Primary hepatic leiomyosarcoma in a young man after Hodgkin's disease: diagnostic pitfalls and therapeutic challenge

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ABSTRACT

Background. Primary leiomyosarcoma of the liver is a rare tumor whose development patterns are unsatisfactorily known.

Patient case. A 26-year-old male patient with a previous history of radiochemotherapy treatment for Hodgkin's lymphoma was referred to our unit with a histological and radiological diagnosis of primary hepatic leiomyosarcoma. Six months before referral, in a workup for hypertension, a CT scan of the abdomen had shown a 2.5-cm lesion in liver segment VII, which was interpreted as an angioma. Shortly before referral the lesion had grown to 7.8 cm, associated with two smaller lesions in segments VIII and III, and a diagnosis of hepatic leiomyosarcoma was made at biopsy. After referral he underwent a right hepatectomy with wedge resection of segment III. This was followed by rapid progression of the disease, in spite of transient stabilization under gemcitabine treatment. Octreotide was also administered after the detection of elevated chromogranin A in serum. The patient died 25 months after liver resection.

Conclusions. The challenges and peculiarities of this case are related to the rarity of the tumor, its accidental discovery without immediate suspicion of its nature, its very aggressive behavior that was only partly controlled by chemotherapy, and the unusual expression of a neuroendocrine phenotypic feature with high serum chromogranin A levels.

Key words: primary hepatic leiomyosarcoma, Hodgkin's disease, liver resection, chromogranin A.

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