Fibroblastic Reticular Cell Tumor

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Abstract

Review on Fibroblastic reticular cell tumor, with data on clinics, and the genes involved.

Keywords
Fibroblastic Reticular Cell Tumor; Cytokeratin-positive interstitial reticulum cell tumor; Fibroblastic dendritic cell tumor; dendritic cell sarcoma

Identity

Other names
Cytokeratin-positive interstitial reticulum cell tumor, Fibroblastic dendritic cell tumor

Clinics and pathology

Disease

Fibroblastic reticular cell tumor (FRCT) is the rarest subgroup of dendritic cell sarcoma (DCS) with few cases published in the literature (Kwon JE et al.,2009; Dalia S et al., 2013).

Fibroblastic reticular cells are stromal support cells located in the parafollicular are and deep cortex of lymph nodes and in the extrafollicular areas of the spleen and tonsils. (Andriko JW et al., 1998).

In general, FRCT are presents in the lymph nodes, but also in the spleen, lung, liver, and soft tissue (Martel M et al., 2003).

Most of the patients presented with a localized disease, and stage have an important effect in the overall survivor of patients.

Phenotype/cell stem origin

Dendritic/reticular cells are structural and functional accessory cells components of immune system. They have been subdivided on the bases of their location and phenotype into 3 major subsets: follicular dendritic cells (FDCs), interdigitating dendritic cells (IDCs), and fibroblastic reticular cells (FRCs). These cells are stromal cells located in the parafollicular area and deep cortex of lymph nodes and also in the extrafollicular areas of the spleen and tonsils (Doglioni C, et al., 1990). These cells are crucial to the interaction between IDCs and T cells in the primary immune response (Andriko JW et al., 1998).

This pathology was also known as cytokeratin-positive interstitial reticulum cell tumor.

Epidemiology

Median age for FRCT was 61 years old (range 13-80 years) with slight male predilection.

Clincs

Information about FRCT generally comes from series of isolated patients mainly reported in the manuscripts by Saygin C et al and Dalia S et al. Both papers reported that isolated nodal disease is more common (84.2%) when compared to other pathological entities. Cervical and mediastinal lymph nodes were the ones most commonly involved during disease course. Extranodal sites included the liver, lung, kidney, adrenal, bone, and soft tissue. Higher-stage disease has a significantly shorter survival rate. Patients with local disease had a 2-year survival rate of 85.7%; median survival was not reached.
Patients with distant disease died in 2 years and had a median survival rate of one year (Saygin C et al., 2013). Most patients present with a newly diagnosed asymptomatic mass that was surgically removed (Saygin C et al., 2013, Martel M et al., 2003).

**Diagnosis** The value of CT scans, bone marrow biopsy, and other staging work in single nodal disease is unknown, but it becomes important in patients with multiple enlarged lymph nodes. Excisional biopsy of the lymph node is the gold-standard diagnostic method for FRCT.

**Pathology**

Morphologically this disorder is described as spindle to ovoid cells with abundant reticulin staining fibers. Immunohistochemistry is positive for vimentin, desmin factor XIIIa, and smooth muscle actin. CD45RB, CD21, CD35, S100, CD65, and CD1a are negative (Saygin C et al., 2013). Ultrastructural evaluation reveals peripherally located fusiform densities, long cytoplasmic extension, and desmosomal-like intercellular attachments (Dalia S et al., 2013). A subset of fibroblastic reticulum cells that express cytokeratine 8 and 18 and various other epithelial markers has been identified. (Golud VE et al., 1995).

**Treatment**

Surgery is the treatment of choice for patients with localized disease. Limited data exist about the role of the radiotherapy. In localized disease, chemotherapy is not indicated. Patient should be included in clinical trials or referred to specialized care centers.

**References**


This article should be referenced as such: