Multiorgan involvement in HHV-8-positive multifocal inflammatory myofibroblastic tumor

Multiorgan tutulumu gösteren HHV-8 pozitif inflamatuvar myofibroblastik tümör

To the Editor,

Inflammatory myofibroblastic tumor (IMT) is an uncommon soft-tissue tumor with intermediate behavioral potential.

An 80-year-old male presented with abdominal pain, fever and weight loss. Subsequent to detection of tumoral lesions in his right scrotum and mesentery of the ileum, partial ileum resection and right orchiectomy were performed. In the ileo-cecal region, a solid lesion measuring 11 x 10 x 9 cm, located on the serosal face, with a smooth surface, lobulated appearance and cream-colored section, was observed (Figure 1). Grossly, the scrotal solid tumor neighboring the epididymis was well-circumscribed and measured 2 cm at its greatest diameter. Both lesions located in the ileum and testis tissue had similar histological features. In the microscopic examination, a tumoral lesion comprised of spindle-shaped myofibroblast-like cells and polygonal shaped 'ganglion-like' giant cells was seen. An intense, mixed type inflammatory cellular infiltration was observed in the fibrocollagenous stroma with myxoid changes. In the tumor cells, cytoplasmic reaction was observed with vimentin and SMA, and a nuclear reaction was observed with human herpes virus-8 (HHV-8).

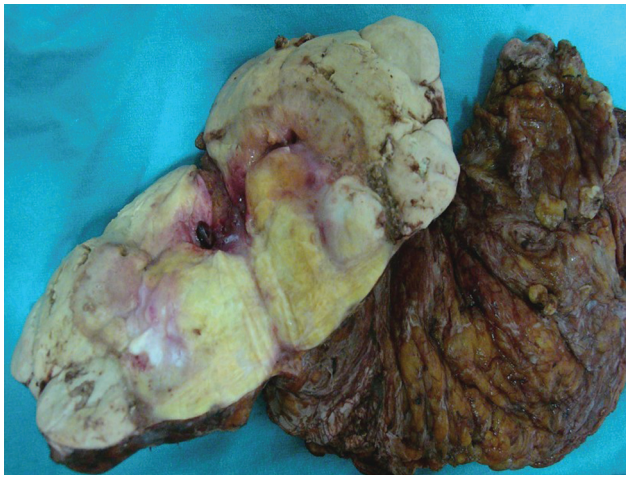

Figure 1. The surgical specimen showed a tumor 11x10x9 cm in size involving the serosal surface of the ileum. The sagittal section showed that the tumor was well-circumscribed, grayish white and firm with foci of hemorrhage.

Inflammatory myofibroblastic tumors (IMTs) are rare lesions with differing clinical and histopathological characteristics. A common histopathological feature of these lesions is spindle cell proliferation with varying amounts of inflammatory infiltration. Ultrastructural studies have shown the myofibroblastic character of the cells that form the lesion (1). IMTs generally occur in children and young adults, with the most common location being the lungs (2). Most cases of IMT reported in the literature are solitary; only a few lesions with a multifocal character have been described (3). In the present case, the lesions were localized to mesenteric and testicular areas, which is unusual for IMT. Moreover, the lesions were present simultaneously. The advanced age of the patient indicates that tumors can occur in various age groups.

The etiology and pathogenesis of IMT are contentious issues. Translocations in ALK, which are found in approximately 50% of cases, suggest that the lesions are true neoplasias, and the dense inflammatory cell content of the tumor and the presence of systemic inflammatory signs in several cases support an underlying infectious etiology (4). In the present case, inflammatory cells were remarkable in both lesions. In both lesions, spindle cells were immuno-positive for HHV-8. Our results suggest that infectious agents may play a part in the etiology of IMTs with predominant inflammatory cells. IMT is rarely diagnosed prior to surgery, and it may clinically and radiologically mimic sarcomas and carcinomas. A definitive diagnosis is reached by histopathological and immunohistochemical examinations (5).

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