Hairy cell sarcoma

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Summary

The first case of a hairy cell sarcoma is presented. Subsequent to surgical resection and post-operative radiotherapy the patient has survived two years without systemic evidence of hairy cell leukaemia.

KEY WORDS: hairy cell sarcoma, leukaemia, reticuloendotheliosis.

Introduction

Extramedullary solid tumours may precede systemic evidence of leukaemia. They are previously unreported in hairy cell leukaemia.

Case report

A 39-year-old coal merchant presented to his family practitioner with a seven month history of swelling of the left side of his face. He was referred to hospital where a left parotid mass was noted. All routine haematological and biochemical investigations were normal. A needle biopsy of the parotid mass, showed, on light microscopy using mercury based fixation, the presence of cells whose appearance was suggestive of hairy cell leukaemia. The cells failed to stain with acid phosphatase, non-specific esterase, chloroacetate esterase, periodic acid schiff and unna pappenheim/alcan blue. Bone marrow aspiration and trephine showed no evidence of leukaemia. Computed tomography of the chest and abdomen revealed no other deposits. The patient proceeded to surgery, where a mass infiltrating the superficial part of the parotid gland was excised. Electron microscopy showed it to be composed of typical hairy cells (Fig. 1). Post-operatively the parotid area and the whole of the lymphatic drainage of both sides of the neck from basicranium to clavicles was irradiated, using a radiocobalt unit. Lateral fields were applied and tissue compensators used. A tumour dose was delivered of 3500 cGy in 25 fractions over 29 days. Two years after presentation the patient is well without systemic evidence of hairy cell leukaemia.

Discussion

Extramedullary leukaemia was first reported in 1811 when Burns described an ocular chloroma. These large solid deposits should be distinguished from microscopic metastatic leukaemia which is common and observed in over 80% of patients at post-mortem (Hustu and Aur, 1978). Common sites of infiltration include lymph nodes, spleen, testes and the central nervous system (Viadara, Bross and Pickren, 1978). Extramedullary solid tumours are reported as occurring in 8% of acute myeloid and 4% of patients with chronic myeloid leukaemia (Liu *et al.*, 1973). They are collectively termed granulocytic sarcoma. When green they are described as chloromas, when colourless, myeloblastomata (Muss and Moloney, 1973). The pigment may be a function of the presence of a porphyrin-containing enzyme myeloperoxidase (Schulz and Rosenthal, 1959). Their distribution is widespread, most commonly in bone (Liu *et al.*, 1973), rarely in breast (Blackwell *et al.*, 1973), skin (Wiernick and Serpick, 1970), central nervous system (Hurwitz, Sutherland and Walker, 1970), uterus (Hartford, 1968) and ovary (Hinkamp, Evanston and Szanto, 1958). Occurring as an isolated finding, they invariably herald systemic leukaemia and the latency period may range up to two years (Mason, Demanee and Margolis, 1973).

Leukaemic reticuloendotheliosis or hairy cell leukaemia was first described in 1958 by Bouroncle, Wiseman and Doar. The diagnosis is established by the presence of the characteristic 'hairy cell', best visualized by electron microscopy (Katovsky *et al.*, 1974). The disease is rare, exhibits a male preponderance, and has a variably benign course, with a mean survival time of 6 years (Bouroncle, 1979). Treatment is controversial, and the choice may include splenectomy, steroids, chemotherapy and splenic irradiation (Bouroncle, 1979; Catovsky *et al.*, 1974; Davis *et al.*, 1976). Infection is a common complication, and the most frequent cause of death (Bouroncle, 1979).

Extra medullary hairy cell sarcoma has not been previously reported. Its outcome is awaited in this patient.

FIG. 1. Electron micrograph. Arrows indicate intercellular hair projections.

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