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Retroperitoneal follicular dendritic cell sarcoma in a young woman: diagnosis and treatment challenges

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Abstract:

Introduction: Follicular dendritic cell sarcoma (FDCC) is an uncommon tumor that usually arises in lymph nodes, especially in the cervical, mediastinal, or axillary areas, but rarely in extranodal sites. Few cases have been reported in English literature so far. The scarcity may be partially due to under-recognition of this entity. Through this case report we analyzed the difficulties of clinical and pathological diagnosis of this rare tumor with its unusual location mistaken it with gynecological cancer's iliac lymph nodes metastases. We also discussed its systemic treatment options.

Case report: A 48-year-old female presented with a loss of weight and epigastralgia. Computed tomography (CT) showed a mass of 5cm of diameter, located close to iliac vessels. Investigation for gynecologic cancers was negative and a partial tumor resection was performed. Pathological examination readdressed the diagnosis of (FDCC). Microscopically, the tumor was composed of a proliferation of spindle to ovoid cells arranged in fascicles, whorls and storiform pattern, accompanied by sprinkling of small lymphocytes. The nuclei of the tumor cells were elongated spindle or ovoid shape with vesicular chromatin and distinct small nuclei. Immunohistochemically, the tumor cells were positive for CD21, CD23 but negative for any type of cytokeratin. Even pathological diagnosis was misleading, therapeutic management was more challenging with this unusual location particularly associated with an aggressive clinical course. Two lines of chemotherapy gave different responses.

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