

Brief report

A case of inflammatory pseudotumor-like follicular/fibroblastic dendritic cell sarcoma

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The patient was female, 79 y/o, and complained abdominal pain and vomiting. Radiographic study revealed several abdominal masses: intra-pelvic tumor (Tp), expansive growth and 8.5cm in diameter, occupied mesentery close to superior mesenteric vein and ileocolic artery, and hepatic expansive tumors occupied both S7 segment (TS7), 6.5cm in diameter, and S8 (TS8), 5.5cm (Fig. 1, 2).

These tumors showed same macroscopic findings: solid, white-and yellow colored, medium-consistency, and spotty necrosis. Microscopic study showed marked inflammatory change and several nodular lesions consisted of polygonal and spindle cells. These nodular areas were found in Tp mass, and showed whorl formation consisted of spindle cells without atypism and separated with thick bands of inflammatory cells around vessels (Fig. 3, 4). Immunohistochemical analysis was positive for CD21, CD35, fascin, and MDM2, which suggested follicular dendritic cell sarcoma (FDCS) (Table 1, Fig. 5). Genetic analysis with both PCR-electrophoresis and real time PCR showed positivity for Epstein-Barr (EBV) infection and negativity for immunoglobulin heavy chain (IgHVDJ) and T cell receptor (TCR) γ .

Based on positive EBV and predilection of aged female patient, "inflammatory pseudotumor-like follicular/fibroblastic dendritic cell (FDC/FRC) sarcoma" was suggested rather than simple "FDC sarcoma".

Key words: nflammatory pseudotumor-like follicular/fibroblastic dendritic cell sarcoma, follicular dendritic cell sarcoma, FDC/FRC, CD21, CD35, fascin, MDM2, Epstein-Barr virus infection, aged female

References

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2. Pileri SA et al. Histiocyte and dendritic cell neoplasms. Fig. 17.01, Table 17.02. In WHO Classification of tumours of haematopoietic and lymphoid tissues. Ed. Swerdlow SH et al. Lyon: International Agency for Research on Cancer, 2017. p466-7.

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和 文 抄 録

短報

炎症性偽腫瘍様濾胞／線維芽細胞肉腫の1症例

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症例は骨盤内腫瘍と肝臓内多発腫瘍を認めた高齢女性で、免疫組織検査で濾胞樹状細胞肉腫が示唆された。高齢・女性、高度の炎症、Epstein-Barr ウイルス感染陽性より、炎症性偽腫瘍様濾胞／線維芽細胞肉腫が妥当と判断された。

キーワード: 炎症性偽腫瘍様濾胞／線維芽細胞肉腫、濾胞樹状細胞肉腫、免疫組織学的検査、FDC/FRC, CD21, CD35, fascin, MDM2, Epstein-Barr ウイルス感染、高齢、女性

Table 1. Immunophenotypic markers of macrophage and dendritic cell (DC)

DC marker	macrophage	dermal/interstitial DC	Langerhans cell	interdigitating DC	plasmacytoid DC	follicular DC	this case
CD21						+	+
CD35						+	+
CD68	+	+			+		
fascin		+		+		+	+
langerin			+				
lysozyme	+						
S100			+	+			
MDM2							+

DC: dendritic cell

Sources: Pileri SA et al. Histiocyte and dendritic cell neoplasms. Fig. 17.01, Table 17.02.

In WHO Classification of tumours of haematopoietic and lymphoid tissues.

Ed. Swerdlow SH et al. Lyon: International Agency for Research on Cancer, 2017. p466-7.

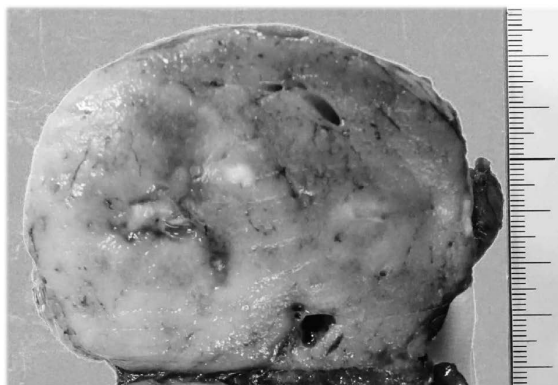


Fig. 1. Intrapelvic tumor (Tp)

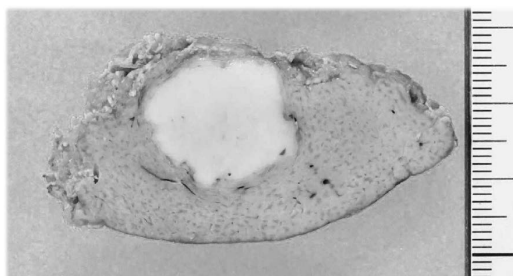


Fig. 2. Hepatic tumor (TS8)

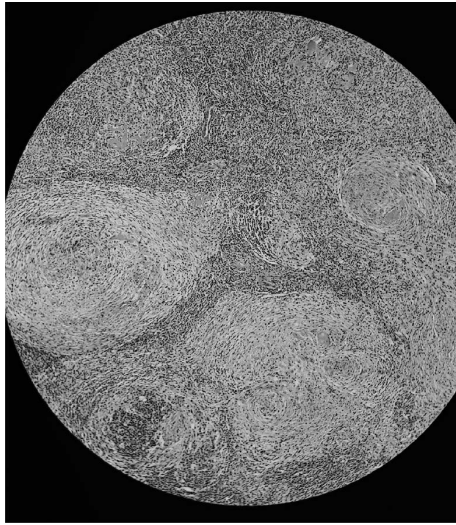


Fig. 3. T_p nodular lesion (Hematoxylin eosin stain, loupe)

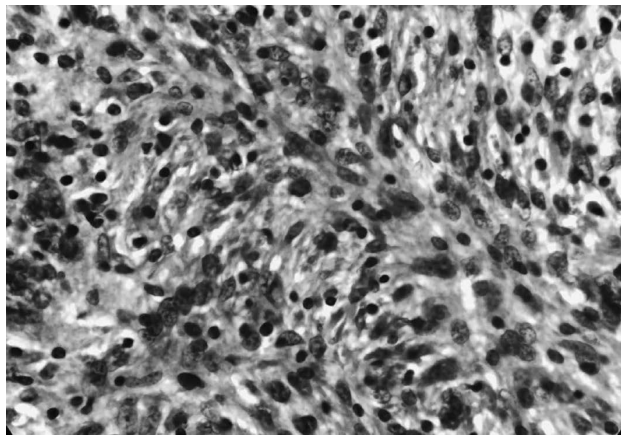


Fig. 4. Whorl formation in Fig. 4 (HE stain)

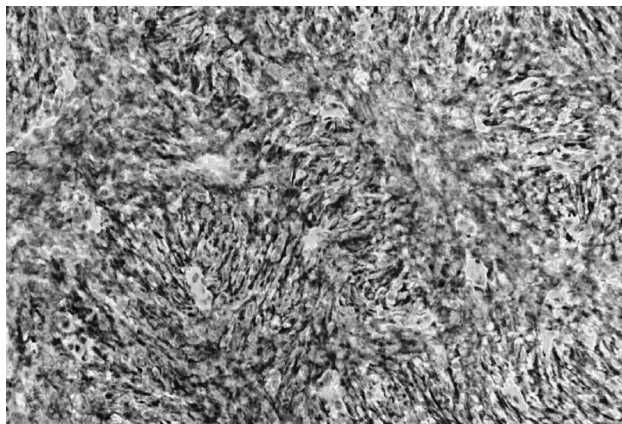


Fig. 5. Positive immunostain for fascin.