The Journal of Medical Research 2022; 8(2):47-49

The Journal of Medical Research

Case Report

JMR 2022; 8(2):47-49
March- April
ISSN:2395-7565

www.medicinearticle.com
Received: 30-03-2022
Accepted: 14-05-2022

Castleman Disease masquerading as the lumbosacral neurofibroma with excellent response to rituximab

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Abstract

Castleman's disease is an uncommon, lymphoproliferative disease. Its etiology is unknown. Spine involvement is very scant in Castleman's disease. The authors report on the case of a 50 year-old man who referred with saddle anesthesia and low back pain from 2.5 years ago. A CT scan showed an extra medullary mass at S1-S2 level of the spinal canal without bone involvement. The signal specs on MRI were resembling to signal of neurofibroma .The patient underwent needle biopsy. Histological examination confirmed the diagnosis of Castleman disease, hyaline-vascular type. He received rituximab and pain was dramatically relieved. Although rare, Castleman disease should be considered in the differential diagnosis of an extramedullary mass.

Keywords: Castleman Disease, Hyaline-Vascular Type, Spine.

INTRODUCTION

There are several non-neoplastic lesions that mimic extramedullary spinal cord neoplasm in their radiographic and clinical presentation. Although MRI is able to pinpoint the exact location of the lesion, it cannot determine its nature, because different lesions have the same density. So histological evaluation is crucial for making a proper diagnosis. Castleman's disease is an uncommon, benignant lymphoproliferative disease, at first explained by Castleman and Towne in 1954 [1]. The disease has three main subtypes: hyaline vascular, plasma cell and mixed variants [2]. The disease is often seen as a single mass in the mediastinum, but it may be seen in the abdomen, neck or other anatomical sites [3]. Rare cases of Castleman's disease have been reported in the spine [2]. There have been previous reports of Castelman's disease in the thoracic spine and lumbosacral involvement in Castelman's disease is actually rare. Here we present a patient with Castleman's disease of the lumbosacral spine. In addition regarding the rarity of the reported cases, there is no consensus on proper treatment.

CASE REPORT

A 50 year old male presented to our hospital with chief complaint of buttock anesthesia and low back pain from 2.5 years ago. The patient's symptoms were progressive and had recently become unbearable. There was nothing significant about his past medical history. General physical examination revealed an afebrile otherwise healthy man with no evidence of peripheral lymphadenopathies or hepatosplenomegaly. Neurologic evaluation showed saddle anesthesia. Laboratory results including tumor markers were within normal limits. Non contrast sagittal T1, sagital T1 W with contrast, T2 weighted and axial T1 weighted with contrast images respectively, demonstrated a soft tissue extradural mass in the spinal canal. It was isointense in comparison to cord. In postcontrast images a homogeneouse enhancing mass in the spinal canal with extension to the left foraminal and paravertebral areas was seen that resulted nerve root compression and canal stenosis (Figure 1). A sagital CT scan demonstrated a mass in the spinal canal with scalloping at posterior of S2 vertebral body without any bone invasion (Figure 2). Based on the imaging findings, the following differential diagnoses were made: nerve sheath tumor including neurofibroma, paraganglioma and lymphoma. 18F-fluoro-2-deoxy-D-glucose positron emission tomography showed no radiotracer uptake by the mentioned mass and there was no other lesions elsewhere in the body .The patient underwent a CT guided needle biopsy and the specimen sent for histological examination. The sample consisted of 3 pieces of cylindrical cream soft tissue each measuring 1 cm in the length and 0. 2 cm in diameter. Microscopic slides showedlymphoid tissue with large follicles containing small regressed

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germinal centers and hyperplastic mantle zones. A few follicles contained double germinal centers. Germinal centers were composed mostly of FDCs. Some follicles showed so-called lollipop features. There was prominent vasculature in the interfollicular areas. Plasma cells population was inconspicuous (Figure 3). Immunohistochemical staining revealed polytypic B cells and T cells and increased FDCs in involuted germinal centers (Figure 4). These findings were compatible with the diagnosis of Castleman's disease, hyaline vascular type. The patient was referred to a hematologist for treatment and received rituximab at a dose of 375 mg/m^2 with a total dose of 800 mg per week for up to three doses. Thereafter pain was dramatically relieved. It is scheduled to take this dose every 21 days and then MRI will be performed.

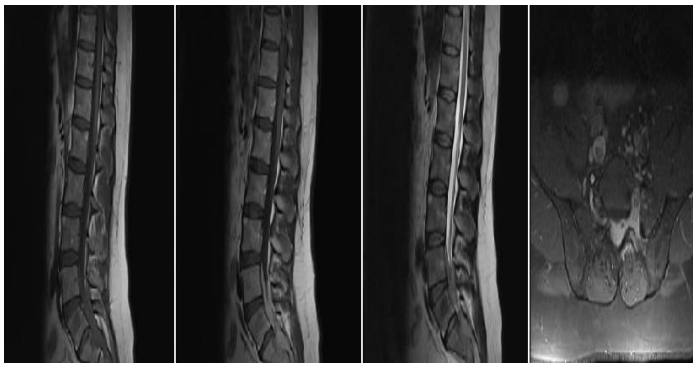

Figure 1: Non contrast sagittal T1 sagital T1 W with contrast, T2 weighted and axial T1 weighted with contrast images respectively, demonstrate soft tissue extradural mass in the spinal canal. It is isointense in comparison to cord. In postcontrast imagesa homogeneouse enhancing mass in the spinal canal with extention to left foraminal and paravertebral is seen that result nerve root compression and canal stenosis

