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Case Report

Langerhans Cell Histiocytosis of Thoracic Vertebra in an Adult: A Case Report and Review of the Literature

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Highlights

As it is a rarely seen and not well-defined disease in adults, LCH may be considered as a differential diagnosis in patients with osteolytic bone lesions.

Since there is not a standard treatment for patients with multisystem complaints, presenting more cases with this disease will have a great contribution.

Abstract

The diagnosis of Langerhans Cell Histiocytosis (LCH) is mostly seen in patients aged from 1 to 3, and seen as cases that are not well-diagnosed in adult patients. In this study, LCH diagnosis of a 37-years old patient, who was suffering from a severe pain in the thoracic spine for 2 years, and his treatment process is presented as a case. Although it is a rarely seen disease in adults, it should be kept in mind in the differential diagnosis of this disease and the number of cases should be increased in order to create a standard treatment procedure.

Keywords: Langerhans cell histiocytosis; Thoracic vertebra; Treatment

Introduction

LCH is a rare disease characterized with abnormal proliferation of histiocytes originated from bone narrow with an unknown etiology [1]. It is usually seen in the pediatric age group and defined in three different ways as Eosinopholic Granuloma, Hand Schuller Christıan and Letteree-Siwe according to clinical and pathological characteristics [2]. The disease can be local or systemic and involvement can be seen mostly in skin, bone, lymph nodes, lungs and central nervous systems. Men are affected more often than women [3].

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Case Report

A 37-year-old male patient has applied to the emergency department several times with in the last 2 years with complaints of severe back pain that makes him difficult to breathe and was spreading to the left arm of the patient. According to the evaluations of cardiology and thoracic specialists, routine laboratory tests, chest X-rays, pulmonary function tests of the patient were normal. The severity of the pain has increased as they started to prevent sleep at night and it didn't respond to analgesics. The patient admitted to the neurosurgery clinic. In the thoracic MRI examination, contrasting was detected in T6 and T12 vertebral corpus (Figures 1 & 2) and bone scintigraphy has been proposed. According to the results of bone scintigraphy, findings detected in thoracic 6th vertebrae, right rib lateral and right ischia (Figure 3) suggest a metastatic spread of a primer malign process and it is suggested to be evaluated with PET-BT. As a result of the PET-BT evaluation, the findings detected in patient's skeleton system and liver were found to be suspicious in terms of metastases linked to primer malignancy. In the abdominal ultrasonography, diffuse solid nodular lesions were found in the liver and LAP was lymphadenopathy was detected in the right inguinal. The tumor was excised with spinal tumor ablation system for palliative purposes since there was advanced defect in the patient's vertebra. In this process histopathologic examination was performed. In the pathologic examination, revealed a proliferation of histiocytes with an infiltration of intense eosinophil leucocytes, for CD1a and S-100. So according the results no malignancy has become clear. At the same time, in the Immunohistochemical examination performed on the liver tissue, CD1a and S-100 were found to be positive in the atypical cells and the diagnosis has become clear.

The patient received 12 mg i.v.-Vinblastine every 21 days and oral 80 mg Prednol for 7 days. Osteovita is administrated on the 2nd day of receiving Vinblastine until the 12th day and Zometa 4 mg infusion once a month (Table 1).

After three months of treatment, significant regression was seen in the patient and the treatment protocol created again, he received Vinblastine 12 mg every month, Prednol 80 mg for 5 days from the date of administration Vinblastine and 6-mercaptopurine 100 mg every day for 12 months as the follow-up treatment. During this period, the patient was checked every 3 months and no tumoral symptoms or recurrence was observed. A full recovery was seen. The patient was smoking 1 pack a day before being diagnosed with LCH. Despite recommendations, he didn't quit smoking during treatment (Figure 1).

Discussion

LCH is considered to be a pediatric disease and it is estimated annual prevalence of 1 case per 560,000 in adults [4]. Its etiology is not yet known and it is still controversial whether it is a neoplastic process or an atypical immunological reaction [5]. According to the studies conducted, the recent findings towards mutation of BRAF [6] and MAP2K1 [7] suggest that the disease has a neoplastic origin [5].

It is reported that the diagnosis of LCH is sometimes difficult and it can be diagnosed lately or even be unnoticed in the adult population

Drugs	Drug Application Methods	1st Day	2nd Day	3rd Day	4th Day	5th Day	6th Day	7th Day
Prednol 40 mg/m²	Oral	+	+	+	+	+	+	+
Vinblastine 6 mg/m²	I.V. Push	+						
Osteovita	Oral		+	+	+	+	+	+ ..For 12 days

Table 1: Treatment Protocol.

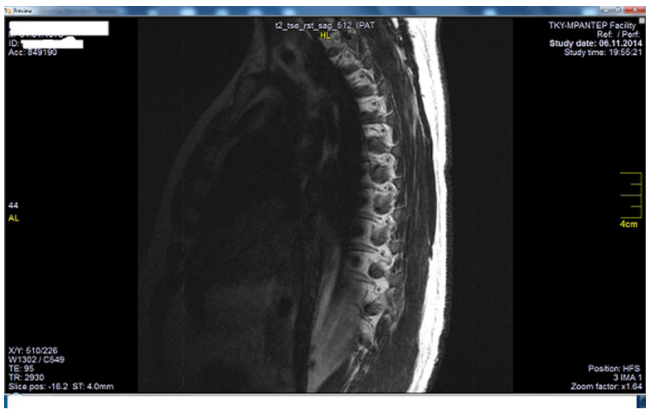

Figure 1: Thorocal MR.

