CASE REPORT

Interdigitating dendritic cell sarcoma occurred alone in axilla

Sun Wook Han, Zi Sun Kim, Hyuk Moon Kim, Jihyoun Lee, Gil Ho Kang, Hyun Deuk Cho, Sung Yong Kim, Nae Kyeong Park, Cheol Wan Lim, Min Hyk Lee

Departments of Surgery and 1Pathology, Soonchunhyang University College of Medicine, Cheonan, Korea

Interdigitating dendritic cell sarcoma (IDCS) is a very rare disease around the world and its prognosis is known to be aggressive. This reports a case diagnosed as IDCS of the axillary region treated in Soonchunhyang University Hospital. A 57-year-old female visited Soonchunhyang University Hospital with a left axillary mass. The mass was hard and fixed. Computed tomography observed a 7 cm lymph node at the left axilla, and core biopsy suspected sarcoma. In another study, there was no specific finding except the axillary lesion. Left axillary lymph node dissection (level I, II) was conducted and the pathologic report finally showed IDCS. The patient was treated with only radiotherapy and followed up without recurrence for 13 months up to now. IDCS is a very rare sarcoma that is hard to diagnose and progresses fast. Thus, treatment is very difficult. Proper treatment can be better established after more experiences.

Key Words: Interdigitating dendritic cell sarcoma, Axilla

INTRODUCTION

Dendritic cells are arachnoid immune accessory cells originating on hematopoietic stem cells, and sarcoma on dendritic cells is divided into various subtypes including Langerhans cell sarcoma, interdigitating dendritic cell sarcoma (IDCS), follicular dendritic cell sarcoma, dermal dendrocyte sarcoma, interstitial dendritic cell sarcoma and veiled cell sarcoma [1]. Most IDCS occurs in the lymph node (particularly in the collum, the mediastinum and axilla) and around one third of it is found in other organs except the lymph node. For the organs, the liver is observed most frequently, while others are the spleen, the skin, the lung, the rhinopharynx, the small intestine and the breast [1,2].

The incidence rate of IDCS was reported to be slightly higher in males than in females (1:1.4), and its mean age at onset was about 46 years but it was shown in all age groups (14 to 80 years) [3]. Its prognosis is highly aggressive.

Against this background, a case diagnosed as IDCS of the axilla and treated in Soonchunhyang University Hospital is reported.
CASE REPORT

A 57-year-old female visited Soonchunhyang University Hospital with a left axillary mass palpated for the last two months as a chief complaint. The first physical examination found no specific abnormality such as pain or burning sensation except the palpable mass in the left axilla. The mass was hard and fixed.

Chest computed tomography (CT) showed an approximately 7 cm mass clearly observed in the left axillary region and several lymph nodes enlarged to be 2 cm around the mass (Fig. 1).

Breast ultrasonography and mammography were performed first to determine whether it was breast cancer or not but revealed no mass in the breast and no calcification lesion, respectively. Next, core biopsy found an abnormality suspected to be sarcoma in the lymph node, so additional tests were conducted to check its primary site.

Colonoscopy and gastroscopy revealed no abnormality and abdomen CT also did not reveal a primary site. Bone marrow biopsy to examine bone marrow involvement recorded a negative result. Lastly, positron emission tomography (PET) CT also found no specific abnormality except the increase of 18-fluoro-deoxyglucose in the left axillary region (Fig. 2).

The patient underwent left axillary lymph node dissection (level I, II) as she was thought to have sarcoma originating in the axillary lymph node. Permanent pathologic report finally diagnosed it as IDCS originating in the axillary lymph node (Fig. 3). The microscopy showed ill-defined, paracortically proliferating irregular nests of spindle to ovoid cells in fascicles. The remaining follicles were frequently seen. The tumor cells showed mild to moderate nuclear atypia. Mitosis was rarely seen and necrosis was not present. Immunohistochemistry showed that S-100 protein, vimentin, CD99, CD68 and cyclin D1 were positive and CD34, CD20, CD21, CD23, CD3, HMB-45, desmin and melanin A were negative. Ki-67 or a tumor marker was around 10%.

After the surgery, the patient was treated only with radiotherapy not chemotherapy as adjuvant therapy and recurrence has not been observed for thirteen months up to now.

DISCUSSION

Interdigitating dendritic cells are an antigen presenting cells observed on T cells of the peripheral lymphoid tissues, and sarcoma originating on it is very rare and is histologically similar with B cell lymphoma or T cell lymphoma and other spindle cell neoplasm [2].

Due to this similarity, it is hard to distinguish through hematoxylin and eosin staining, which is conducted frequently for pathologic diagnoses [1]. So, immunohistochemistry is necessary to confirm IDCS. For the sarcoma, S-100 and CD45RB protein, HLA-DR and vimentin are positive and CD1a, CD21, and CD35 protein are negative while CD68 is positive or negative. Another property in differentiating it is its structural characteristics. It has been reported that interdigitating cytoplasmic processes or one characteristic of interdigitating dendritic cells existed on the surface of cells but

Fig. 1. Axillary lymph node enlargement at computed tomography. (A) Arrow shows about 7 cm sized lymph node. (B) Arrow shows about 2 cm sized lymph node around main.