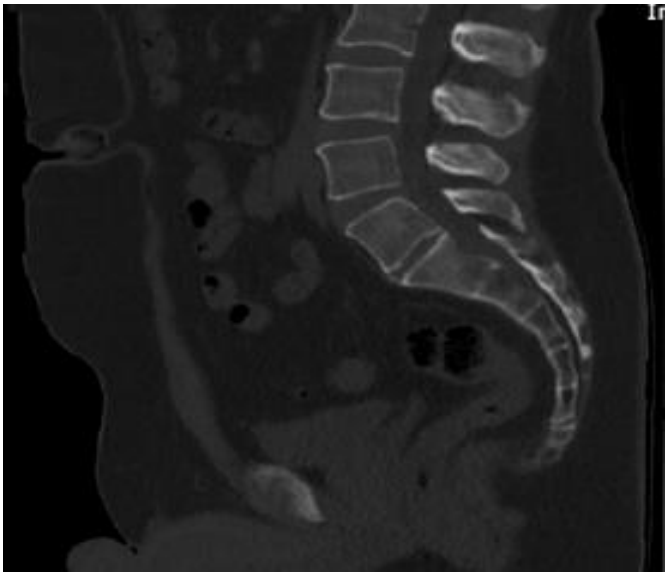

Figure 2: Sagital CT scan shows a mass in the spinal canal with scalloping at posterior of S2 vertebral body without any bone invasion

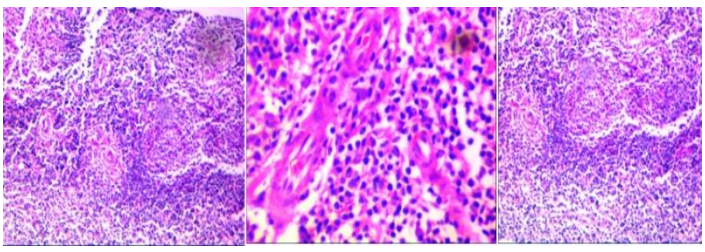

Figure 3: Microscopic slides showed lymphoid tissue with large follicles containing small regressed germinal centers and hyperplastic mantle zones. A few follicles contained double germinal centers (H&E stain)

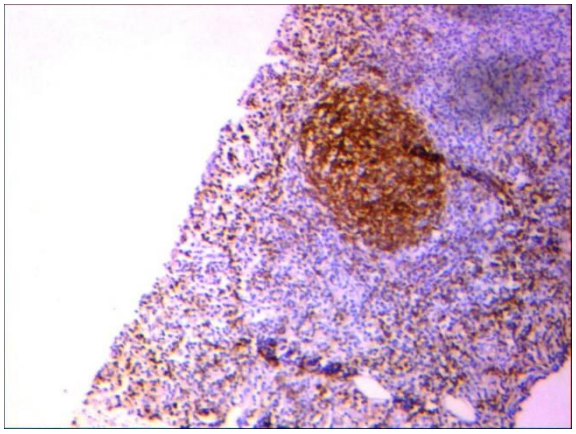

Figure 4: Immunohistochemical staining revealed increased FDCs in involuted germinal centers (IHC stain)

DISCUSSION

Various non-neoplastic spinal masses can mimic the tumor and make it difficult to assess the patient. Benjamin Castleman explained a peculiar but benignant disease in 1954. It was determined by lymphoid tissue hyperplasia [4]. The most common type, unicentric-form, is seen in men and women between the ages of 20 and 30. It can be asymptomatic or have a compressive effect related to the mass [2]. The disease has been reported in nodal or extra nodal sites. In most cases, Castleman disease is seen in the chest, although it can be seen in other parts of the body [5]. Castleman disease, which affects the spine, is very rare and only a few cases have been reported so far [6-10]. In four of these patients have been reported, there were myelopathic symptoms due to cord compression. The disease should be considered in the differential diagnosis of extramedullary masses such as meningioma, pyogenic abscess, schwannoma, neurofibroma, paraganglioma, sarcoidosis, and lymphoma. Its etiology is unknown [1]. On MRI, Castleman disease display mild hyperintensity in T1W and T2W imaging in comparison with skeletal muscle [11]. MRI is exclusively useful to determine the extent of the mass and its communication to adjoining tissues [12]. It is said that in FDG-PET/CT, Castleman disease revealed just moderate radiotracer uptake [13]. On the other hand, malignant lymphomas exhibit higher mean radiotracer uptake [14]. However, there are some degrees of overlap regarding radiotracer uptake between Castleman disease and malignant lymphoma. In the presence of internal calcification, necrosis, or fibrosis, Castleman disease may show a heterogeneous appearance on CT [15]. Imaging methods cannot establish a definitive diagnosis. As a result a histological examination is substantial for confirming an undoubted diagnosis. Although there are a few reports of the effectiveness of steroid therapy and cytotoxic drugs [16], surgical management has been introduced as modulus treatment. A complete surgical excision is preferable for the reason of infiltrative nature of growth of the lesion. Neo-adjuvant radiotherapy can be applied for unrespectable masses. Because secondary malignancy such as non-Hodgkin's lymphoma and Kaposi's sarcoma are more likely in these patients than general population, continuous monitoring of them is essential [6]. The current patient received rituximab and pain was dramatically relieved.

CONCLUSION

Castleman disease is an uncommon disease that should be kept in mind in the differential diagnosis of a patient with an extramedullary mass. Preoperative diagnosis of Castleman's disease-especially when located in extramedullary area is hard due to its rarity and nonspecific imaging results.

Acknowledgement

The authors acknowledge centers and individuals that contributed in this case.

Conflict of Interest

None declared.

Financial Support

None declared.

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