and hyaline-vascular plasma cell type (HV-PC) [2,3]. These types are detected mostly as a unicentric indolent disease. A multicentric disease which is characterized with multiple lymphadenopathy, organomegaly, laboratory abnormalities and progressive course with potential of malignancy [4,5]. CD has been associated with acquired immunodeficiency syndrome and Kaposi’s sarcoma, thus, that tendency implies a viral or autoimmune etiology for this condition.

CD typically presents as a localized mass, generally in the mediastinum or abdomen, rarely with systemic manifestations and responds to local treatment including surgical resection or partial resection. Radiotherapy, chemotherapy, and steroid therapy have been employed to completely unresectable disease [6,7]. Clinical presentation, histologic subtype, regional involvement and laboratory findings have been used to predict outcome of selected treatment [8].

Although CD usually involves the mediastinum or abdomen, it may occur in other locations. In this case report, we described a 29 years old man with localized Castleman disease as a epidural lumbar disease which growth as a retroperitoneal space mass. To our knowledge and according to literature, this involvement of CD is very rare.

Case Report

A 30 year old male patients was admitted to outpatient clinic with 2 months history of back pain and radicular symptoms. He had severe blunt left sided pain which was unrelated with standing or walking and there was radiating pain posteriorly to left buttock and lower extremity. He was complain of weakness of his legs, he was able to walk only short distances. No incontinence of bowel or bladder was reported.

On his physical examination at admission, he showed tenderness while palpating left lumbar region. Although weakness of lower extremities was reported, there was no apparent any pathologic condition on the manual testing. He had no any neurologic deficit with 5/5 muscle strength. His mental status, cranial nerves, strength, and deep tendon reflexes in both the upper and lower extremities were normal. Although Romberg test was negative, straight leg raise test was positive. No organomegaly was evident.

On his laboratory findings, all parameters were normal except for minimal elevated neutrophil levels (5.95 10^9/L - Normal: 1.78 to 5.38 10^9/L) which was not interpreted as directly sign of any pathologic condition. Chest, spinal x-rays and the result
of electrocardiography were normal. T1 weighted magnetic resonance imaging demonstrated a posterior intraforaminal, extradural mass which displayed an anterior extension in the retroperitoneal space toward to posterior of the peritoneum. Spinal cord compression were not observed. There were no any intrapelvic or intra abdominal invasion. (Figure 1) The lesion enhanced with uniformly with gadolinium. The lesion was well bordered with dura and surrounding tissue.

With the aim of pathological examination, anterior portion of the lesion were submitted with anterior oblique approach to retroperitoneum. The squash preparation for cytology was performed intraoperatively. The working diagnosis was benign lymphocytic lesion. On the basis of this finding, due to intraforaminal extension of the mass, the accessible portion of the mass was extracted and posterior extension was left in place and second approach from posterior was planned intraoperatively. The entire postoperative lesion consisted of a well encapsulated with the 8x6x4 cm largest measuring. Light pink nodular areas with fatty tissues were also observed. Slices through the mass also revealed same light pink nodular areas (Figure 2). Microscopic examination showed lymphoid follicles with regressed germinal centers surrounded by a broad mantle zone composed of concentric rings of small lymphocytes and the hyalinized blood vessels with plump endothelial cells penetrated radially into germinal centers. Interfollicular areas showed extensive hyalinized blood vessels without aggregates of plasma cells.

Immunohistochemically, the follicles were composed of CD20 positive B cells. CD21 immunostaining revealed follicular dendritic network. A few scattered plasma cells were polyclonal. Kappa and lambda immunoglobulin light chains were present with approximately equal distribution (Figure 3). The hyaline vascular variant of Castleman’s disease was determined by histopathologic examination.

Postoperatively, surgical excision or radiotherapy were considered for the rest of the lesion and complications of the each treatment modality were discussed with patients. Radiotherapy with 200 cGy per day a total dose of 4000 cGy was administered to the residual lesion and regression with pure cure was observed. All laboratory findings and control screenings were normal at the one year follow-up.

