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Primary extra nodal Hodgkin disease: Bone presentation

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Summary	
Background:	Extra nodal and extra lymphatic propagation of Hodgkin's disease is a characteristic of the fourth stage of disease when the organs are affected. Primary appearances of the disease outside the lymph node is a rare event. Therefore, it makes diagnostic problem. Skeletal system is possible localization of primary extra nodal Hodgkin's disease.
Case Report:	Women, 42-years-old, was admitted to hospital because of swelling and pain in the right shoulder. After imaging and histological examination diagnosed Hodgkin's nodular sclerosing histological subtype disease has been established. The patient starts to receive chemotherapy.
Conclusions:	Primary extra nodal Hodgkin's disease of bone is manifested with painful swelling in geared area. Imaging method shows destruction of the affected bone, with swelling of the soft tissues. Propagation in soft tissue is not accompanied by their destruction, but rather manifested swelling of the surrounding soft tissue.
Keywords:	Hodgkin's Disease • Painful Shoulder • Bone Disease
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Background

Hodgkin (HL) extranodal presentation is a terminal form of this disease, with lymphoma spreading outside the lymphatic tissue.

Hodgkin's disease may involve every organ, but the most common extranodal location of HL is digestive tract, lungs, central nervous system, bone tissue [1]. The primary manifestation of Hodgkin's disease outside the lymph nodes (that remain normal in such cases), is extremely rare [2]. Primary extranodal presentation is noted in 0.25% of patients with HL [3]. Until today, 19 cases with primary manifestation of HL in bone have been published in the literature. Twelve patients had solitary lesions and seven presented with multifocal lesions in the bones. The patients had no affected lymph nodes [4]. We presented a patient with this rare manifestation of HL together with diagnostic options.

Case Report

A woman, 42-years-old, admitted for examination due to swelling and pain in the right shoulder. The symptoms

started a few months ago. There was no positive response to standard pain-relieving therapy. Radiography of the right shoulder joint was performed (Figure 1). Because of the observed changes, CT (Figure 2) and MRI (Figure 3) of the right shoulder region were performed. Imaging found osteolysis and destruction of acromion of the right scapula, as well as expansion of the process into the supraspinatus region of the right shoulder. No other abnormal focal uptake was found in the rest of the body.

Biopsy and histology examination were performed. Pathological diagnosis was Hodgkin's lymphoma subtype nodular sclerosing. CT examination of the thorax and abdomen to detect enlarged lymph node was negative. Chemotherapy was initiated. After three sessions of chemotherapy, which continues until now, clinical improvement was noted (reduced swelling in the right shoulder and pain reduction). Symptomatic therapy and chemotherapy were continued.

Discussion

Clinically, the primary form of extra-nodal HL presents as intermittent pain that lasts for months. Local swelling



Figure 1. Radiography of the shoulder. White arrow is pointed at the right scapula acromion. See the destruction and the fields of sclerosis.



Figure 2. CT oblique section through both acromions of scapula. White arrow is pointed at the right scapula acromion. There is expansion of the right-sided acromion. Osteolysis with field of osteosclerosis are seen. Lacks its distal portion of right acromion. It is noticeable swelling in the region of supraspinatus muscle from the right side. Left acromion seems normal.

and palpable mass occur. Common symptoms of lymphoma such as weight loss and fever may be helpful for diagnosis [5]. Unlike skeletal expansion of Hodgkin's disease (which is painless), primary bone Hodgkin's disease is painful [6]. Pain from osteolytic lesions in bones presents a problem for differential diagnosis as it needs to be distinguished from osteomyelitis [7]. Only the biopsy of lesions can lead to a correct diagnosis. Primary bone HL can have a solitary or multifocal form [6]. Solitary form can be easily misinterpreted as a non-neoplastic disease after diagnostic imaging.

Location of solitary primary HL in bones is nonspecific. Primary bone HL lesions were found in spine, pelvis, ribs, mandible, femur, tibia and scapula [6]. Primary HL lesions in the bones cannot be correlated with specific parts of bone, bone types, age, or gender [8]. This makes diagnosis difficult.

Imaging features of bone in Hodgkin's disease are nonspecific [9]. Bone changes are predominantly inhomogeneous

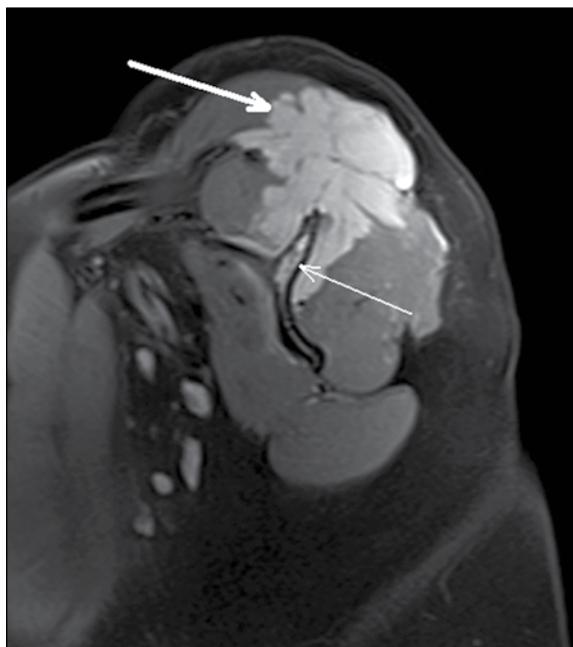


Figure 3. MRI T1 sequence weight. The thin arrow points right acromion that increased signal. Great swelling of the right shoulder muscle mass thick arrow indicates.

osteolytic. Any periosteal reaction, if it occurs, is laminar. Sometimes the chronic form shows osteosclerotic reactions [10]. The lytic-destructive pattern is the most common radiographic appearance of primary bone lymphoma [11]. CT examination shows lesions better than radiography. MRI features of HL lesions include reduced bone marrow signal in the affected region on T1-weighted pulse sequences. On T2-weighted images, regions affected with the process appear bright. Peritumoral reactive marrow edema and lesions can also produce high signal intensity on T2-weighted images. When contrast material is administered, MRI images show areas of enhancement in the lesion [12]. Involved soft-tissue masses around bone lesions on MRI constitute a very common finding [13].

Conclusions

Primary Hodgkin's disease in bones is rare. It is accompanied by pain of the affected region. Imaging method shows destruction of the affected bone, with swelling of soft tissues. Diagnosis is based on histopathology. Prognosis depends on histological forms. Ten-year survival is good.

Conflict of interest

The authors have no conflict of interest.

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