Histiocytic Sarcoma

Histiocytic sarcoma (HS) is an extremely rare neoplasia, of hematopoietic origin that accounts for less than 1% of hematologic malignancies. Previously known as “true histiocytic lymphoma”, the tumor follows an aggressive clinical course. According with the World Health Organization classification HS is characterized by the proliferation of malignant cells that have the morphological and immunohistochemical characteristics of mature histiocytes. HS can be diagnosed in all age group, but it is frequently seem in adults [1].

The term HS was first introduced by Mathe et al. [2] based on an analysis of 110 cases of “reticulosarcomas” that were grouped into two varieties: histoblastic and histiocytic types. They noted that both types had the same duration of evolution, but with the difference that the presence of cutaneous lesions was predominant in the histiocytic type.

HS can be localized or disseminated. The majorities of lesions are reported with presentation at extranodal sites, most frequently in the soft tissues and skin [3]. Other anatomical sites and can also be affected including the intestinal tract, the central nervous system, head and neck structures like thyroid and parotid among others [4-6]. Cases of HS associated with malignant leukemia or lymphoma have been also reported, but the nature of this association has not been established [7].

Recent studies have shown that HS has a prominent inflammatory background and is immunoreactive for CD45, CD163, CD68, and lysozyme. The differential diagnosis includes metastatic carcinoma, metastatic melanoma, and large cell non-Hosgkin lymphoma and should be excluded by immunhistochemistry. The CD163, a recently characterized hemoglobin scavenger receptor, appears to be a specific marker of histiocytic lineage and a promising diagnostic tool for HS [8].

Most patients presenting HS are being treated on an individual basis and the outcomes are still poor, particularly in those advanced or disseminated disease. The largest series published of HS treated with upfront surgery with the intent of diagnosis or treatment included 14 patients. All patients presented with a solitary mass with sizes ranging from 1.8 cm to 12 cm. Seven tumors arose in soft tissue, 5 in the gastrointestinal tract, 1 in the nasal cavity, and 1 in the lung. Six patients were treated with postoperative radiation and 7 with chemotherapy (CHOP or PROMACE-MOPP). The follow up was available for 10 patients: 2 of them recurrent locally, and 5 patients developed distant metastasis [9].

The international Lymphoma Study Group stained 61 tumors of suspected histiocytic/dendritic cell type with a panel of 15 antibodies and found 18 malignant histiocytic tumors, with 15 cases of HS and 3 cases of malignant histiocytosis with disseminated disease. In this series there was a predominance of males, adults (median age: 46 years) and extranodal (72%) presentation. The following phenotype was observed: CD68 (100%), LYS (94%), CD1a (0%), S100 (33%), CD21/35 (0%). Nine patients (50%) had stage III or stage IV disease, and seven patients (58%) died of the disease [10].

Despite the rarity of the disease and paucity of data, radiotherapy has being used in some instances with a relative success in the local control. Median radiation doses, fractions of radiotherapy was 60 Gy and 30 fractions, respectively.

Chen et al. [11] recently published a case of a patient with oropharyngeal HS and regional lymph node involvement that was successfully treated with a combination of CHOP-E and adjuvant radiotherapy (50 Gy given in 25 fractions). The patient had no evidence of recurrent disease after 3 years of the end of the treatment.

Recent reviews reported the use of thalidomide in patients with HS after systemic failure [12,13]. The potential mechanisms of antitumoral activity of thalidomide include inhibition of both vascular and fibroblast growth factors, cytokine regulation, apoptosis induction and oxidative DNA damage by free radicals. One review of the use of thalidomide in pediatric patients concluded that it should be used as a last resort when all other therapies fail [14].

Currently there is no standard treatment recommended for HS. The treatment protocols available in the literature are diverse, with most patients being treated on an individual basis. Although HS is
considered a potentially fatal disease, some cases do not pursue such as an aggressive clinical course. Surgery and/or chemotherapy are the most commonly employed treatments, but radiotherapy also seems to be a treatment option in patients with localized disease. Further research is still needed to explore the use of new treatment combinations in this entity.

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