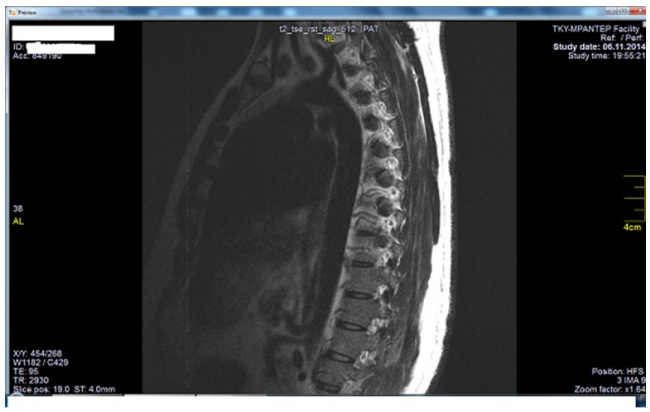

Figure 2: Thorocal MR.

in particular [1,5]. Although PET-BT application is useful in the diagnosis, it has been reported to be insufficient to establish a final diagnosis [8]. There are not any symptoms, physical examinations or laboratory findings that are specific to the disease, and it is diagnosed by immunohistochemical analysis. The definitive diagnosis is obtained by showing the Langerhans cells stained with S-100 and CD1a antigen in the biopsy [9,10]. These cells are mostly eosinophils, leukocytes, neutrophil leukocytes, lymphocytes or plasma cells [10].

Three different clinical forms have been described as Eosinopholic Granuloma (EG), Hand Schuller Christıan and Letteree-Siwe. EG is characterized by either a single or a large number of bone lesions and it can involve all the bones, especially the skull and it is mostly reported to be involved in pelvis and ribs [11]. Hand Schuller Christıan and Letteree-Siwe are more aggressive forms that holds in visceral organs such as lung, liver and skin. The single organ involvement is reported as second skin after bone involvement [14]. Isolated skin LCH has a good prognosis in general, however reactivation in the skin or progression to the disseminated, sometimes fatal, form can also occur [1].

In our case, due to the complaints of the patient, thoracic MRI scan was performed and contrasting patterns were observed in the T6 vertebra corpus, T12 vertebra inferior in a smaller size and in the level of spinal cord right postero lateral segment on the level of T7-8 disc in millimeter-level. According to the bone scan results, suggested the metastatic spread of a malignant process; thus, PET-BT is proposed to be used for further evaluation. Reconstructive surgery applications are recommended in large bone defects [13]. Due to advanced defects in vertebrae, spinal tumor ablation was applied on our patient and the diagnosis was clarified by the pathology results.

Treatment of LCH varies depending on whether the disease is localized or widespread and the surgery, radiotherapy, local and systemic steroid therapy and chemotherapy may be used [14]. LCH is examined in two groups as a single system or multisystem involvement depending on the organ involvement [9,12,15]. In our patient, LCH was diagnosed by the pathology received from liver outside the thoracic vertebrae. According to the LCH classification, the patient was considered to be in the high risk group since he had multisystem involvement. Patients with multisystem involvement are recommended to receive combined treatments and they usually receive both systemic chemotherapy and systemic steroid therapy [9,15]. Due to the lack of a wide range of treatment models in the literature, different treatment models are applied in different centers. Inci et al. have observed a significant regression in a patient with lung and bone involvement after applying 12 mg/week i.v. Vinblastine, 80 mg/ day Metilprednizolol, 100 mg/ day 6-merkaptopürin for 3 months [9]. It is similar to our patient treatment plan in this review.

Conclusion

In this study, diagnosis and treatment of LCH disease in thoracic vertebrae and soft tissues of adult patient was presented. LCH is a rare disease of unknown etiology. Radiological tests help in its finding and immunohistochemical analysis are confirmatory. LCH syndromes have a relatively bening course in adult patients and treatment has got good results. Due to it is a rarely seen and not well-defined disease in adults, LCH may be considered as a differential diagnosis in patients with osteolytic bone lesions. Since there is not a standard treatment for patients with multisystem complaints, presenting more cases with this disease will have a great contribution.

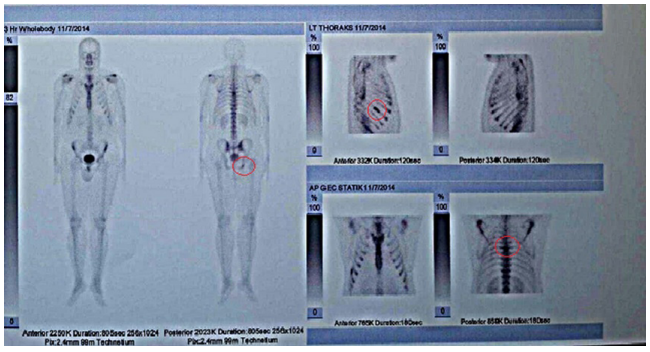

Figure 3: Bone Scintigraphy.

Conflict of Interest

The author(s) declare no potential conflicts of interest, with respect to the research, authorship, and/or publication of this article.

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