Discussion

The mediastinum is the most common location for Castleman’s disease which was first described in a group of patients with localized lymph node hyperplasia resembling thymoma in the mediastinum. Since first described of CD, various locations for disease have been reported, including the retroperitoneum, orbit, neck, axilla, muscle, paravertebral space and CNS [9,10,11]. There are two main histological subtypes of Castleman’s disease described by Keller, including hyaline vascular type and plasma cell type, and later a third mix type added [2,3]. Hyaline vascular type accounts for approximately %90 of all cases and usually features a localized non-symptomatic mass. Conversely, plasma cell type, which composed %10 of all cases, may present as a symptomatic disease including fever and other laboratory abnormalities (elevated ESR, anemia, hypergammaglobulinemia). Localized Castleman’s disease regardless of subtype is mostly treated with surgical resections. Adjuvant radiotherapy has been used for incomplete surgical removal.

The differential diagnosis is challenging because presenting features can suggest infection, malignancy or collagen vascular disease [10]. The differential diagnosis with respect to our case would be various disease with spinal mass, including lymphoma, meningioma, plasma cell granuloma, schwannoma and sarcoidosis. Multicentric castleman disease, which is usually plasma cell type, present institutional symptoms such as anemia, increased ESR, fever and various physical findings including hepatomegaly, splenomegaly and skin rashes. Kaposi sarcoma and AIDS are usually accompany with such multicentric type of disease and the clinical course could be rapidly fatal [6,12,13].

The etiology of Castleman’s disease yet to be known. Keller et al. suggested that it can occur after inflammatory or infectious process according to its histological features resembling to the lymph nodes and clinical presentation including fever, anemia and elevated ESR [2]. Due to unusual presentation in localization of the disease, some authors assume that it originates as a hamartomatous process [6]. High incidence of Castleman’s disease in the patients with Kaposi’s sarcoma and AIDS tend to support an immune mediated cause after viral infection. Furthermore, that histological similarity with other autoimmune-related diseases such as rheumatoid arthritis and Sjogren syndrome, supports the immune system related mechanism.

Neurological involvement and symptoms occur up to %30 of the patients with Castleman’s disease [4]. Although, common neurological involvement manifests as a severe motor - sensory neuropathy, plasma cell neoplasm induced POEMS syndrome (organomegaly, endocrinopathy, skin changes, papilledema, thrombocytosis and peripheral neuropathy) is distinguished from Castleman’s disease variant of POEMS syndrome which has little or no peripheral neuropathy.[14] Most of the rare intracranial lesions consistent with Castleman’s disease have been interpreted to meningiomas before pathological examination. All of the lesions resected totally except in one case of subtotal resection of a left frontal lesion. In this case 4500 cGy radiation was administered for 5 weeks, postoperatively [15,16]. There is an another Castleman disease presenting as a spinal epidural mass with spinal cord compression which was treated successfully with subtotal resection and radiotherapy administration [10]. Although surgery is considered standard therapy for unicentric Castleman’s disease, patients treated with only radiotherapy demonstrated favorable results. In case of multicentric disease, chemotherapy also has been added to treatment modality [17].

Castleman’s disease should be included in a differential diagnosis of a spinal extradural mass even though it is a very rare lesion. Excellent results were reported with surgical excision of the Castleman’s disease. Although surgical excision is considered as gold standard, radiotherapy treatment has been described especially for unicentric disease. In this case, we represented a intraforaminal spinal lesion which extend to retroperitoneal...
space. After confirming of the benign characteristic of the lesion through squash preparation, intraspinal unicentric Castleman’s disease could be treated with surgical excision via relatively safe approach to the lesion and additional radiotherapy for the residual mass, with the aim of avoiding to root injury which is one of the major complications of the surgical excision of the intraforaminal lesions.

References